

Δημοσιευμένες περιλήψεις (ενδεικτική επιλογή) για την Γιγαντοκυτταρική Αρτηρίτιδα (Giant Cell Arteritis, GCA)

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Δημοσιευμένες περιλήψεις (ενδεικτική επιλογή) για την γίγαντοκυτταρική αρτηρίτιδα (Giant Cell Arteritis, GCA)

1. Use of Noninvasive Imaging in Giant Cell Arteritis.

Halbach C, McClelland CM, Chen J², Li S, Lee MS.

Abstract

Giant cell arteritis (GCA) requires a prompt diagnosis to avoid significant morbidity among the elderly. An accurate diagnosis is also paramount given the side effect profile of long-term corticosteroid treatment. Temporal artery biopsy (TAB) has long remained the gold standard for the diagnosis of GCA but requires an invasive procedure that is not without risk. This article discusses the argument for and against the use of noninvasive imaging including ultrasound, magnetic resonance imaging, and positron emission tomography scanning for the diagnosis of GCA. It also provides a suggested diagnostic algorithm for when to consider noninvasive imaging versus TAB.

Source: Asia Pac J Ophthalmol (Phila). 2018 Jul 13. doi: 10.22608/APO.2018133.

<https://www.ncbi.nlm.nih.gov/pubmed/30003767>

2. Ophthalmic manifestations of giant cell arteritis.

Vodopivec I, Rizzo JF 3rd.

Abstract

GCA, the most common systemic arteritis, affects medium-sized and larger extradural arteries that have the internal elastic lamina. Involvement of the ophthalmic artery and its branches results in visual loss, which is often complete but is usually painless. Visual loss may be monocular or binocular developing simultaneously or sequentially. Rarely, it stems from occipital lobe infarct that result in homonymous hemianopia, a visual field defect involving the two identical halves (right or left) of the visual fields of both eyes. Visual hallucinations and diplopia are less common. All visual symptoms, including those that are transient, require urgent ophthalmological evaluation and treatment with high-dose glucocorticoids to avoid permanent visual loss.

Source: Rheumatology (Oxford). 2018 Feb 1;57(suppl_2):ii63-ii72. doi: 10.1093/rheumatology/kex428.

<https://www.ncbi.nlm.nih.gov/pubmed/29986083>

3. Ultrasound in the diagnosis and management of giant cell arteritis.

Schmidt WA

Abstract

US has become an important diagnostic tool for musculoskeletal diseases. Because of its wide availability in rheumatology practice, US has also been applied in other rheumatic diseases such as GCA. In acute GCA, US displays a non-compressible, hypoechoic, most commonly concentric arterial wall thickening. Temporal and axillary arteries should be examined in patients with suspected GCA and PMR. Additionally, almost all other large arteries, with the exception of the thoracic aorta, can be easily delineated by US. Many studies and several meta-analyses have been conducted to evaluate the diagnostic performance of US. US is more sensitive than temporal artery biopsy (TAB) because TAB evaluates only a limited anatomical region in a systemic disease. Most US studies arrive at specificities between 90 and 100% compared with the final clinical diagnosis. Reliability for reading US images and videos is excellent and comparable to reliability for reading TAB specimens. The advantage of US over other imaging techniques in GCA is its availability, safety and tolerability and its high resolution of 0.1 mm. Rheumatology departments are increasingly establishing fast-track clinics. Physicians can refer patients with suspected GCA within 24 h. Patients receive clinical and US examination by experienced specialists, establishing a clear diagnosis either before TAB or without the need for TAB. The introduction of fast-track clinics has led to a significant reduction of permanent vision loss. Furthermore, a process that primarily includes US is significantly more cost-effective than TAB.

Source: Rheumatology (Oxford). 2018 Feb 1;57(suppl_2):ii22-ii31. doi: 10.1093/rheumatology/kex461
<https://www.ncbi.nlm.nih.gov/pubmed/29982780>

4. Prevention of glucocorticoid morbidity in giant cell arteritis.

Buttgereit F, Matteson EL, Dejaco C, Dasgupta B.

Abstract

Glucocorticoids are the mainstay of treatment for GCA. Patients often require long-term treatment that may be associated with numerous adverse effects, depending on the dose and the duration of treatment. Trends in recent decades for glucocorticoid use in GCA suggest increasing cumulative doses and longer exposures. Common adverse events (AEs) reported in glucocorticoid-treated GCA patients include osteoporosis, hypercholesterolaemia, hypertension, posterior subcapsular cataract, infections, diabetes mellitus, Cushingoid appearance, adrenal insufficiency and aseptic necrosis of bone. AEs considered most worrisome by patients and rheumatologists include weight gain, psychological effects, osteoporosis, cardiometabolic complications and infections. The challenge is to maximize the benefit-risk ratio by giving the maximum glucocorticoid treatment necessary to control GCA initially and then to prevent relapse but to give the minimum treatment possible to avoid glucocorticoid-related AEs. We discuss the safety issues associated with long-term glucocorticoid use in patients with GCA and strategies for preventing glucocorticoid-related morbidity.

Source: Rheumatology (Oxford). 2018 Feb 1;57(suppl_2):ii11-ii21. doi: 10.1093/rheumatology/kex459.
<https://www.ncbi.nlm.nih.gov/pubmed/29982779>

5. Large-vessel giant cell arteritis: diagnosis, monitoring and management.

Koster MJ, Matteson EL, Warrington KJ.

Abstract

GCA is a chronic, idiopathic, granulomatous vasculitis of medium and large arteries. It comprises overlapping phenotypes including classic cranial arteritis and extra-cranial GCA, otherwise termed large-vessel GCA (LV-GCA). Vascular complications associated with LV-GCA may be due, in part, to delayed diagnosis, highlighting the importance of early identification and prompt initiation of effective therapy.

Advancements in imaging techniques, including magnetic resonance angiography, CT angiography, PET and colour duplex ultrasonography, have led to improvements in the diagnosis of LV-GCA; however, the role imaging modalities play in the assessment of disease activity and long-term outcomes remains unclear. Glucocorticoids are the mainstay of therapy in LV-GCA, but their prolonged use is associated with multiple, sometimes serious, adverse effects. Recent data suggest that biologic therapies, such as tocilizumab, may be effective and safe steroid-sparing options for patients with GCA. However, data specifically evaluating the management of LV-GCA are limited.

Source: Rheumatology (Oxford). 2018 Feb 1;57(suppl_2):ii32-ii42. doi: 10.1093/rheumatology/kex424.

<https://www.ncbi.nlm.nih.gov/pubmed/29982778>

6. Pathogenesis of giant-cell arteritis: how targeted therapies are influencing our understanding of the mechanisms involved.

Terrades-Garcia N, Cid MC

Abstract

GCA is a chronic granulomatous vasculitis that affects large- and medium-sized vessels. Both the innate and the adaptive immune system are thought to play an important role in the initial events of the pathogenesis of GCA. Amplification cascades are involved in the subsequent development and progression of the disease, resulting in vascular inflammation, remodelling and occlusion. The development of large-vessel vasculitis in genetically modified mice has provided some evidence regarding potential mechanisms that lead to vascular inflammation.

However, the participation of specific mechanistic pathways in GCA has not been fully established because of the paucity and limitations of functional models. Treatment of GCA is evolving, and novel therapies are being incorporated into the GCA treatment landscape. In addition, to improve the management of GCA, targeted therapies are providing functional proof of concept of the relevance of particular pathogenic mechanisms in the development of GCA and in sustaining vascular inflammation.

Source: Rheumatology (Oxford). 2018 Feb 1;57(suppl_2):ii51-ii62. doi: 10.1093/rheumatology/kex423.

<https://www.ncbi.nlm.nih.gov/pubmed/29982777>

7. Assessing Vasculitis in Giant Cell Arteritis by Ultrasound: Results of OMERACT Patient-based Reliability Exercises.

Schäfer VS, Chrysidis S, DeJaco C, Duftner C, Iagnocco A, Bruyn GA, Carrara G, D'Agostino MA, De Miguel E, Diamantopoulos AP, Fredberg U, Hartung W, Hocevar A, Juche A, Kermani TA, Koster MJ, Lorenzen T, Macchioni P, Milchert M, Døhn UM, Mukhtyar C, Ponte C, Ramiro S, Scirè CA, Terslev L, Warrington KJ, Dasgupta B, Schmidt WA.

Abstract

OBJECTIVE:

To test the reliability of Outcome Measures in Rheumatology Clinical Trials (OMERACT) consensus-based ultrasound definitions for normal and vasculitic temporal and axillary arteries in patients with giant cell arteritis (GCA) and in controls.

METHODS:

A preliminary 1-day meeting and a full 3-day meeting fulfilling OMERACT Ultrasound Group guidelines were held. Temporal and axillary arteries were examined at 2 timepoints by 12 sonographers on 4 patients with GCA and 2 controls. The aim was to test inter- and intrareader reliability for normal findings, halo sign, and compression sign. In both meetings, patients had established GCA. Pathology was more recent in the full meeting, which was preceded by 6 h of training. Scanning time was 15-20 min instead of 10-13 min.

RESULTS:

In the preliminary exercise, interreader reliabilities were fair to moderate for the overall diagnosis of GCA (Light κ 0.29-0.51), and poor to fair for identifying vasculitis in the respective anatomical segments (Light κ 0.02-0.46). Intrareader reliabilities were moderate (Cohen κ 0.32-0.64). In the main exercise, interreader reliability was good to excellent (Light κ 0.76-0.86) for the overall diagnosis of GCA, and moderate to good (Light κ 0.46-0.71) for identifying vasculitis in the respective anatomical segments. Intrareader reliability was excellent for diagnosis of GCA (Cohen κ 0.91) and good (Cohen κ 0.71-0.80) for the anatomical segments.

CONCLUSION:

OMERACT-derived definitions of halo and compression signs of temporal and axillary arteries are reliable in recent-onset GCA if experienced sonographers (> 300 examinations) have 15-20 min for a standardized examination with prior training and apply > 15 MHz probes.

Source: J Rheumatol. 2018 Jul 1. pii: jrheum.171428. doi: 10.3899/jrheum.171428.

<https://www.ncbi.nlm.nih.gov/pubmed/29961687>

8. Predictors of positive ¹⁸F-FDG PET/CT-scan for large vessel vasculitis in patients with persistent polymyalgia rheumatica.

Prieto-Peña D, Martínez-Rodríguez I, Loricera J, Banzo I, Calderón-Goercke M, Calvo-Río V, González-Vela C³, Corrales A, Castañeda S, Blanco R, Hernández JL, González-Gay MÁ.

Abstract

OBJECTIVE:

Polymyalgia rheumatica (PMR) is often the presenting manifestation of giant cell arteritis (GCA). Fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (PET/CT) scan often discloses the presence of large vessel vasculitis (LVV) in PMR patients. We aimed to identify predictive factors of a positive PET/CT scan for LVV in patients classified as having isolated PMR according to well-established criteria.

METHODS:

A set of consecutive patients with PMR from a single hospital were assessed. All of them underwent PET/CT scan between January 2010 and February 2018 based on clinical considerations. Patients with PMR associated to other diseases, including those with cranial features of GCA, were excluded. The remaining patients were categorized in classic PMR (if fulfilled the 2012 EULAR/ACR classification criteria at disease diagnosis; n = 84) or atypical PMR (who did not fulfill these criteria; n = 16). Only information on patients with classic PMR was assessed.

RESULTS:

The mean age of the 84 patients (51 women) with classic PMR was 71.4 ± 9.2 years. A PET/CT scan was positive in 51 (60.7%). Persistence of classic PMR symptoms was the most common reason to perform a PET/CT scan. Nevertheless, patients with positive PET/CT scan often had unusual symptoms. The best set of predictors of a positive PET/CT scan were bilateral diffuse lower limb pain (OR = 8.8, 95% CI: 1.7-46.3; p = 0.01), pelvic girdle pain (OR = 4.9, 95% CI: 1.50-16.53; p = 0.01) and inflammatory low back pain (OR = 4.7, 95% CI: 1.03-21.5; p = 0.04).

CONCLUSION:

Inflammatory low back pain, pelvic girdle and diffuse lower limb pain are predictors of positive PET/CT scan for LVV in PMR.

Source: Semin Arthritis Rheum. 2018 May 18. pii: S0049-0172(18)30215-4. doi: 10.1016/j.semarthrit.2018.05.007.

<https://www.ncbi.nlm.nih.gov/pubmed/29903537>

9. Arterial lesions in giant cell arteritis: A longitudinal study.

Kermani TA, Diab S, Sreih AG, Cuthbertson D, Borchin R, Carette S, Forbess L, Koenig CL, McAlear CA, Monach PA, Moreland L, Pagnoux C, Seo P, Spiera RF, Warrington KJ, Ytterberg SR, Langford CA, Merkel PA, Khalidi NA; Vasculitis Clinical Research Consortium.

Abstract**OBJECTIVES:**

To evaluate large-vessel (LV) abnormalities on serial imaging in patients with giant cell arteritis (GCA) and discern predictors of new lesions.

METHODS:

Clinical and imaging data from patients with GCA (including subjects diagnosed by LV imaging) enrolled in a prospective, multicenter, longitudinal study and/or a randomized clinical trial were included. New arterial lesions were defined as a lesion in a previously unaffected artery.

RESULTS:

The study included 187 patients with GCA, 146 (78%) female, mean (\pm SD) age at diagnosis 68.5 ± 8.5 years; 39% diagnosed by LV imaging. At least one arterial lesion was present in 123 (66%) on the first study. The most frequently affected arteries were subclavian (42%), axillary (32%), and thoracic aorta (20%). In 106 patients (57%) with serial imaging, new arterial lesions were noted in 41 patients (39%), all of whom had a baseline abnormality, over a mean (\pm SD) follow-up of 4.39 (2.22) years. New abnormalities were observed in 33% patients by year 2; clinical features of active disease were present at only 50% of these cases. There were no differences in age, sex, temporal artery biopsy positivity, or disease activity in patients with or without new lesions.

CONCLUSIONS:

In this cohort of patients with GCA, LV abnormalities on first imaging were common. Development of new arterial lesions occurred in patients with arterial abnormalities at first imaging, often in the absence of symptoms of active disease. Arterial imaging should be considered in all patients with GCA at diagnosis and serial imaging at least in patients with baseline abnormalities.

Source: Semin Arthritis Rheum. 2018 May 9. pii: S0049-0172(18)30094-5. doi: 10.1016/j.semarthrit.2018.05.002.

<https://www.ncbi.nlm.nih.gov/pubmed/29880442>

10. Current and emerging diagnosis tools and therapeutics for giant cell arteritis.

González-Gay MA, Pina T, Prieto-Peña D, Calderon-Goercke M, Blanco R, Castañeda S.

Abstract

Giant cell arteritis (GCA) is the most common large-vessel vasculitis in individuals older than 50 years from Western countries. The goal of the treatment is to achieve improvement of symptoms and clinical remission as well as decrease the risk of severe vascular complications. Areas covered: The review summarizes the main epidemiological and clinical features of GCA and discusses in depth both the classic and the new therapies used in the management of GCA. Expert commentary: Prednisone/prednisolone of 40-60 mg/day is the mainstay in GCA therapy. It yields improvement of clinical features and reduces the risk of permanent visual loss in patients with GCA. Other drugs are used in patients who experience relapses (flares of the disease) or side effects related to glucocorticoids. Methotrexate is the most common conventional immunosuppressive drug used as a glucocorticoid sparing agent. Among the new biologic agents, the most frequently used is the recombinant humanized anti-IL-6 receptor antibody, which is effective to improve clinical symptoms, decrease the cumulative prednisone dose, and reduce the frequency of relapses in these patients. Antitumor necrosis factor- α therapy is not useful in GCA. Experience with other biologic agents, such as abatacept or ustekinumab, looks promising but it is still scarce.

Source: Expert Rev Clin Immunol. 2018 Jul;14(7):593-605. doi: 10.1080/1744666X.2018.1485491.

<https://www.ncbi.nlm.nih.gov/pubmed/29877748>

11. Definitions and reliability assessment of elementary ultrasound lesions in giant cell arteritis: a study from the OMERACT Large Vessel Vasculitis Ultrasound Working Group.

Chrysidis S, Duftner C, Dejaco C, Schäfer VS, Ramiro S, Carrara G, Scirè CA, Hocevar A, Diamantopoulos AP, Iagnocco A, Mukhtyar C, Ponte C, Naredo E, De Miguel E, Bruyn GA, Warrington KJ, Terslev L, Milchert M, D'Agostino MA, Koster MJ, Rastalsky N, Hanova P, Macchioni P, Kermani TA, Lorenzen T, Døhn UM, Fredberg U, Hartung W, Dasgupta B, Schmidt WA.

Abstract

OBJECTIVES:

To define the elementary ultrasound (US) lesions in giant cell arteritis (GCA) and to evaluate the reliability of the assessment of US lesions according to these definitions in a web-based reliability exercise.

METHODS:

Potential definitions of normal and abnormal US findings of temporal and extracranial large arteries were retrieved by a systematic literature review. As a subsequent step, a structured Delphi exercise was conducted involving an expert panel of the Outcome Measures in Rheumatology (OMERACT) US Large Vessel Vasculitis Group to agree definitions of normal US appearance and key elementary US lesions of vasculitis of temporal and extracranial large arteries. The reliability of these definitions on normal and abnormal blood vessels was tested on 150 still images and videos in a web-based reliability exercise.

RESULTS:

Twenty-four experts participated in both Delphi rounds. From originally 25 statements, nine definitions were obtained for normal appearance, vasculitis and arteriosclerosis of cranial and extracranial vessels. The 'halo' and 'compression' signs were the key US lesions in GCA. The reliability of the definitions for normal temporal and axillary arteries, the 'halo' sign and the 'compression' sign were excellent with inter-rater agreements of 91-99% and mean kappa values of 0.83-0.98 for both inter-rater and intra-rater reliabilities of all 25 experts.

CONCLUSIONS:

The 'halo' and the 'compression' signs are regarded as the most important US abnormalities for GCA. The inter-rater and intra-rater agreement of the new OMERACT definitions for US lesions in GCA was excellent.

Source: *RMD Open*. 2018 May 17;4(1):e000598. doi: 10.1136/rmdopen-2017-000598.

<https://www.ncbi.nlm.nih.gov/pubmed/29862043>

12. Primary Care Vasculitis: Polymyalgia Rheumatica and Giant Cell Arteritis.

Pioro MH.

Abstract

Polymyalgia rheumatica (PMR) and giant cell arteritis (GCA) are related inflammatory diseases of adults aged 50 years or older. The diagnosis of PMR is based on morning stiffness, proximal shoulder and pelvic girdle pain, and functional impairment. GCA is characterized by headache, jaw claudication, and visual disturbances. Constitutional symptoms and elevated inflammatory markers are common to both conditions. Temporal artery biopsy remains the gold standard for diagnosis of GCA. Glucocorticoids are the cornerstone of therapy, with tapering regimens individualized to the patient. Prompt diagnosis and treatment are essential to avert vision loss in GCA. Tocilizumab increases remission rates in GCA.

Source: *Prim Care*. 2018 Jun;45(2):305-323. doi: 10.1016/j.pop.2018.02.007.

<https://www.ncbi.nlm.nih.gov/pubmed/29759126>

13. Risk Associated with Cumulative Oral Glucocorticoid Use in Patients with Giant Cell Arteritis in Real-World Databases from the USA and UK.

Gale S, Wilson JC, Chia J, Trinh H, Tuckwell K, Collinson N, Dimonaco S, Jick S, Meier C, Mohan SV, Sarsour K.

Abstract

INTRODUCTION:

Treatment of giant cell arteritis (GCA) involves immediate initiation of high-dose glucocorticoid therapy with slow tapering of the dose over many months. Chronic exposure to glucocorticoids is associated with serious comorbidities. The objective of this analysis was to determine the glucocorticoid exposure and risk of glucocorticoid-related adverse events (AEs) in real-world patients with GCA.

METHODS:

Data from the Truven Healthcare MarketScan® database (from January 1, 2000, to June 30, 2015) and the Clinical Practice Research Datalink (CPRD; from January 1, 1995, to August 31, 2013) were used to retrospectively analyze patients aged ≥ 50 years with GCA in the USA and UK, respectively. Outcomes included oral glucocorticoid use (cumulative prednisone-equivalent exposure), glucocorticoid-related AEs and the association of AE risk with glucocorticoid exposure over 52 weeks.

RESULTS:

Of the 4804 patients in the US MarketScan database and 3973 patients in the UK CPRD database included, 71.3 and 74.6% were women and mean age was 73.4 and 73.0 years, respectively. Median starting glucocorticoid dose and cumulative glucocorticoid dose at 52 weeks were 20-50 mg/day and 4000-4800 mg, respectively. The most frequent glucocorticoid-related AEs were hypertension and eye, bone health, and glucose tolerance conditions. In the first year after diagnosis, the likelihood of any glucocorticoid-related AE was significantly increased for each 1 g increase in cumulative glucocorticoid dose in the US and UK cohorts (odds ratio [95% CI], 1.170 [1.063, 1.287] and 1.06 [1.03, 1.09], respectively; $P < 0.05$ for both). Similar trends were observed for the risk of glucocorticoid-related AEs over full follow-up (mean, USA: 3.9 years, UK: 6.3 years).

CONCLUSIONS:

In real-world patients with GCA, increased cumulative glucocorticoid exposure was associated with an increased risk of glucocorticoid-related AEs.

FUNDING:

F. Hoffmann-La Roche Ltd. Plain language summary available for this article.
Source: Rheumatol Ther. 2018 May 11. doi: 10.1007/s40744-018-0112-8.
<https://www.ncbi.nlm.nih.gov/pubmed/29752705>

14. EULAR recommendations for the use of imaging in large vessel vasculitis in clinical practice.

Dejaco C, Ramiro S, Duftner C, Besson FL, Bley TA, Blockmans D, Brouwer E, Cimmino MA, Clark E, Dasgupta B, Diamantopoulos AP, Direskeneli H, Iagnocco A, Klink T, Neill L, Ponte C, Salvarani C, Slart RHJA, Whitlock M, Schmidt WA.

Abstract

To develop evidence-based recommendations for the use of imaging modalities in primary large vessel vasculitis (LVV) including giant cell arteritis (GCA) and Takayasu arteritis (TAK). European League Against Rheumatism (EULAR) standardised operating procedures were followed. A systematic literature review was conducted to retrieve data on the role of imaging modalities including ultrasound, MRI, CT and [^{18}F]-fluorodeoxyglucose positron emission tomography (PET) in LVV. Based on evidence and expert opinion, the task force consisting of 20

physicians, healthcare professionals and patients from 10 EULAR countries developed recommendations, with consensus obtained through voting. The final level of agreement was voted anonymously. A total of 12 recommendations have been formulated. The task force recommends an early imaging test in patients with suspected LVV, with ultrasound and MRI being the first choices in GCA and TAK, respectively. CT or PET may be used alternatively. In case the diagnosis is still in question after clinical examination and imaging, additional investigations including temporal artery biopsy and/or additional imaging are required. In patients with a suspected flare, imaging might help to better assess disease activity. The frequency and choice of imaging modalities for long-term monitoring of structural damage remains an individual decision; close monitoring for aortic aneurysms should be conducted in patients at risk for this complication. All imaging should be performed by a trained specialist using appropriate operational procedures and settings. These are the first EULAR recommendations providing up-to-date guidance for the role of imaging in the diagnosis and monitoring of patients with (suspected) LVV. **Source:** Ann Rheum Dis. 2018 May;77(5):636-643. doi: 10.1136/annrheumdis-2017-212649. Epub 2018 Jan 22.

15. **Why do temporal arteries go wrong? Principles and pearls from a clinician and a pathologist.**

Banz Y, Stone JH.

Abstract

Early diagnosis and treatment of GCA is essential to prevent complications of the disease, including permanent vision loss. Temporal artery biopsy has been intrinsically linked with the diagnosis of GCA for several decades. A negative predictive value of > 90% has been reported for temporal artery biopsy; however, a negative result does not reliably indicate the absence of GCA because inflammation of the temporal artery is not always evident because of segmental involvement or other reasons. This is demonstrated by a case study of a patient hospitalized following acute vision loss to the right eye whose glucocorticoid treatment was suspended after temporal artery biopsy revealed no evidence of GCA. The patient subsequently lost sight in the left eye 6 weeks after stopping glucocorticoid therapy. The specificity of temporal artery biopsy for the diagnosis of GCA is variable and influenced by many factors, including length of biopsy specimens, vasculitis in vessels other than the temporal artery (ophthalmic, retinal and posterior ciliary vessels), unilateral versus bilateral biopsy, expertise of the surgeon, interpretation of histology, effects of treatment and confounding factors such as atherosclerosis or other non-GCA diseases that can affect the temporal artery. Considering the limitations of temporal artery biopsy, collaboration and education between the clinician, the pathologist and the patient, taking into account a thorough examination of patient history, recognizing signs and symptoms, and potentially involving newer imaging studies with trained technicians and physicians, are essential in confirming or eliminating diagnosis of GCA. **Source:** Rheumatology (Oxford). 2018 Feb 1;57(suppl_2):ii3-ii10. doi:10.1093/rheumatology/kex524.

16. **Long-term glucocorticoid treatment in patients with polymyalgia rheumatica, giant cell arteritis, or both diseases: results from a national rheumatology database.**

Albrecht K, Huscher D, Buttgereit F, Aringer M, Hoese G, Ochs W, Thiele K, Zink A.

Abstract

The objective of this study was to evaluate glucocorticoid (GC) use in patients with polymyalgia rheumatica (PMR), giant cell arteritis (GCA) or both diseases (PMR + GCA) under rheumatological care. Data from patients with PMR (n = 1420), GCA (n = 177) or PMR + GCA (n = 261) from the National Database of the German Collaborative Arthritis Centers were analyzed regarding GCs and related comorbidities (osteoporosis, diabetes and cardiovascular disease), stratified by disease duration (DD). Longitudinal data were analyzed for all patients with a DD ≤ 2 years at database entry (n = 1397). Three-year data were available for 256 patients. Predictors of GC use ≥ 3 years were examined by logistic regression analyses. A total of 76% received GCs, and 19% (PMR) to 40% (GCA) received methotrexate. Median GC doses were 12.5 mg (PMR), 11.3 mg (GCA), and 20.0 mg/day (PMR + GCA) in a 0-6-month DD. Median GC doses ≤ 5 mg/day were reached at a 13-18-month DD in PMR patients and at a 19-24-month DD in GCA or PMR + GCA patients. In the multivariate analysis, baseline methotrexate (OR 2.03, [95% CI 1.27-3.24]), GCs > 10 mg/day (OR 1.65, [1.07-2.55]), higher disease activity (OR 1.12, [1.02-1.23]) (median 0.6 years DD), and female sex (OR 1.63 [1.09-2.43]) were predictive for GC therapy at ≥ 3 years. Of the examined comorbidities, only osteoporosis prevalence increased within 3 years. GC use for ≥ 3 years was reported in one-fourth of all the patients. A difficult-to-control disease activity within the first year was a good predictor of long-term GC need.

Source: Rheumatol Int. 2018 Apr;38(4):569-577. doi: 10.1007/s00296-017-3874-3. Epub 2017 Nov 9.

17. **Value of temporal artery biopsy length in diagnosing giant cell arteritis.**

Oh LJ, Wong E, Gill AJ, McCluskey P, Smith JEH.

Abstract

BACKGROUND:

Giant cell arteritis (GCA) is considered an ophthalmological emergency with severe sight and life-threatening sequelae. Temporal artery biopsy (TAB) is the current gold standard for the diagnosis of GCA; however, the required length of biopsy remains an issue of contention in the literature.

METHODS:

Retrospective case-control study of a consecutive cohort of 545 patients who had undergone TABs across five hospitals between 1 January 1992 and 1 January 2016. In patients with either positive or negative TABs, we collected age, sex, biopsy length and erythrocyte sedimentation rate (ESR).

RESULTS:

A total of 538 patients were included in the final analysis. Of these, 23.4% of TABs were positive, with the average length being 17.6 mm. There was a significant difference in means for positive (19.9 mm) and negative (16.8 mm) biopsies (P = 0.0009). Each millimetre increase in TAB length increased the odds of a positive TAB by 3.4% (P = 0.024). A cut-off point of ≥15 mm increased the odds of a positive TAB by 2.25 compared with a TAB <15 mm (P = 0.003). We also found that ESR ≥50 mm/h was a very strong predictor for a positive TAB result (P < 0.0001).

CONCLUSION:

Biopsy length and ESR were significant predictors of a pathological diagnosis of GCA. We also found that the optimal length threshold predictive for GCA was 15 mm in order to avoid a false-negative GCA diagnosis. Although TAB remains the gold standard for diagnosis, clinicians should refer to both clinical and pathological data to guide their management.

Source: ANZ J Surg. 2018 Mar;88(3):191-195. doi: 10.1111/ans.13822. Epub 2016 Nov 1.

18. Spectrum of Aortic Disease in the Giant Cell Arteritis Population

Kebed DT, Bois JP, Connolly HM, Scott CG, Bowen JM, Warrington KJ, Makol A, Greason KL, Schaff HV, Anavekar NS.

Abstract

We report the spectrum of aortic involvement in patients with giant cell arteritis (GCA) following review of medical records of 4,006 patients including those with imaging studies. A total of 1,450 patients (36%) had a confirmed diagnosis of GCA. Of these, 974 had aortic imaging. Of the 974 patients with imaging, 435 (45%) had an identified aortopathy. The most common aortopathy was aneurysm/dilation (69%). Overall, an annual aneurysmal growth rate of 1.5 mm/y was calculated. In patients with aneurysm/dilation, aortic dissection occurred in 18 patients (6%), and these patients had a significantly higher aneurysmal growth rate compared with those without dissection (4.5 vs 1.4 mm/y, $p = 0.005$). The median size of the aorta at the time of dissection was 51 mm, with 7 (39%) occurring with a maximal aortic aneurysm/dilation <50 mm. In conclusion, our findings indicate higher aneurysmal growth rate in GCA compared with that reported for degenerative aortic disease. Moreover, patients who develop dissection had a significantly higher growth rate than those without dissection with over a third of these patients suffering dissection at a caliber <50 mm.

Source: Am J Cardiol. 2018 Feb 15;121(4):501-508. doi: 10.1016/j.amjcard.2017.11.011. Epub 2017 Nov 23.

19. Comparative effectiveness of ^{18}F -FDG PET-CT and contrast-enhanced CT in the diagnosis of suspected large-vessel vasculitis.

Vaidyanathan S, Chattopadhyay A, Mackie SL, Scarsbrook AF.

Abstract

OBJECTIVE:

Large-vessel vasculitis (LVV) is a serious illness with potentially life-threatening consequences. (^{18}F Fluorine) fluorodeoxyglucose positron emission tomography-computed tomography (^{18}F -FDG PET-CT) has emerged as a valuable diagnostic tool in suspected LVV, combining the strengths of functional and structural imaging. This study aimed to compare the accuracy of FDG PET-CT and contrast-enhanced CT (CECT) in the evaluation of patients with LVV.

METHODS:

A retrospective database review for LVV patients undergoing CECT and PET-CT between 2011 to 2016 yielded demographics, scan interval and vasculitis type. Qualitative and quantitative PET-CT analyses included aorta:liver FDG uptake, bespoke FDG uptake distribution scores and vascular maximum standardised uptake values (SUV_{max}). Quantitative CECT data were assessed for wall thickness and mural-lumen

ratio. Receiver operating characteristics (ROC) curves were constructed to evaluate comparative diagnostic accuracy and a correlational analysis was conducted between SUV_{max} and wall thickness.

RESULTS:

36 adults (17 LVV, 19 controls) with a mean age (range) 63 (38-89) years, of which 17 (47%) were males were included. Time interval between CT and PET was mean [standard deviation (SD)] 1.9 (1.2) months. Both SUV_{max} and wall thickness demonstrated a significant difference between LVV and controls, with a mean difference [95% confidence interval (CI)] for SUV_{max} 1.6 (1.1, 2.0) and wall thickness 1.25 (0.68, 1.83) mm, respectively. These two parameters were significantly correlated ($p < 0.0001$, $R = 0.62$). The area under the curve (AUC) (95% CI) for SUV_{max} was 0.95 (0.88-1.00), and for mural thickening was 0.83 (0.66-0.99).

CONCLUSION:

FDG PET-CT demonstrated excellent accuracy whilst CECT mural thickening showed good accuracy in the diagnosis of LVV. Both parameters showed a highly significant correlation. In hospitals without access to FDG PET-CT or in patients unsuitable for PET-CT (e.g. uncontrolled diabetes) CECT offers a viable alternative for the assessment of LVV. Advances in knowledge: FDG PET-CT is a highly accurate test for the diagnosis of LVV. Aorta:liver SUV_{max} ratio is the most specific parameter for LVV. In hospitals without PET-CT or in unsuitable patients e.g. diabetics, CECT is a viable alternative.

Source: Br J Radiol. 2018 Jul 5:20180247. doi: 10.1259/bjr.20180247. [Epub ahead of print]

20. FDG-PET/CT(A) imaging in large vessel vasculitis and polymyalgia rheumatica: joint procedural recommendation of the EANM, SNMMI, and the PET Interest Group (PIG), and endorsed by the ASNC.

Slart RHJA; Writing group; Reviewer group; Members of EANM Cardiovascular; Members of EANM Infection & Inflammation; Members of Committees, SNMMI Cardiovascular; Members of Council, PET Interest Group; Members of ASNC; EANM Committee Coordinator.
Collaborators (93)

Abstract

Large vessel vasculitis (LVV) is defined as a disease mainly affecting the large arteries, with two major variants, Takayasu arteritis (TA) and giant cell arteritis (GCA). GCA often coexists with polymyalgia rheumatica (PMR) in the same patient, since both belong to the same disease spectrum. FDG-PET/CT is a functional imaging technique which is an established tool in oncology, and has also demonstrated a role in the field of inflammatory diseases. Functional FDG-PET combined with anatomical CT angiography, FDG-PET/CT(A), may be of synergistic value for optimal diagnosis, monitoring of disease activity, and evaluating damage progression in LVV. There are currently no guidelines regarding PET imaging acquisition for LVV and PMR, even though standardization is of the utmost importance in order to facilitate clinical studies and for daily clinical practice. This work constitutes a joint procedural recommendation on FDG-PET/CT(A) imaging in large vessel vasculitis (LVV) and PMR from the Cardiovascular and Inflammation & Infection Committees of the European Association of Nuclear Medicine (EANM), the Cardiovascular Council of the Society of Nuclear Medicine and Molecular Imaging (SNMMI), and the PET Interest Group (PIG), and endorsed by the American Society of Nuclear Cardiology (ASNC). The aim of this joint paper is to provide recommendations and statements, based on the available evidence in the literature and consensus of experts in the field, for patient preparation, and FDG-

PET/CT(A) acquisition and interpretation for the diagnosis and follow-up of patients with suspected or diagnosed LVV and/or PMR. This position paper aims to set an internationally accepted standard for FDG-PET/CT(A) imaging and reporting of LVV and PMR.

Source: Eur J Nucl Med Mol Imaging. 2018 Jul;45(7):1250-1269. doi: 10.1007/s00259-018-3973-8. Epub 2018 Apr 11.

21. Recent Advances in Giant Cell Arteritis

Guevara M, Kollipara CS

Abstract

PURPOSE OF REVIEW:

Giant cell arteritis (GCA) is the most common systemic vasculitis. GCA is categorized as a granulomatous vasculitis of large and medium size vessels. Majority of the symptoms and signs of GCA result from involvement of the aorta and its branches intra- and extracranial. Temporal artery biopsy continues to be the cardinal diagnostic procedure despite new imaging modalities for diagnosing GCA with cranial involvement. Great advances in awareness have led to improvement in preventing irreversible vision loss due to early diagnosis.

RECENT FINDINGS:

The cause of GCA has not been elucidated but major progress has been made in the knowledge of its pathogenesis leading to new therapeutic targets, particularly inhibition of interleukin 6. IL 6 plays a key role in the regulation of TH17/Tregs imbalance in GCA and appears to correlate with clinical disease activity in GCA. All of this has led to the first FDA (food and drug administration) approved treatment for GCA, Tocilizumab. Abatacept and Ustekinumab are promising targets for therapy in LVV but still need further research. This paper is a review of the recent progress in the understanding of GCA pathogenesis, diagnosis, treatment, and prognosis.

Source: Curr Rheumatol Rep. 2018 Apr 2;20(5):25. doi: 10.1007/s11926-018-0737-1.

22. Prognosis and monitoring of giant cell arteritis and associated complications

Kermani TA, Warrington KJ.

Abstract

Giant cell arteritis (GCA) is the most common systemic vasculitis in people over the age of 50 years. Prospective imaging studies in GCA highlight the systemic nature of this vasculitis. Areas covered: This review summarizes literature using PubMed on complications of GCA and its treatment. Emphasis was placed on articles published within the past 5 years. Disease associated complications including vision loss from arteritic anterior ischemic optic neuropathy, large-artery stenoses and ischemia, and, aortic aneurysms and dissections. Glucocorticoids are effective but have serious adverse effects. Furthermore, relapses are frequent and treatment- or disease-associated damage may accrue. Tocilizumab is the first treatment that showed efficacy in a large randomized prospective trial as a glucocorticoid sparing agent for GCA. Patients with GCA are also at increased risk for multiple cardiovascular diseases and venous thromboembolism. Monitoring for large-vessel involvement, particularly late manifestations like aortic aneurysms is important. Expert commentary: Advances in, and the incorporation of, imaging in GCA have led to better recognition and diagnosis of patients with large-vessel involvement. Prompt treatment with

glucocorticoids is essential in preventing the occurrence or progression of vision loss. Therapeutics that allow sustained remission and reduce vessel damage in patients with GCA will play a crucial role.

Source: Expert Rev Clin Immunol. 2018 May;14(5):379-388. doi: 10.1080/1744666X.2018.1467758. Epub 2018 Apr 26.

23. Negative temporal artery biopsy: predictive factors for giant cell arteritis diagnosis and alternate diagnoses of patients without arteritis.

Bornstein G, Barshack I, Koren-Morag N, Ben-Zvi I, Furie N⁴, Grossman C.

Abstract

To investigate whether among patients with a negative temporal artery biopsy (TAB) there are clinical features that may differentiate between patients with an eventual diagnosis of giant cell arteritis (GCA) and those without arteritis, and to assess the eventual diagnoses of patients without arteritis. Retrospective analysis of patients with a negative TAB performed between 1/1/2000 and 31/12/2015. Information collected included baseline clinical and laboratory data. Patients' final diagnoses were obtained from medical records. Patients eventually diagnosed with GCA were compared with those without arteritis, and predictive features for GCA diagnosis were assessed. A total of 154 patients with a negative TAB were included in the study. Among them, 31 (20%) were eventually diagnosed with GCA. The leading alternative diagnoses of patients without arteritis were self-limited disease (23%), isolated polymyalgia rheumatica (PMR) (18%), and neurological conditions (17%). In the multivariate analysis, predictors for diagnosis of GCA among patients with a negative TAB included PMR (OR = 2.86, 95% CI 1.06-7.69), platelet count (OR = 1.28, 95% CI 1.07-1.53), and ACR score > 2 (OR = 13.4, 95% CI 4.27-42.03). Among patients with a negative TAB, the best predictors for diagnosis of GCA are fulfillment of the ACR criteria, a clinical diagnosis of PMR, and high platelet levels. These features may aid in the diagnostic work-up of patients with a negative TAB.

Source: Clin Rheumatol. 2018 Mar 17. doi: 10.1007/s10067-018-4068-4. [Epub ahead of print]

24. Risk of fracture among patients with polymyalgia rheumatica and giant cell arteritis: a population-based study

Paskins Z, Whittle R, Sultan AA, Muller S, Blagojevic-Bucknall M, Helliwell T, Hider S, Roddy E, Mallen C.

Abstract

BACKGROUND:

Glucocorticoids are associated with increased fracture risk and are the mainstay of treatment in polymyalgia rheumatica (PMR) and giant cell arteritis (GCA). However, fracture risk in these conditions has not been previously quantified. The aim of this study was to quantify the risk of fracture among patients with PMR and GCA.

METHODS:

A retrospective cohort study was conducted using primary care records from the UK-based Clinical Practice Research Datalink. Individuals aged 40 years and over, with incident diagnoses of PMR or GCA were separately identified from 1990-2004 and followed up until 2015. For each exposed individual, four age-, sex- and practice-matched controls were randomly selected. Incidence rates of fracture per 10,000 person-years were calculated for each disease group and hazard rates were compared to the unexposed using Cox regression models.

RESULTS:

Overall, 12,136 and 2673 cases of PMR and GCA, respectively, were identified. The incidence rate of fracture was 148.05 (95% CI 141.16-155.28) in PMR and 147.15 (132.91-162.91) in GCA per 10,000 person-years. Risk of fracture was increased by 63% in PMR (adjusted hazard ratio 1.63, 95% CI 1.54-1.73) and 67% in GCA (1.67, 1.49-1.88) compared to the control populations. Fewer than 13% of glucocorticoid-treated cases were prescribed bisphosphonates.

CONCLUSIONS:

This study reports, for the first time, a similar increase in fracture risk for patients with PMR and GCA. More needs to be done to improve adherence to guidelines to co-prescribe bisphosphonates. Further research needs to identify whether lower glucocorticoid starting doses and/or aggressive dose reduction reduces fracture risk.

Source: BMC Med. 2018 Jan 10;16(1):4. doi: 10.1186/s12916-017-0987-1.

25. Giant cell arteritis and polymyalgia rheumatica: current challenges and opportunities

Dejaco C, Brouwer E, Mason JC, Buttgerit F, Matteson EL, Dasgupta B.

Abstract

The fields of giant cell arteritis (GCA) and polymyalgia rheumatica (PMR) have advanced rapidly, resulting in a new understanding of these diseases. Fast-track strategies and improved awareness programs that prevent irreversible sight loss through early diagnosis and treatment are a notable advance. Ultrasonography and other imaging techniques have been introduced into routine clinical practice and there have been promising reports on the efficacy of biologic agents, particularly IL-6 antagonists such as tocilizumab, in treating these conditions. Along with these developments, which should improve outcomes in patients with GCA and PMR, new questions and unmet needs have emerged; future research should address which pathogenetic mechanisms contribute to the different phases and clinical phenotypes of GCA, what role imaging has in the early diagnosis and monitoring of GCA and PMR, and in which patients and phases of these diseases novel biologic drugs should be used. This article discusses the implications of recent developments in our understanding of GCA and PMR, as well as the unmet needs concerning epidemiology, pathogenesis, imaging and treatment of these diseases.

Source: Nat Rev Rheumatol. 2017 Oct;13(10):578-592. doi: 10.1038/nrrheum.2017.142. Epub 2017 Sep 14.

26. The spectrum of giant cell arteritis and polymyalgia rheumatica: revisiting the concept of the disease.

Dejaco C, Duftner C, Buttgerit F, Matteson EL, Dasgupta B

Abstract

GCA and PMR are conditions of older persons that frequently overlap. The traditional concept of GCA has focused on cranial symptoms such as headache and visual disturbance, but extra-cranial manifestations such as constitutional symptoms, polymyalgia and limb claudication have also long been recognized. These symptoms may coincide with cranial GCA, occur as an independent clinical subset [large-vessel (LV) GCA] or overlap with PMR. Imaging studies have demonstrated that up to one-third of patients with PMR have subclinical LV inflammation at disease outset. The implication of this finding for PMR management is unclear. Pathophysiological studies have emphasized the pivotal role

of dendritic cells (DCs) and T cells in the pathogenesis of GCA, and the activation of certain pattern recognition receptors on DCs may determine the clinical subset of GCA. In patients with only PMR clinically, it is conceivable that transmural arterial inflammation has either not yet started or is prevented by unexplored regulatory pathways. This concept is supported by vasculitis of peri-adventitial small-vessels and activated DCs in the adventitia of temporal arteries, in the absence of media-infiltrating T cells. This review examines the clinical and pathophysiological spectrum of GCA and its subsets with PMR, the role of newer imaging techniques for GCA diagnosis and the management of these diseases.

Source: *Rheumatology* (Oxford). 2017 Apr 1;56(4):506-515. doi: 10.1093/rheumatology/kew273.

27. What Is the Current Evidence for Disease Subsets in Giant Cell Arteritis?

van der Geest KSM, Sandovici M, van Sleen Y, Sanders JS, Bos NA, Abdulhad WH, Stegeman CA, Heeringa P, Rutgers A, Kallenberg CGM, Boots AMH, Brouwer E.

Abstract

Giant cell arteritis (GCA) is an autoimmune vasculitis affecting large and medium-sized arteries. Ample evidence indicates that GCA is a heterogeneous disease in terms of symptoms, immune pathology, and response to treatment. In the current review, we discuss the evidence for disease subsets in GCA. We describe clinical and immunologic characteristics that may impact the risk of cranial ischemic symptoms, relapse rates, and long-term glucocorticoid requirements in patients with GCA. In addition, we discuss both proven and putative immunologic targets for therapy in patients with GCA who have an unfavorable prognosis. Finally, we provide recommendations for further research on disease subsets in GCA.

Source: *Arthritis Rheumatol*. 2018 Apr 12. doi: 10.1002/art.40520. [Epub ahead of print]

28. CT analysis of the aorta in giant-cell arteritis: a case-control study

Berthod PE, Aho-Glélé S, Ornetti P, Chevallier O, Devilliers H, Ricolfi F, Bonnotte B, Loffroy R, Samson M.

Abstract

OBJECTIVES:

Giant cell arteritis (GCA) is a large-vessel vasculitis whose diagnosis is confirmed by temporal artery biopsy. However, involvement of large vessels, especially the aorta, can be shown by imaging, which plays an increasing role in GCA diagnosis. The threshold above which aortic wall thickening, as measured by computed tomography (CT), is considered pathological is controversial, with values ranging from 2 to 3 mm. This study assessed aortic morphology by CT scan and its diagnostic value in GCA.

METHODS:

Altogether, 174 patients were included (64 with GCA, 43 with polymyalgia rheumatica and 67 controls). All patients had a CT scan at diagnosis or at inclusion for controls. Aortic wall thickness, aortic diameter and scores for atheroma were measured. Assessor was blinded to each patient's group.

RESULTS:

Aortic diameters and atheroma scores were similar between groups. Aortic wall thickness was greater in the GCA group, even after the exclusion of GCA patients with aortic wall thickness ≥ 3 mm. The receiver operating characteristic (ROC) curve showed that a wall thickness of 2.2 mm was the optimal threshold to diagnose GCA (sensitivity, 67%; specificity, 98%).

CONCLUSIONS:

Measuring aortic wall thickness by CT scan is effective to diagnose GCA. The optimal threshold to regard aortic wall thickening as pathological was ≥ 2.2 mm.

Source: Eur Radiol. 2018 Mar 29. doi: 10.1007/s00330-018-5311-8. [Epub ahead of print]

29. Giant-Cell Arteritis: Do We Treat Patients with Large-Vessel Involvement Differently?

de Boysson H, Liozon E, Lambert M, Dumont A, Boutemy J, Maigné G, Martin Silva N, Ly KH, Manrique A, Bienvenu B, Aouba A.

Abstract**PURPOSE:**

We aimed to describe the initial treatment that was used in a common hospital-based practice in patients with giant-cell arteritis with and without large-vessel involvement at diagnosis as well as the outcomes in both groups.

METHODS:

This retrospective multi-center cohort included patients with giant-cell arteritis diagnosed between 2005 and 2015, all of whom had fluorodeoxyglucose (FDG) positron emission tomography combined with computed tomography (FDG-PET/CT) performed at giant-cell arteritis diagnosis and were followed up for ≥ 12 months. We compared the features, treatment, and outcomes of patients with large-vessel involvement demonstrated on FDG-PET/CT with those of patients with a negative PET/CT.

RESULTS:

Eighty patients (50 women, median age: 71 [53-87] years) were included, 40 of whom had large-vessel involvement demonstrated on FDG-PET/CT and 40 who did not. After a median 56-month follow-up time, 42 (53%) patients had discontinued glucocorticoid (GC) treatment. Patients with and without large-vessel involvement were indistinguishable in the initial median dose of prednisone (0.74 mg/kg vs 0.75 mg/kg, $P = .56$), overall GC duration ($P = .77$), GC discontinuation rate ($P = .65$), relapse rate ($P = .50$), frequency of GC-dependent disease requiring GC-sparing treatments ($P = .62$), and fatality rate ($P = .06$).

CONCLUSION:

In the setting of tertiary hospital recruitment, large-vessel involvement at giant-cell arteritis diagnosis using a PET/CT study had no influence on the choice of initial GC dose and had no impact on outcomes. Prospective studies are required to confirm these findings.

Source: Am J Med. 2017 Aug;130(8):992-995. doi: 10.1016/j.amjmed.2017.03.054. Epub 2017 Apr 29.

30. Serious adverse effects associated with glucocorticoid therapy in patients with giant cell arteritis(GCA): A nested case-control analysis.

Wilson JC, Sarsour K, Collinson N, Tuckwell K, Musselman D, Klearman M, Napalkov P, Jick SS, Stone JH, Meier CR.

Abstract

OBJECTIVE:

Giant cell arteritis (GCA) is an inflammatory vasculitis preferentially affecting large and medium-sized arteries. High-dose oral glucocorticoids (GCs) are the mainstay of GCA therapy. Using data from the UK Clinical Practice Research Datalink (CPRD), we examined the risk of oral GC-related serious adverse events (SAEs) in a UK population of patients with giant cell arteritis (GCA).

METHODS:

We conducted a series of nested case-control analyses in GCA patients to examine the effect of increasing dose of prednisolone on the risk of developing diabetes, glaucoma, osteoporosis, fractures, serious infection requiring hospitalization, and death. We used conditional logistic regression to calculate the unadjusted and multivariate odds ratios (ORs) with 95% CIs for the associations between prednisolone use and the risks of all outcomes of interest. We stratified the analyses by increasing cumulative prednisolone use and average daily dose.

RESULTS:

In the multivariate analyses, we observed a trend of increasing risk of diabetes and osteoporosis with increasing cumulative dose of oral prednisolone ($p^{\text{trend}} < 0.05$). GCA patients in the highest daily dose category (30mg/d) had an increased risk of diabetes (adjusted OR, 95% CI) (4.7, 2.8-7.8), osteoporosis (1.9, 1.2-2.9), fractures (2.6, 1.6-4.3), glaucoma (3.5, 2.0-6.1), serious infection (3.3, 2.2-5.2), and death (2.1, 1.3-3.5) compared to those with lower average daily prednisolone doses (5mg/d).

CONCLUSION:

Compared to lower average daily prednisolone doses, high average daily doses were associated with an increased risk of serious adverse effects.

Source: Semin Arthritis Rheum. 2017 Jun;46(6):819-827.

doi: 10.1016/j.semarthrit.2016.11.006. Epub 2016 Nov 28.

31. Association between glucocorticoid therapy and incidence of diabetes mellitus in polymyalgia rheumatica and giant cell arteritis: a systematic review and meta-analysis.

Lai LYH, Harris E, West RM, Mackie SL.

Abstract

BACKGROUND:

Polymyalgia rheumatica (PMR) and giant cell arteritis (GCA) are almost always treated with glucocorticoids (GCs), but long-term GC use is associated with diabetes mellitus (DM). The absolute incidence of this complication in this patient group remains unclear.

OBJECTIVE:

To quantify the absolute risk of GC-induced DM in PMR and GCA from published literature.

METHODS:

We identified literature from inception to February 2017 reporting diabetes following exposure to oral GC in patients with PMR and/or GCA without pre-existing diabetes. A random-effects meta-analysis was performed to summarize the findings.

RESULTS:

25 eligible publications were identified. In studies of patients with GCA, mean cumulative GC dose was almost 1.5 times higher than in studies of PMR (8.2 g vs 5.6 g), with slightly longer treatment duration and longer duration of follow-up (6.4 years vs 4.4 years). The incidence proportion (cumulative incidence) of patients who developed new-onset DM was 6% (95% CI 3% to 9%) for PMR and 13% (95% CI 9% to 17%) for GCA. Based on UK data on incidence rate of DM in the general population, the expected background incidence rate of DM over 4.4 years in patients with PMR and 6.4 years in patients with GCA (follow-up duration) would be 4.8% and 7.0%, respectively. Heterogeneity between studies was high ($I^2=79.1\%$), as there were differences in study designs, patient population, geographical locations and treatment. Little information on predictors of DM was found.

CONCLUSION:

Our meta-analysis produced plausible estimates of DM incidence in patients with PMR and GCA, but there is insufficient published data to allow precise quantification of DM risk.

Source: RMD Open. 2018 Feb 28;4(1):e000521. doi: 10.1136/rmdopen-2017-000521. eCollection 2018.

32. Five-year prospective multi-center cohort study of patients with giant cell arteritis in Greece

Christina Tsalapaki, Eleni Nikitopoulou, Kyriaki A. Boki, Dimitrios Boumpas, Petros P. Sfikakis, Georgios Vosvotekas, Paraskevi V. Voulgari, Dimitrios Vassilopoulos

Abstract

Giant cell arteritis (GCA) is the most common systemic vasculitis in the aged population associated with significant morbidity due to the long term administration of corticosteroids and the presence of various comorbidities. Data regarding its current treatment modalities, comorbidities, morbidity and mortality in Greece are limited. In this multi-center, prospective study that begun at the end of 2015 patients with newly diagnosed GCA according to the modified 1990 ACR criteria, as well as individuals with established or relapsing disease have been included. During the 1st phase of the study that is still ongoing, data are being collected concerning demographic and clinical characteristics of the patients, treatment at the onset of the disease and at relapses, relapses, adverse events of therapy, comorbidities, hospitalizations and deaths. During the 2nd and 3rd phase of the study patients will be reevaluated 2 and 5 years after their 1st evaluation. The study is expected to provide valuable data regarding the current clinical characteristics, co-morbidities, therapeutic regimens used, relapse rate, morbidity and mortality of patients with GCA.

Source: *Mediterr J Rheumatol* 2018;29(2):103-5 <https://doi.org/10.31138/mjr.29.2.103>

33. Infections and vasculitis.

Thomas K, Vassilopoulos D.

Abstract

PURPOSE OF REVIEW:

To review recent evidence for infection rates in patients with systemic vasculitides, the role of specific infectious agents in the pathogenesis of vasculitis and recent breakthroughs in the treatment of virus-associated vasculitides.

RECENT FINDINGS:

In well-designed recent studies, infections were found to be common during the first 6-12 months in patients with anti-neutrophil cytoplasmic antibodies (ANCA)-associated vasculitides (AAV) and giant cell arteritis (GCA) and to contribute significantly to increased mortality during this period. New therapeutic schemes with lower cyclophosphamide doses and shorter corticosteroid courses were associated with decreased infectious rates in elderly patients with AAV whereas a prednisone dose greater than 10mg/day at the end of the first year were associated with increased infectious-related mortality in patients with GCA. Recently, a potential role for varicella zoster virus in GCA pathogenesis has been proposed but more data are needed in order to establish a causal relationship. Finally, preliminary data show excellent short-term efficacy and safety of the new, interferon-free, oral antiviral agents in the treatment of hepatitis C virus-associated cryoglobulinemic vasculitis.

SUMMARY:

Infections continue to be one of the main causes of mortality in patients with systemic vasculitides, emphasizing the need for safer immunosuppressive therapies and appropriate prophylaxis.

Source: Curr Opin Rheumatol. 2017 Jan;29(1):17-23.

34. What is the impact of giant cell arteritis on patients' lives? A UK qualitative study.

Liddle J, Bartlam R, Mallen CD, Mackie SL, Prior JA, Helliwell T, Richardson JC.

Abstract**OBJECTIVES:**

Clinical management of giant cell arteritis (GCA) involves balancing the risks and burdens arising from the disease with those arising from treatment, but there is little research on the nature of those burdens. We aimed to explore the impact of giant cell arteritis (GCA) and its treatment on patients' lives.

METHODS:

UK patients with GCA participated in semi-structured telephone interviews. Inductive thematic analysis was employed.

RESULTS:

24 participants were recruited (age: 65-92 years, time since diagnosis: 2 months to >6 years). The overarching themes from analysis were: ongoing symptoms of the disease and its treatment; and 'life-changing' impacts. The overall impact of GCA on patients' lives arose from a changing combination of symptoms, side effects, adaptations to everyday life and impacts on sense of normality. Important factors contributing to loss of normality were glucocorticoid-related treatment burdens and fear about possible future loss of vision.

CONCLUSIONS:

The impact of GCA in patients' everyday lives can be substantial, multifaceted and ongoing despite apparent control of disease activity. The findings of this study will help doctors better understand patient priorities, legitimise patients' experiences of GCA and work with patients to set realistic treatment goals and plan adaptations to their everyday lives.

Source: BMJ Open. 2017 Aug 23;7(8):e017073. doi: 10.1136/bmjopen-2017-017073.

35. The Association Between Giant Cell Arteritis and Ischemic Heart Disease: A Population Based Cross-Sectional Study.

Dagan A, Mahroum N, Segal G, Tiosano S, Watad A, Comaneshter D, Cohen AD, Amital H

Abstract

BACKGROUND:

Patients with giant cell arteritis (GCA) suffer from inflammatory diseases often treated by large amounts of corticosteroids. Whether this inflammatory burden also carries an increased risk for cardiovascular morbidity, and especially ischemic heart disease, is not clearly established.

OBJECTIVES:

To clarify the linkage between GCA and ischemic heart disease.

METHODS:

In a cross-sectional study, we assessed the association between GCA and ischemic heart disease, adjusting for cardiovascular risk factors, among GCA patients and matched controls using the database of the largest healthcare provider in Israel.

RESULTS:

The study group was comprised of 5659 GCA patients and 28,261 age and gender matched controls. The proportion of ischemic heart disease was higher in the GCA group (27.5% vs. 12.5% among controls, odds ratio 2.65). Diabetes mellitus, hypertension, hyperlipidemia and smoking were also found to have higher concurrency in GCA. After stratifying for those cardiovascular co-morbidities using logistic regression, GCA remained independently associated with ischemic heart disease with an odds ratio of 1.247 (1.146-1.357 P < 0.001).

CONCLUSIONS:

GCA is associated with both cardiovascular risk factors and ischemic heart disease. Healthcare professionals should not overlook this aspect of the disease when managing GCA patients.

Source: Isr Med Assoc J. 2017 Jul;19(7):411-414.

36. Use of imaging techniques in large vessel vasculitis and related conditions.

Versari A, Pipitone N, Casali M, Jamar F, Pazzola G.

Abstract

Giant cell arteritis (GCA) and Takayasu's arteritis (TA) are large vessel vasculitis (LVV) primarily affecting the aorta and its major branches, mainly differentiated by the onset age (>50 years GCA and <40 years TA). In addition, temporal artery involvement and polymyalgia rheumatica are typical features of GCA, but not TA. Imaging techniques are required to secure the diagnosis of large-vessel vasculitides, and to monitor the disease course. Both morphological and metabolic imaging are involved. Morphological imaging is represented mainly by computerized tomography (CT), CT angiography, magnetic resonance (MR), MR angiography, color-Doppler sonography (CDS) and high-resolution CDS. Metabolic aspects of inflammatory process in LVV can be well studied by positron emission tomography/computed tomography with [18F]deoxyglucose ([18F]FDG PET/CT). It has an important increasing role in diagnosis, extent assessment and disease

activity and therapy response evaluation. In the near future the concomitant development of increasingly powerful PET/CT scanners, of new radiopharmaceuticals more specific for inflammation, and of new PET/MRI hybrid scanners probably will lead to a further new step forward in the diagnosis and clinical management of LVV.

Source: Q J Nucl Med Mol Imaging. 2018 Mar;62(1):34-39. doi: 10.23736/S1824-4785.17.03044-8. Epub 2017 Nov 22.

37. The proposed role of ultrasound in the management of giant cell arteritis in routine clinical practice.

Monti S, Floris A, Ponte CB, Schmidt WA, Diamantopoulos AP, Pereira C, Vaggers S, Luqmani RA.

Abstract

OBJECTIVE:

To develop and explore a protocol for using colour duplex sonography (CDS) in the routine care of GCA.

METHODS:

We tested CDS of temporal arteries and axillary arteries (AXs) on consecutive patients with suspected or established GCA, between July 2014 and September 2016.

RESULTS:

We assessed 293 patients [age 72 (10), female/male 196/97], of whom 118 had clinically confirmed GCA. Seventy-three percent of patients had already received high-dose glucocorticoids (GCs) for 17 (33) days. Among new referrals with <7 days of GC treatment (n = 55), the sensitivity of CDS was 63.3% (95% CI: 44%, 80%), specificity 100% (95% CI: 83%, 100%), positive predictive value 100% and negative predictive value 64.5% (95% CI: 53%, 74%). Sensitivity rose to 81.8% in patients with jaw claudication and high inflammatory markers. During the observation period, the rate of temporal artery biopsies decreased from 72 (42%) to 36 (25%) (P = 0.002). CDS was positive in 21% of 89 follow-up scans in asymptomatic individuals, compared with 37% in patients experiencing clinical flares. Over time, the number of halos reduced; only new or flaring patients showed a halo in four or more sites. The diameter of axillary halos reduced from referral [1.6 (0.4) mm] to follow-up [1.4 (0.2) mm, P = 0.01] or flares [1.4 (0.2) mm, P = 0.02].

CONCLUSION:

CDS provides high positive predictive value for diagnosing GCA and allows for a significant reduction in temporal artery biopsies. We explored the role of CDS in detecting flares and demonstrated a relationship to the extent of the distribution of halos, but not to their size.

Source: Rheumatology (Oxford). 2018 Jan 1;57(1):112-119. doi: 10.1093/rheumatology/kex341.

38. Usefulness of PET in recognizing and managing vasculitides.

Pipitone NAM, Versari A, Salvarani C.

Abstract

PURPOSE OF REVIEW:

The aim of this article was to review the recent contributions to the scoring methods of PET in vasculitis as well as to its role in the diagnostic work-up.

RECENT FINDINGS:

Both visual and semiquantitative scoring methods can be used to interpret PET scans. PET has been shown to be both sensitive and specific in the diagnosis of large-vessel vasculitis. In addition, it also has a role in predicting vascular complications.

SUMMARY:

There is a need to better standardize the scoring methods used to interpret PET scans. In clinical practice, PET is useful to diagnose untreated individuals with suspected large-vessel vasculitis and contributes to identify patients at risk for vascular complications.

Source: *Curr Opin Rheumatol*. 2018 Jan;30(1):24-29.

doi: 10.1097/BOR.0000000000000459.

39. The use of ultrasound to assess giant cell arteritis: review of the current evidence and practical guide for the rheumatologist.

Monti S, Floris A, Ponte C, Schmidt WA, Diamantopoulos AP, Pereira C, Piper J, Luqmani R.

Abstract

Colour duplex sonography (CDS) of temporal arteries and large vessels is an emerging diagnostic tool for GCA. CDS can detect wall oedema, known as a halo, throughout the length of the vessel and shows higher sensitivity compared with biopsy. Specificity reaches 100% in case of bilateral halos. A positive compression sign has been demonstrated to be a robust marker with excellent inter-observer agreement. The assessment of other large vessels, particularly the axillary arteries, is recognized to further increase the sensitivity and to reliably represent extra-cranial involvement in other areas. Nevertheless, CDS use is still not widespread in routine clinical practice and requires skilled sonographers. Moreover, its role in the follow-up of patients still needs to be defined. The aim of this review is to provide the current evidence and technical parameters to support the rheumatologist in the CDS evaluation of patients with suspected GCA.

Source: *Rheumatology (Oxford)*. 2018 Feb 1;57(2):227-235.

doi: 10.1093/rheumatology/kex173.

40. Recent advances in our understanding of giant cell arteritis pathogenesis.

Samson M, Corbera-Bellalta M, Audia S, Planas-Rigol E, Martin L⁴ Cid MC, Bonnotte B.

Abstract

Giant cell arteritis (GCA) is a granulomatous vasculitis affecting large arteries, especially the aorta and the extracranial branches of the external carotid artery. Its exact pathogenesis is not fully understood but major progress has been made in recent years, leading to new therapeutic targets like inhibition of the interleukin-6 pathway or the modulation of immune checkpoints. The cause of GCA has not been clearly identified but it is thought that GCA occurs on a genetic background and is triggered by unknown environmental factors that could activate and lead to the maturation of dendritic cells localized in the adventitia of normal arteries. These activated dendritic cells then produce chemokines which trigger the recruitment of CD4⁺ T cells, which in turn become activated, proliferate and polarize into Th1 and Th17 cells, which produce IFN- γ and IL-17, respectively. Exposed to IFN- γ , endothelial cells and vascular smooth muscle cells produce chemokines leading to the recruitment of further Th1 cells, CD8⁺ T cells and monocytes. The latter differentiate into macrophages, which,

when persistently exposed to IFN- γ , form giant cells, the histological hallmark of GCA. With the contribution of vascular smooth muscle cells, immune cells then trigger the destruction and remodeling of the arterial wall, thus leading to the formation of a neo-intima resulting in progressive occlusion of the arterial lumen, which is responsible for the ischemic symptoms of GCA. In this paper, we review recent progress in our understanding of GCA pathogenesis in the fields of genetics, epigenetics, infections, immunology and vascular remodeling.

Source: Autoimmun Rev. 2017 Aug;16(8):833-844. doi: 10.1016/j.autrev.2017.05.014. Epub 2017 May 28.

41. **Challenges of diagnosis and management of giant cell arteritis in general practice: a multi-methods study.**

Helliwell T, Muller S, Hider SL, Prior JA, Richardson JC, Mallen CD.

Abstract

BACKGROUND:

In the UK, general practitioners (GPs) are usually the first medical contact for patients with suspected giant cell arteritis(GCA). While rare, it is critical not to miss, as delayed treatment can lead to significant complications including permanent visual loss. To date, little is known about the approach and challenges to diagnosis and management of GCA by GPs.

OBJECTIVE:

To investigate the diagnosis and management of patients with suspected GCA in UK general practice.

DESIGN AND PARTICIPANTS:

A multi-methods approach was taken, comprising a postal survey of 5000 randomly selected UK GPs and semi-structured telephone interviews of 24 GPs from across the UK.

SETTING:

UK general practice.

RESULTS:

1249 questionnaires were returned. 879 responders (70%) indicated that they had diagnosed and managed a patient with GCA. A variety of clinical features were used to identify GCA. 21.9% suggested that they would exclude GCA as a diagnosis if headache was absent and around one-third do not routinely initiate glucocorticoid treatment prior to referral. Significant regional variations in referral pathways were reported. Thematic analysis of interview transcripts highlighted fears relating to a missed diagnosis of GCA and the non-specific nature of early GCA presentation. Accessing specialist care was highlighted as challenging by many GPs and that a national standard fast-track pathway is lacking to support this patient group. Additionally, there were significant concerns regarding potential adverse effects relating to long-term treatment with glucocorticoids.

CONCLUSION:

GPs appear to over-rely on headache to identify GCA and marked geographical differences in management, with conflicting referral pathways and difficulties in accessing appropriate services exist in the UK. A national standard for fast-tracking patients with suspected GCA to relevant specialists would be beneficial to improve care and outcomes for patients with GCA.

Source: BMJ Open. 2018 Feb 3;8(2):e019320. doi: 10.1136/bmjopen-2017-019320.

42. Evaluation of damage in giant cell arteritis.

Kermani TA, Sreih AG, Cuthbertson D, Carette S, Hoffman GS, Khalidi NA, Koenig CL, Langford CA, McAlear CA, Monach PA, Moreland L, Pagnoux C, Seo P, Warrington KJ, Ytterberg SR, Merkel PA; Vasculitis Clinical Research Consortium.

Abstract

OBJECTIVES:

To evaluate damage and variables associated with damage in GCA.

METHODS:

Patients with GCA enrolled in a prospective, multicentre, longitudinal study were included. Per-protocol assessments were made with the Vasculitis Damage Index and the Large-Vessel Vasculitis Index of Damage.

RESULTS:

The study included 204 patients: 156 women (76%), mean age at diagnosis 71.3 years (s.d. 8.3), mean follow-up of 3.5 years (s.d. 1.9). One or more damage item was present in 54% at baseline and 79% at the last follow-up on the Vasculitis Damage Index, and 60% at baseline and 82% at the last follow-up on the Large-Vessel Vasculitis Index of Damage. The most frequently observed damage items were large-artery complications (29% cohort) and ocular (22%). Among 117 patients with new damage, most new items were ocular (63 patients), cardiac/vascular (48) and musculoskeletal (34). Of these, treatment-associated items were frequently observed, including cataracts (46 patients), osteoporosis (22) and weight gain (22). Disease-associated new damage included ischaemic optic neuropathy (3 patients), limb claudication (13), arterial occlusions (10) and damage requiring vascular intervention (10). In univariate analysis, the risk of damage increased 22% for every additional year of disease duration [odds ratio (OR) 1.22 (95% CI 1.04, 1.45)]. In 94 patients enrolled within ≤ 90 days of diagnosis of GCA, the risk of new damage at the last follow-up decreased 30% for each additional relapse [OR 0.70 (95% CI 0.51, 0.97)].

CONCLUSIONS:

Large-artery complications and ocular manifestations are the most commonly occurring items of damage in GCA. Most new damage is associated with treatment. These findings emphasize the cumulative burden of disease in GCA.

Source: Rheumatology (Oxford). 2018 Feb 1;57(2):322-328.

doi: 10.1093/rheumatology/kex397.

43. Imaging in diagnosis, outcome prediction and monitoring of large vessel vasculitis: a systematic literature review and meta-analysis informing the EULAR recommendations.

Duftner C, DeJaco C, Sepriano A, Falzon L, Schmidt WA, Ramiro S.

Abstract

OBJECTIVES:

To perform a systematic literature review on imaging techniques for diagnosis, outcome prediction and disease monitoring in large vessel vasculitis (LVV) informing the European League Against Rheumatism recommendations for imaging in LVV.

METHODS:

Systematic literature review (until 10 March 2017) of diagnostic and prognostic studies enrolling >20 patients and investigating ultrasound, MRI, CT or positron emission tomography (PET) in patients with suspected and/or established primary LVV. Meta-analyses were conducted, whenever possible, obtaining pooled estimates for sensitivity and specificity by fitting random effects models.

RESULTS:

Forty-three studies were included (39 on giant cell arteritis (GCA), 4 on Takayasu arteritis (TAK)). Ultrasound ('halo' sign) at temporal arteries (8 studies, 605 patients) and MRI of cranial arteries (6 studies, 509 patients) yielded pooled sensitivities of 77% (95% CI 62% to 87%) and 73% (95% CI 57% to 85%), respectively, compared with a clinical diagnosis of GCA. Corresponding specificities were 96% (95% CI 85% to 99%) and 88% (95% CI 81% to 92%). Two studies (93 patients) investigating PET for GCA diagnosis reported sensitivities of 67%-77% and specificities of 66%-100% as compared with clinical diagnosis or temporal artery biopsy. In TAK, one study each evaluated the role of magnetic resonance angiography and CT angiography for diagnostic purposes revealing both a sensitivity and specificity of 100%. Studies on outcome prediction and monitoring disease activity/damage were limited and mainly descriptive.

CONCLUSIONS:

Ultrasound and MRI provide a high diagnostic value for cranial GCA. More data on the role of imaging for diagnosis of extracranial large vessel GCA and TAK, as well as for outcome prediction and monitoring in LVV are warranted.

Source: RMD Open. 2018 Feb 2;4(1):e000612. doi: 10.1136/rmdopen-2017-000612. eCollection 2018.

44. Defining conditions where long-term glucocorticoid treatment has an acceptably low level of harm to facilitate implementation of existing recommendations: viewpoints from an EULAR task force.

Strehl C, Bijlsma JW, de Wit M, Boers M, Caeyers N, Cutolo M, Dasgupta B, Dixon WG, Geenen R, Huizinga TW, Kent A, de Thurah AL, Listing J, Mariette X, Ray DW, Scherer HU, Seror R, Spies CM, Tarp S, Wiek D, Winthrop KL, Buttgerit F.

Abstract

There is convincing evidence for the known and unambiguously accepted beneficial effects of glucocorticoids at low dosages. However, the implementation of existing recommendations and guidelines on the management of glucocorticoid therapy in rheumatic diseases is lagging behind. As a first step to improve implementation, we aimed at defining conditions under which long-term glucocorticoid therapy may have an acceptably low level of harm. A multidisciplinary European League Against Rheumatism task force group of experts including patients with rheumatic diseases was assembled. After a systematic literature search, breakout groups critically reviewed the evidence on the four most worrisome adverse effects of glucocorticoid therapy (osteoporosis, hyperglycaemia/diabetes mellitus, cardiovascular diseases and infections) and presented their results to the other group members following a structured questionnaire for final discussion and consensus

finding. Robust evidence on the risk of harm of long-term glucocorticoid therapy was often lacking since relevant study results were often either missing, contradictory or carried a high risk of bias. The group agreed that the risk of harm is low for the majority of patients at long-term dosages of ≤ 5 mg prednisone equivalent per day, whereas at dosages of > 10 mg/day the risk of harm is elevated. At dosages between > 5 and ≤ 10 mg/day, patient-specific characteristics (protective and risk factors) determine the risk of harm. The level of harm of glucocorticoids depends on both dose and patient-specific parameters. General and glucocorticoid-associated risk factors and protective factors such as a healthy lifestyle should be taken into account when evaluating the actual and future risk.

Source: Ann Rheum Dis. 2016 Jun;75(6):952-7. doi: 10.1136/annrheumdis-2015-208916. Epub 2016 Mar 1.

45. EULAR evidence-based and consensus-based recommendations on the management of medium to high-dose glucocorticoid therapy in rheumatic diseases.

Duru N, van der Goes MC, Jacobs JW, Andrews T, Boers M, Buttgereit F, Caeyers N, Cutolo M, Halliday S, Da Silva JA, Kirwan JR, Ray D, Rovensky J, Severijns G, Westhovens R, Bijlsma JW.

Abstract

To develop recommendations for the management of medium to high-dose (ie, > 7.5 mg but ≤ 100 mg prednisone equivalent daily) systemic glucocorticoid (GC) therapy in rheumatic diseases. A multidisciplinary EULAR task force was formed, including rheumatic patients. After discussing the results of a general initial search on risks of GC therapy, each participant contributed 10 propositions on key clinical topics concerning the safe use of medium to high-dose GCs. The final recommendations were selected via a Delphi consensus approach. A systematic literature search of PubMed, EMBASE and Cochrane Library was used to identify evidence concerning each of the propositions. The strength of recommendation was given according to research evidence, clinical expertise and patient preference. The 10 propositions regarded patient education and informing general practitioners, preventive measures for osteoporosis, optimal GC starting dosages, risk-benefit ratio of GC treatment, GC sparing therapy, screening for comorbidity, and monitoring for adverse effects. In general, evidence supporting the recommendations proved to be surprisingly weak. One of the recommendations was rejected, because of conflicting literature data. Nine final recommendations for the management of medium to high-dose systemic GC therapy in rheumatic diseases were selected and evaluated with their strengths of recommendations. Robust evidence was often lacking; a research agenda was created.

Source: Ann Rheum Dis. 2013 Dec;72(12):1905-13. doi: 10.1136/annrheumdis-2013-203249. Epub 2013 Jul 19.

46. Corticosteroid-related adverse events in patients with giant cell arteritis: A claims-based analysis.

Broder MS, Sarsour K, Chang E, Collinson N, Tuckwell K, Napalkov P, Klearman M.

Abstract

OBJECTIVE:

Corticosteroids (CS) are standard treatment for giant cell arteritis (GCA), but concerns persist over toxicities associated with long-term use. In this retrospective study of medical claims data, we estimated risks for adverse events (AEs) in CS-treated GCA patients.

METHODS:

Cox regression analyses with CS use as a time-dependent variable were conducted on data from the 2003 to 2012 Truven Health Analytics MarketScan Database. Patients 50 years of age and older who had ≥ 2 claims of newly diagnosed GCA, ≥ 1 filled oral CS prescription, and no AEs before GCA diagnosis were included. The primary outcome was presence of a new CS-related AE.

RESULTS:

In total, 2497 patients were included. Their mean age was 71.0 years, and 71% were women. Follow-up was 9680 patient-years (PY). CS treatment continued for a mean (SD) of 1.196 (729.2) days; mean (SD) prescribed cumulative CS dose was 6983.3mg (6519.9). The overall AE rate was 0.43 events/PY; the most frequent AEs were cataract and bone disease. For each 1000-mg increase in CS exposure, the hazard ratio (HR) increased by 3% (HR = 1.03; 95% CI: 1.02-1.05; $P < 0.001$). Additionally, statistically significant individual associations between increased CS exposure and AE risk were observed for bone-related AEs ($P < 0.001$), cataract ($P < 0.001$), glaucoma ($P = 0.005$), pneumonia ($P = 0.003$), and diabetes mellitus ($P < 0.001$ in a subset of patients with no previous history of diabetes).

CONCLUSION:

CS exposure significantly increased risk for potentially serious AEs, emphasizing a need for new treatment options for GCA patients.

Source: Semin Arthritis Rheum. 2016 Oct;46(2):246-52.

doi: 10.1016/j.semarthrit.2016.05.009. Epub 2016 Jun 2.

47. Predictors of relapse and treatment outcomes in biopsy-proven giant cell arteritis: a retrospective cohort study.

Labarca C, Koster MJ, Crowson CS, Makol A, Ytterberg SR, Matteson EL, Warrington KJ.

Abstract**OBJECTIVE:**

To evaluate characteristics of relapse, relapse rates, treatment and outcomes among patients with biopsy-proven GCA in a large, single-institution cohort.

METHODS:

We conducted a retrospective review of all patients with biopsy-proven GCA from 1998 to 2013. Demographic, clinical, laboratory and treatment data at presentation and during follow-up were collected. Comparisons by relapse rate were performed using chi-square tests. Prednisone discontinuation by initial oral dose ≤ 40 and >40 mg/day was compared using Cox models.

RESULTS:

The cohort included 286 patients [74% female, mean age at diagnosis 75.0 years (s.d. 7.6), median follow-up 5.1 years). During follow-up, 73 patients did not relapse, 80 patients had one relapse and 133 had two or more relapses. The first relapse occurred during the first year in 50% of patients, by 2 years in 68% and by 5 years in 79%. More patients with established hypertension ($P = 0.007$) and diabetes ($P = 0.039$) at GCA diagnosis were in the high relapse rate group (≥ 0.5 relapses/year) and more females were in the low or high relapse groups than in the no relapse group ($P = 0.034$). Patients receiving an initial oral prednisone dose >40 mg/day were able to reach a dose of <5 mg/day [hazard ratio (HR) 1.46 (95% CI 1.09, 1.96)] and discontinue prednisone [HR 1.56 (95% CI 1.09, 2.23)] sooner than patients receiving ≤ 40 mg/day without an increase in observed glucocorticoid-associated adverse events.

CONCLUSION:

Females and patients with hypertension or diabetes at GCA diagnosis have more relapses during follow-up. Patients treated with an initial oral prednisone dose >40 mg/day achieved earlier prednisone discontinuation.

Source: Rheumatology (Oxford). 2016 Feb;55(2):347-56.

doi: 10.1093/rheumatology/kev348. Epub 2015 Sep 18.