



Facial nerve palsy in giant-cell arteritis: case-based review

Eveline Claeys^{1,4} · Olivier Gheysens² · Wouter Meersseman¹ · Eric Verbeken³ · Daniel Blockmans¹ · Liesbet Henckaerts¹

Received: 26 March 2020 / Accepted: 2 August 2020 / Published online: 13 August 2020
© Springer-Verlag GmbH Germany, part of Springer Nature 2020

Abstract

Acute peripheral facial nerve palsy is most frequently idiopathic (Bell's palsy) or virally induced, but can also be due to several other conditions. A rare cause is underlying systemic or autoimmune disease. A 79-year-old man presented with peripheral facial nerve palsy, malaise, and fever. Physical examination revealed tenderness of the left temporal artery and reduced pulsatility. ¹⁸F-FDG-PET/CT and biopsy of the temporal artery confirmed the diagnosis of giant cell arteritis (GCA). Prompt institution of corticosteroid therapy produced rapid decrease in inflammatory markers and gradual improvement of the facial nerve palsy. We searched the MEDLINE, Embase, and Scopus databases to identify previous reports of peripheral nerve palsy in GCA, other vasculitides, and autoimmune diseases. Facial nerve palsy as the presenting symptom of GCA has very rarely been reported. Although temporal artery biopsy is the gold standard for diagnosis, it may be negative in up to one-third of cases. In doubtful cases, imaging can help establish the diagnosis. Ultrasound, 3 T MRI, and ¹⁸F-FDG-PET/CT have all been previously reported to be useful. Peripheral facial nerve palsy may very rarely be the presenting symptom of GCA. Early correct diagnosis is essential for starting appropriate therapy. In patients with atypical features, ¹⁸F-FDG-PET/CT may be useful for establishing the diagnosis.

Keywords Giant cell arteritis · Vasculitis · Autoimmune disease · Facial nerve disease

Introduction

Giant cell arteritis (GCA) is the most common type of vasculitis in adults over 50 years of age, with an incidence of 20 per 100,000 in northern European populations. It affects large and medium-sized vessels, ultimately causing luminal occlusion and tissue ischemia. Patients typically present with new-onset severe headache and raised inflammatory markers [1]. Visual loss may occur in GCA, but other neurologic

manifestations are uncommon. Less than ten cases of GCA presenting with peripheral facial nerve palsy have been reported to date.

Acute peripheral facial nerve palsy has a variety of etiologies: it may be congenital or iatrogenic, or may be secondary to infections, malignancy, otologic disease, or systemic or autoimmune disease. Bell's palsy, which is the idiopathic form, and infections (varicella zoster, herpes simplex, or Lyme disease) are the most frequent causes; peripheral facial nerve palsy due to autoimmune disease is rare [2].

Treatment varies with the etiology of the nerve palsy. While Bell's palsy requires a short course of corticosteroid, nerve palsy due to systemic or autoimmune disease requires prolonged treatment with corticosteroid drugs, with or without other immunosuppressants. Therefore, peripheral facial palsy presenting with atypical features warrants thorough investigation.

In patients with GCA, early diagnosis and prompt institution of treatment (with corticosteroid and/or steroid-sparing agents) are crucial to preserve visual function [1, 3]. Temporal artery biopsy (TAB) is considered the gold standard for diagnosis of GCA, but it can be negative in 30% of patients.

✉ Eveline Claeys
eveline.claeys@uzleuven.be

¹ Department of General Internal Medicine, Department of Microbiology, Immunology and Transplantation, KU Leuven, University Hospitals Leuven, Herestraat 49, 3000 Leuven, Belgium

² Present Address: Department of Nuclear Medicine, Cliniques Universitaires Saint-Luc, Brussels, Belgium

³ Department of Translational Cell and Tissue Research, Department of Imaging and Pathology, KU Leuven, University Hospitals Leuven, Leuven, Belgium

⁴ Katholieke Universiteit Leuven Universitaire Ziekenhuizen Leuven, Leuven, Belgium

In doubtful cases, imaging studies may be useful. We report a patient with peripheral facial nerve palsy due to GCA in whom ultrasound and hybrid ^{18}F -fluorodeoxyglucose positron emission tomography with computed tomography scan (^{18}F -FDG-PET/CT) helped establish the diagnosis.

Case report

A 79-year-old man with history of aortic valve replacement and arterial hypertension presented to the emergency department with complaints of fever, general malaise, and headache for 5 days. On examination, he had definite left-sided peripheral facial nerve palsy (House–Brackmann grade III) along with some facial swelling. There were no signs of meningeal irritation, reduced visual acuity, or herpes zoster oticus. Tenderness and reduced pulsations of the left temporal artery raised suspicion of GCA with vasculitic neuropathy. Clinical examination was otherwise normal.

Laboratory analysis showed a markedly increased C-reactive protein (CRP) of 180 mg/L (reference range 0–5 mg/L). Antinuclear antibodies were positive at 1/320, but subtyping and anti-double-stranded DNA antibodies were negative. Cultures of blood and urine, and serology for *Borrelia burgdorferi*, cytomegalovirus, Epstein–Barr virus, and human immunodeficiency virus, were negative. Cranial CT, magnetic resonance imaging (MRI) of the brain, transesophageal echocardiography, and cerebrospinal fluid analysis were normal. Ultrasound of the temporal arteries

demonstrated bilateral halo sign. ^{18}F -FDG-PET/CT showed increased ^{18}F FDG-uptake in the maxillary, temporal, vertebral, carotid, subclavian, brachial, internal thoracic, celiac, lumbar, and femoral arteries, and to a lesser extent in the aortic wall (Fig. 1). Temporal artery biopsy confirmed the diagnosis of GCA (Fig. 2).

Treatment was immediately started with high-dose corticosteroid (methylprednisolone 32 mg orally daily). CRP decreased to normal level within days, and facial nerve function gradually returned to normal over 2 months.

Literature review

We used the keywords “facial nerve disease”, “autoimmune disease”, and “vasculitis” to search the MEDLINE, Embase, and Scopus databases for articles related to peripheral facial nerve palsy published up to 4 July 2020. A total of 1799 articles were identified. The titles and abstracts were first screened, and papers on patients < 18 years were excluded. The full texts of the remaining articles were then examined, and 37 papers were selected for inclusion in this review. Additional relevant papers were identified from the reference lists and from a search of the grey literature.

Results of the review

In a large retrospective review of 1989 patients with facial palsy, only 1.6% of cases were attributable to autoimmune disease (i.e., Guillain–Barré, Melkersson–Rosenthal,

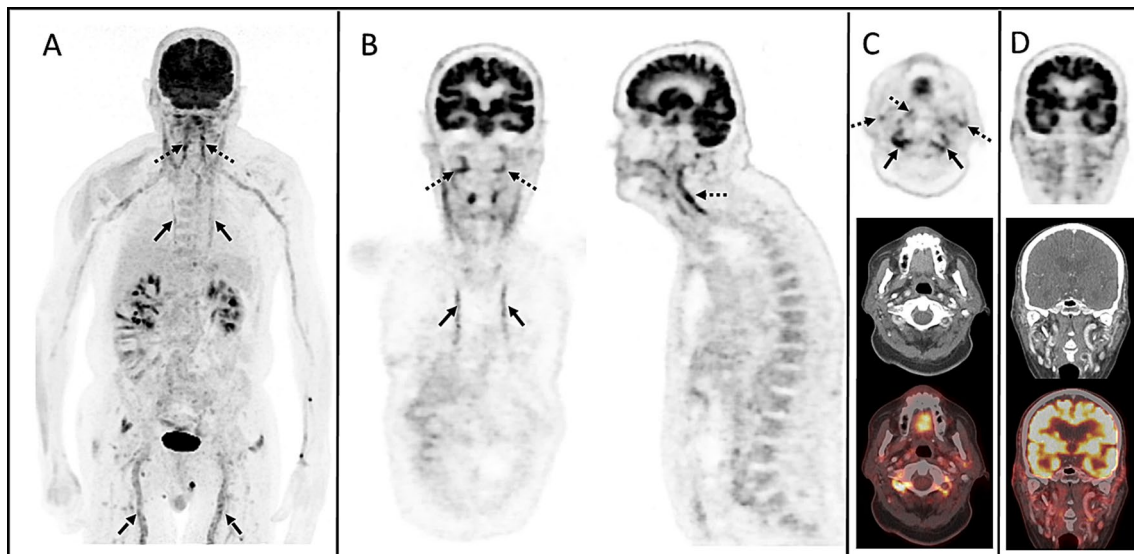


Fig. 1 ^{18}F -FDG PET images. **a** ^{18}F -FDG PET maximum intensity projection image show moderately to strongly increased tracer uptake in the vertebral, carotid, subclavian, brachial, and internal thoracic arteries. **b** Coronal and sagittal PET images show increased uptake in the internal thoracic (full arrow) and vertebral (dashed arrow) arteries.

c Transaxial PET, CT, and fused PET-CT images show increased uptake in the maxillary artery and its branches (dashed arrows) and the vertebral arteries (full arrow). **d** Coronal PET, CT, and fused PET-CT images reveal increased activity at the temporal artery (dashed arrow)

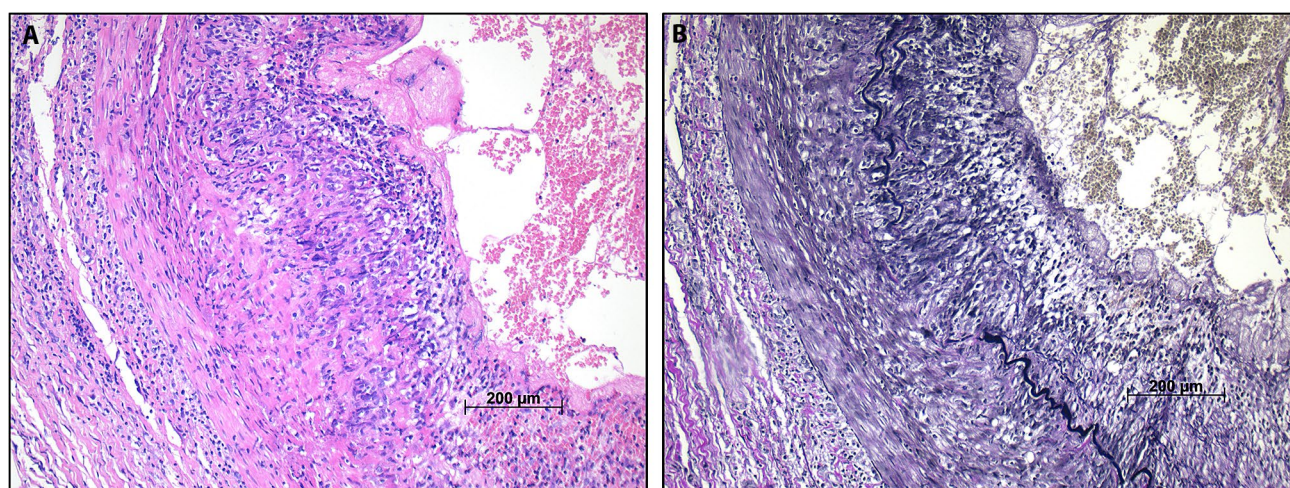


Fig. 2 Histopathology of the temporal artery. **a:** Giant cell (arrow); hematoxylin and eosin stain. **b:** Disruption of the internal elastic lamina; Verhoeff -van Gieson stain

Table 1 Facial nerve palsy in autoimmune systemic diseases

Vasculitis

- Giant cell arteritis [5–8]
- Takayasu's arteritis [9]
- Polyarteritis nodosa [10, 11]
- Granulomatosis with polyangiitis [12–14]
- Eosinophilic granulomatosis with polyangiitis [15]

Connective tissue disease

- Systemic lupus erythematosus [16]
- Sjögren syndrome [16, 17]
- Behçet's disease [18, 19]
- Sarcoidosis [2]
- Systemic sclerosis [16, 20]
- Amyloidosis [2]
- Antiphospholipid syndrome [2]

(Neurologic) autoimmune disease

- Guillain-Barré syndrome [2]
- Multiple sclerosis [2]
- Melkersson-Rosenthal syndrome [2]

sarcoidosis, multiple sclerosis, amyloidosis, and antiphospholipid syndrome) [2].

The autoimmune conditions associated with peripheral facial palsy (Table 1) can be categorized into three broad groups: (1) vasculitis, (2) connective tissue disease, and (3) neurological autoimmune syndrome (the last category is not further discussed in this paper) [2, 4].

Facial nerve palsy in vasculitis

Facial nerve palsy has been reported in several of the vasculitides. In small- and medium-vessel vasculitides,

such as polyarteritis nodosa (PAN) and the antineutrophil cytoplasmic antibody-associated vasculitides, neuropathy is due to ischemia caused by arteritis of the vasa nervorum. Necrotizing arteritis is also observed in the setting of vasculitis secondary to other systemic diseases [21, 22].

Facial nerve palsy is reported in 5–8% of patients with granulomatosis with polyangiitis (GPA), and is widely believed to be due to necrotizing vasculitis of the facial nerve vasa nervorum [12]; however, some case reports suggest that facial nerve palsy may be secondary to the otitis media or mastoiditis often seen in GPA [13, 14]. PAN and eosinophilic granulomatosis with polyangiitis are less common causes of facial nerve palsy [10, 11, 15].

Peripheral facial nerve palsy as the presenting symptom of GCA, as in our patient, has rarely been reported. The most feared (and probably best known) neurologic complication of GCA is anterior ischemic optic neuropathy, that can lead to permanent visual loss [3]. Other, less common, neurological manifestations include diplopia, diabetes insipidus, seizure, paralysis, neuropathies, ataxia, and vertigo [23]. Caselli et al. reported neurological disease in 31% of 166 biopsy-proven cases of GCA. Among the 166 patients, 31% had large-artery abnormalities (leading to stroke or transient ischemic attack), 21% had neuro-ophthalmologic problems, and 14% had peripheral mono- or polyneuropathies [24]. Less than ten cases of GCA presenting with peripheral facial nerve palsy have been reported to date, the most recent one being in 1991 [5–8]. The exact pathophysiology remains unclear. Meadows et al. reported a case of GCA with facial palsy secondary to brain-stem infarction caused by occlusive arteritis of the vertebral arteries [25].

Our literature search yielded two reports of facial nerve palsy in patients with other types of large-vessel vasculitides,

one with Takayasu arteritis and another with isolated cerebral granulomatous vasculitis with giant cells [9, 26].

Facial nerve palsy in connective tissue disease

Peripheral facial nerve palsy has been recorded in several connective tissue diseases. Cranial neuropathy (including facial weakness) or peripheral neuropathy may occur in up to 15% of patients with systemic lupus erythematosus; isolated facial nerve palsy has also been reported in a few cases [16]. The exact aetiology remains unclear. Proposed pathophysiological mechanisms include thrombotic vasculopathy mediated by antiphospholipid antibodies, development of autoimmune antibodies against neuronal antigen, or infection-triggered production of inflammatory cytokines (causing neuritis) [27].

Sjögren's syndrome may manifest with both central and peripheral nervous system symptoms. Cranial nerve involvement is usually of the trigeminal nerve, but a few cases of peripheral facial nerve palsy have been reported [16, 17]. Although vasculitis is not a prominent feature of Sjögren's syndrome (present in < 10% of patients), it has been proposed as the mechanism causing mononeuropathy and multiple neuropathy [28]. Cryoglobulinemia in Sjögren's syndrome may cause vasculitis in a subset of patients and lead to sensorimotor polyneuropathy [29, 30].

Trigeminal sensory neuropathy and other cranial neuropathies (such as facial nerve palsy) have been described in patients with cutaneous system sclerosis [16, 20]. A study investigating the pathophysiological mechanism underlying neuropathy in scleroderma found that small-caliber unmyelinated nerve fibers were prominently affected, especially in patients with vascular bed changes suggestive of ischemia [31].

Recurrent peripheral facial palsy is a very rare clinical manifestation of Behçet's disease [18, 19]. Electroneuromyographic studies in patients with peripheral nervous involvement in Behçet's disease showed demyelination, chronic denervation, and even myogenic involvement; however, the exact pathogenesis remains unclear [32].

Finally, our literature search also found two reports that mention IgG4-related disease as the cause of facial nerve palsy, probably via involvement of the parotid gland, inner ear, or lateral skull base [33, 34].

Discussion

Early correct identification of the cause of peripheral facial nerve palsy is essential for starting appropriate immunosuppressive treatment and preventing irreversible damage.

The diagnosis of GCA is mainly based on clinical findings, raised inflammatory markers, and the typical histopathological findings on TAB (vasculitis with granulomatous inflammation or mononuclear cells, often with multinucleated giant cells) [1, 3]. However, TAB may be negative in up to one-third of patients with GCA, due to the segmental nature of the inflammatory infiltrate [35]. In doubtful cases, imaging can be useful. The halo sign on ultrasound of the temporal artery has sensitivity of 77% and specificity of 96% for diagnosis of GCA [36]. In a recent prospective study, 3 T MRI had sensitivity of 93.6% and specificity of 77.9% for diagnosis of temporal artery involvement in GCA [35, 37]. Computed tomography angiography is useful for detection of acute stenotic complications or late complications, such as aneurysm formation, but its usefulness for diagnosis of GCA remains to be established [38].

The present case report demonstrates that ^{18}F -FDG-PET/CT can be helpful for early diagnosis of GCA in patients with atypical presentation [39]. The temporal arteries, due to their small caliber and their location just superficial to the high physiological FDG uptake of the brain, are generally considered difficult to evaluate with ^{18}F -FDG-PET/CT. The latest European League Against Rheumatism (EULAR) guidelines do not yet recommend ^{18}F -FDG-PET/CT for assessment of inflammation in cranial arteries. However, as demonstrated in our patient, recent advances in imaging equipment and reconstruction have made visualization of significant FDG uptake in the temporal and other cranial arteries possible. A recent retrospective case–control study in 44 glucocorticoid-naïve GCA patients has also demonstrated the usefulness of ^{18}F -FDG-PET/CT [40, 41].

Conclusion

We report an extremely rare case of GCA presenting with peripheral facial nerve palsy and review the literature on the association between autoimmune diseases (including the vasculitides) and peripheral facial nerve palsy. Early correct diagnosis is essential for selection of the appropriate immunosuppressant therapy. ^{18}F -FDG-PET/CT can be useful for establishing the diagnosis in doubtful cases.

Author contributions EC and LH performed the literature search and wrote the manuscript in consultation with OG, BD, EV and WM. OG interpreted the PET-CT images and provided corresponding images and caption. EV interpreted the anatomopathological data and provided corresponding images and caption. All authors have read and approved the final version of the manuscript as submitted, and agree to be accountable for all aspects of the work.

Funding The authors received no specific funding for this work.

Compliance with ethical standards

Conflict of interest The authors report no conflict of interest relevant to this work.

Informed consent We obtained written informed consent of the patient prior to the submission of this paper.

References

- Weyand CM, Goronzy JJ (2014) Giant-cell arteritis and polymyalgia rheumatica. *N Engl J Med* 371:1653. <https://doi.org/10.1056/NEJMc1409206>
- Hohman MH, Hadlock TA (2014) Etiology, diagnosis, and management of facial palsy: 2000 patients at a facial nerve center. *Laryngoscope* 124:E283–293. <https://doi.org/10.1002/lary.24542>
- Gonzalez-Gay MA, Martinez-Dubois C, Agudo M, Pompei O, Blanco R, Llorca J (2010) Giant cell arteritis: epidemiology, diagnosis, and management. *Curr Rheumatol Rep* 12:436–442. <https://doi.org/10.1007/s11926-010-0135-9>
- Tm O (2018) Medical management of acute facial paralysis. *Otolaryngol Clin N Am* 51:1051–1075. <https://doi.org/10.1016/j.otc.2018.07.004>
- Balquet MH, Gosset D, Bouchez B, Mellin D, Hatron PY, Devulder B (1991) Peripheral nerve diseases disclosing Horton's disease (2 cases). *Rev Med Intern* 12:57–59. [https://doi.org/10.1016/s0248-8663\(05\)81356-6](https://doi.org/10.1016/s0248-8663(05)81356-6)
- Taillan B, Fuzibet JG, Vinti H, Castela J, Gratecos N, Dujardin P, Pesce A (1989) Peripheral facial paralysis disclosing Horton's disease. *Rev Rhum Mal Osteoartic* 56:342
- Treves R, Archambeaud-Mouveau F, Liozon E (1991) Rare neurologic forms disclosing Horton's disease. 2 cases. *Ann Med Intern* 142:318–319
- Roomet A, Allen JS (1974) Temporal arteritis heralded by facial nerve palsy. *JAMA* 228:870–871
- Sotoudeh Anvari M, Masoudkabar F, Abbasi K, Boroumand MA, Zarghampour M, Goodarzynjad H (2016) Takayasu's arteritis presenting with headache and peripheral facial palsy: a case report. *J Tehran Heart Cent* 11:195–197
- Joglekar S, Deroee AF, Morita N, Cureoglu S, Schachern PA, Paparella M (2010) Polyarteritis nodosa: a human temporal bone study. *Am J Otolaryngol* 31:221–225. <https://doi.org/10.1016/j.amjoto.2009.02.006>
- Teichman JM, Mattrey RF, Demby AM, Schmidt JD (1993) Polyarteritis nodosa presenting as acute orchitis: a case report and review of the literature. *J Urol* 149:1139–1140. [https://doi.org/10.1016/s0022-5347\(17\)36322-x](https://doi.org/10.1016/s0022-5347(17)36322-x)
- Iannella G, Greco A, Granata G, Manno A, Pasquariello B, Angeletti D, Didona D, Magliulo G (2016) Granulomatosis with polyangiitis and facial palsy: literature review and insight in the autoimmune pathogenesis. *Autoimmun Rev* 15:621–631. <https://doi.org/10.1016/j.autrev.2016.02.005>
- Renz LD, Seidl R, Dreyer J, Niedobitek G, Ernst A (2019) Case report: a 20 years old patient with chronic mastoiditis and facial nerve palsy within a major-recidive of a granulomatosis with polyangiitis. *Laryngo Rhino Otol* 98:1
- Preuss SF, Stenner M, Beutner D, Laudes M, Klusmann JP (2008) Fatal course of Wegener's granulomatosis with bilateral otomastoiditis and bilateral facial nerve palsy. *Otolaryngol Head Neck* 138:799–800. <https://doi.org/10.1016/j.otohns.2007.12.045>
- Byun JH, Lee JH, Choi IS (2014) Churg-strauss syndrome presented with hearing impairment and facial palsy. *Ann Rehabil Med* 38:852–855. <https://doi.org/10.5535/arm.2014.38.6.852>
- Cikes N (2006) Central nervous system involvement in systemic connective tissue diseases. *Clin Neurol Neurosurg* 108:311–317. <https://doi.org/10.1016/j.clineuro.2005.11.009>
- Hadithi M, Stam F, Donker AJ, Dijkmans BA (2001) Sjogren's syndrome: an unusual cause of Bell's palsy. *Ann Rheum Dis* 60:724–725. <https://doi.org/10.1136/ard.60.7.724>
- Hatzitolios A, Savopoulos C, Ntaios G, Tsirogianni E, Baltatzis M, Apostolopoulou M, Dimitrakoudi E (2008) Recurrent peripheral facial paresis may constitute the sole clinical manifestation in neuro-behçet disease. *Neurologist* 14:77. <https://doi.org/10.1097/NRL.0b013e31815879f5>
- Tsirogianni ES, Hatzitolios AI, Savopoulos CG, Baltatzis MS, Apostolopoulou MI, Magripli CM, Dimitrakoudi ET (2004) Neuro-Behçet's syndrome may present with peripheral paresis of the facial nerve. *J Peripher Nerv Syst* 9:59–61. <https://doi.org/10.1111/j.1085-9489.2004.009203.x>
- Amaral TN, Peres FA, Lapa AT, Marques-Neto JF, Appenzeller S (2013) Neurologic involvement in scleroderma: a systematic review. *Semin Arthritis Rheum* 43:335–347. <https://doi.org/10.1016/j.semarthrit.2013.05.002>
- Vrancken AF, Said G (2013) Vasculitic neuropathy. *Handbook of clinical neurology*, pp 463–483. <https://doi.org/10.1016/B978-0-444-52902-2.00026-6>
- Imboden JB (2017) Involvement of the peripheral nervous system in polyarteritis nodosa and antineutrophil cytoplasmic antibodies-associated vasculitis. *Rheum Dis Clin N Am* 43:633–639. <https://doi.org/10.1016/j.rdc.2017.06.011>
- Nesher G (2000) Neurologic manifestations of giant cell arteritis. *Clin Exp Rheumatol* 18:S24–26
- Caselli RJ, Daube JR, Hunder GG, Whisnant JP (1988) Peripheral neuropathic syndromes in giant cell (temporal) arteritis. *Neurology* 38:685–689. <https://doi.org/10.1212/wnl.38.5.685>
- Meadows SP (1966) Temporal or giant cell arteritis. *Proc R Soc Med* 59:329–334
- Broussalis E, Luthringshausen G, Kunz AB, McCoy M, Weiss S, Killer-Oberpfalzer M, Trinka E, Kraus J (2011) Isolated facial palsy and ophthalmoplegia—first symptoms of a cerebral granulomatous vasculitis with giant cells—a diagnostic challenge. *Clin Neurol Neurosurg* 113:686–688. <https://doi.org/10.1016/j.clineuro.2011.04.007>
- Kazzaz NM, El-Rifai R (2013) Unusual aetiology of isolated lower motor neuron facial palsy: systemic lupus erythematosus presenting with cranial nerve palsy and nephritis. *BMJ Case Rep* 2013:200378. <https://doi.org/10.1136/bcr-2013-200378>
- Pavakis PP, Alexopoulos H, Kosmidis ML, Mamali I, Moutsopoulos HM, Tzioufas AG, Dalakas MC (2012) Peripheral neuropathies in Sjogren's syndrome: a critical update on clinical features and pathogenetic mechanisms. *J Autoimmun* 39:27–33. <https://doi.org/10.1016/j.jaut.2012.01.003>
- Sene D, Jallouli M, Lefacheur JP, Saadoun D, Costedoat-Chalumeau N, Maisonobe T, Diemert MC, Musset L, Haroche J, Piette JC, Amoura Z, Cacoub P (2011) Peripheral neuropathies associated with primary Sjogren syndrome: immunologic profiles of nonataxic sensory neuropathy and sensorimotor neuropathy. *Medicine* 90:133–138. <https://doi.org/10.1097/MD.0b013e31820fd2d1>
- Ramos-Casals M, Anaya JM, Garcia-Carrasco M, Rosas J, Bove A, Claver G, Diaz LA, Herrero C, Font J (2004) Cutaneous vasculitis in primary Sjogren syndrome: classification and clinical significance of 52 patients. *Medicine* 83:96–106. <https://doi.org/10.1097/01.md.0000119465.24818.98>
- Paik JJ, Mammen AL, Wigley FM, Shah AA, Hummers LK, Polydefkis M (2016) Symptomatic and electrodiagnostic features

- of peripheral neuropathy in scleroderma. *Arthritis Care Res* 68:1150–1157. <https://doi.org/10.1002/acr.22818>
32. Siva A, Saip S (2009) The spectrum of nervous system involvement in Behcet's syndrome and its differential diagnosis. *J Neurol* 256:513–529. <https://doi.org/10.1007/s00415-009-0145-6>
 33. Farahani SA, Hosseini MME, Nozarian Z, Yazdani N (2019) IgG4-Related disease causing facial and recurrent laryngeal nerve palsies: a case report. *J Clin Diagn Res* 13:3
 34. Wick CC, Zachariah J, Manjila S, Brown WC, Malla P, Katirji B, Cohen M, Megerian CA (2016) IgG4-related disease causing facial nerve and optic nerve palsies: Case report and literature review. *Am J Otolaryngol* 37:567–571. <https://doi.org/10.1016/j.amjoto.2016.08.005>
 35. Ninan JV, Lester S, Hill CL (2017) Giant cell arteritis: beyond temporal artery biopsy and steroids. *Intern Med J* 47:1228–1240. <https://doi.org/10.1111/imj.13483>
 36. Schmidt WA (2018) Ultrasound in the diagnosis and management of giant cell arteritis. *Rheumatology (Oxford)* 57:22–31. <https://doi.org/10.1093/rheumatology/kex461>
 37. Rheaume M, Rebello R, Pagnoux C, Carette S, Clements-Baker M, Cohen-Hallaleh V, Doucette-Preville D, Stanley Jackson B, Salama Sargious Salama S, Ioannidis G, Khalidi NA (2017) High-resolution magnetic resonance imaging of scalp arteries for the diagnosis of giant cell arteritis: results of a prospective cohort study. *Arthritis Rheumatol* 69:161–168. <https://doi.org/10.1002/art.39824>
 38. Slart R, Writing G, Reviewer G, Members of EC, Members of EI, Inflammation, Members of Committees SC, Members of Council PETIG, Members of A, Coordinator EC (2018) 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. *Eur J Nucl Med Mol* 45:1250–1269. <https://doi.org/10.1007/s00259-018-3973-8>
 39. Salvarani C, Soriano A, Muratore F, Shoenfeld Y, Blockmans D (2017) Is PET/CT essential in the diagnosis and follow-up of temporal arteritis? *Autoimmun Rev* 16:1125–1130. <https://doi.org/10.1016/j.autrev.2017.09.007>
 40. Hellmich B, Agueda A, Monti S, Buttgereit F, de Boysson H, Brouwer E, Cassie R, Cid MC, Dasgupta B, Dejaco C, Hatemi G, Hollinger N, Mahr A, Mollan SP, Mukhtyar C, Ponte C, Salvarani C, Sivakumar R, Tian X, Tomasson G, Turesson C, Schmidt W, Villiger PM, Watts R, Young C, Luqmani RA (2020) 2018 Update of the EULAR recommendations for the management of large vessel vasculitis. *Ann Rheum Dis* 79:19–30. <https://doi.org/10.1136/annrheumdis-2019-215672>
 41. Nielsen BD, Hansen IT, Kramer S, Haraldsen A, Hjorthaug K, Bogsrud TV, Ejlersen JA, Stolle LB, Keller KK, Therkildsen P, Hauge EM, Gormsen LC (2019) Simple dichotomous assessment of cranial artery inflammation by conventional 18F-FDG PET/CT shows high accuracy for the diagnosis of giant cell arteritis: a case-control study. *Eur J Nucl Med Mol Imaging* 46:184–193. <https://doi.org/10.1007/s00259-018-4106-0>

Publisher's Note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.