Letter to the Editor (Case report)

Rheumatology Advances in Practice;0:1–3
doi: 10.1093/rap/rky020

Ophthalmoplegia in an elderly woman with giant cell arteritis

Key message
- GCA can present atypically. Neurological abnormalities are important in considering GCA as a differential diagnosis.

SIR, an 87-year-old woman reported a 2-week history of binocular diplopia, progressive left-sided ptosis and left-sided scalp tenderness. Her symptoms developed within days of a fall from a stepladder, when she sustained a minor head injury with no loss of consciousness. She was seen in her local emergency room and had a head CT scan that was remarkable only for a left subgaleal haematoma. She was discharged home with no specific treatment and advised to follow up with her family practitioner. Two weeks later, she presented to her optometrist with a gradual onset of left-sided ptosis and lateral gaze deviation. He sent her to the emergency room, where she was referred to a neurologist. On probing, she did endorse new-onset jaw claudication and some scalp tenderness with palpation around the area where she sustained the head injury. She denied any constitutional symptoms.

On examination, there was full left ptosis and a pupil-involving left third nerve palsy (Fig. 1A and B). Written consent was obtained for all photographs. She also had restricted left eye abduction, suggesting simultaneous sixth cranial nerve palsy. Her visual acuity was normal. Fundoscopic examination revealed a normal optic disc, with no evidence of anterior ischaemic optic neuropathy. The remainder of the cranial nerve, motor, sensory, coordination and gait examinations were unremarkable. There was no temporal tenderness or proximal muscle weakness. Temporal artery pulsations were normal bilaterally, as were peripheral pulses, with no bruits. She had mildly elevated CRP of 10.4 mg/l (upper limit of normal <6 mg/l), and her ESR was 30 mm/h, which is normal for her age.

Owing to her history of jaw claudication and temporal artery tenderness on palpation, although this was not elucidated on her physical examination, her neurologist was suspicious of GCA and referred the patient to a rheumatologist. The neurologist’s differential also included any lesions that would affect both third and sixth cranial nerves, including lesions in the orbital apex, a cavernous sinus thrombosis, a posterior communicating artery aneurysm, a brainstem lesion affecting both the nerve nuclei or fascicles or a diffuse leptomeningeal process. For this reason, the neurologist ordered an MRI and angiography (MRI/MRA), which came back as normal apart from an incidental 1.3 mm middle cerebral artery aneurysm.

Owing to the lack of inflammatory markers and other typical findings of GCA, her rheumatologist initially suspected a traumatic cause for her ptosis and lateral gaze deviation. As she knew she could not fully explain all the symptoms by trauma and she knew the ophthalmological consequences of leaving GCA untreated, she prudently ordered an urgent US and temporal artery biopsy and started the patient on oral prednisone 50 mg daily empirically. The temporal artery biopsy showed histological features consistent with temporal arteritis. In follow-up 2 weeks later, her palsies were significantly improved (Fig. 1C and D).

GCA is a chronic inflammatory vasculitis that affects large arteries. Up to 70% present with the classic cranial symptoms: new-onset headache, jaw or tongue claudication, scalp tenderness or neck pain.

Diagnosis becomes challenging when patients present with non-classic symptoms, including ophthalmological symptoms. Ophthalmological symptoms can include anterior ischaemic optic neuropathy, retinal ischaemia, choroidal ischaemia, scleritis, peripheral ulcerative keratitis and ophthalmoplegia [1].

Anterior ischaemic optic neuropathy is the most common ocular manifestation, with involvement of posterior ciliary branches of the ophthalmic artery [2]. This can lead to permanent vision loss without treatment. Ophthalmoplegia is an uncommon manifestation, with proposed pathomechanisms including muscular dysfunction, neuronal ischaemia or orbital pseudotumour [1,3,4]. Extraocular muscle ischaemia involves the blood vessels directly feeding those muscles supplied by the lateral and medial muscular branches of the ophthalmic artery [3,5]. Ophthalmoplegia can also occur as a result of microvascular ischaemia of cranial nerves supplied by a complex network of branches derived from the anterior and posterior cerebral circulations [4]. The most commonly involved is the third cranial nerve. In GCA, the third nerve palsy is usually pupil sparing, similar to other microvascular third nerve palsies, because the parasympathetic fibres to the pupil are located circumferentially and receive collateral blood supply; they are, therefore, relatively protected from ischaemia but more prone to compression [5]. There are only a handful of published case reports in which a third nerve palsy is the initial manifestation of GCA and only three that describe pupil involvement [6–8].

This case report serves to remind practitioners that GCA can present with predominantly, and sometimes solely, ophthalmic manifestations. It is important when assessing an elderly patient with new-onset ophthalmic symptoms to screen for other GCA symptoms because...
they may uncover subtle symptoms that the patient had
not noticed themselves and lead to the correct diagno-
sis. If the patient lacks systemic features and inflamma-
tory markers, but the diagnosis remains elusive, recent
advances in imaging modalities (US and high-resolution
MRI of the scalp arteries), if available, can assist the cli-

Funding: No specific funding was received from any fund-
ing bodies in the public, commercial or not-for-profit sec-
tors to carry out the work described in this manuscript.

Disclosure statement: The authors have declared no
conflict of interest.

Ryan Quinn1, Christine Hawkes2, Amina Lodhi3,
Shangguo Tang4, Karen A. Beattie3,
Brian van Adel2 and Maggie J. Larche4
1Department of Medicine, 2Department of Medicine,
Division of Neurology, 3Department of Medicine, Division
of Rheumatology, 4Department of Pathology and
Molecular Medicine, McMaster University, Hamilton, ON,
Canada
Accepted 17 May 2018
Correspondence to: Karen A. Beattie, Department of
Medicine, Division of Rheumatology, McMaster University,
501-25 Charlton Avenue East, Hamilton, ON L8N 1Y2,
Canada. E-mail: beattik@mcmaster.ca

Fig. 1 Patient as presented before and after treatment

Ptosis of the left eye (A), with the characteristic ‘down and out’ displacement of the eye and a mydriatic pupil on the
left side (B), typical of a third nerve palsy (C), and complete resolution by 2 weeks following high-dose prednisone (D).

https://academic.oup.com/rheumap
Ophthalmoplegia in an elderly woman

References

1 De Smit E, O’Sullivan E, Mackey DA, Hewitt AW. Giant cell arteritis: ophthalmic manifestations of a systemic disease. Graefe’s Arch Clin Exp Ophthalmol 2016;254:2291–306.

2 Hayreh SS, Podhajsky PA, Zimmerman B. Ocular manifestations of giant cell arteritis. Am J Ophthalmol 1998;125:509–20.

3 Mehler MF, Rabinowich L. The clinical neuro-ophthalmologic spectrum of temporal arteritis. Am J Med 1988;85:839–44.

4 Hendrix P, Griessenauer CJ, Foreman P et al. Arterial supply of the upper cranial nerves: a comprehensive review. Clin Anat 2014;27:1159–66.

5 Hamed LM, Guy JR, Moster ML, Bosley T. Giant cell arteritis in the ocular ischemic syndrome. Am J Ophthalmol 1992;113:702–5.

6 Thurtell MJ, Longmuir RA. Third nerve palsy as the initial manifestation of giant cell arteritis. J Neuroophthalmol 2014;34:243–5.

7 Asensio-Sánchez VM, Morales-Gómez I, Rodríguez-Vaca I. [Third nerve palsy as the only manifestation of occult temporal arteritis]. Arch Soc Esp Oftalmol 2009;84:395–8.

8 Rabinowich L, Mehler MF. Parasympathetic pupillary involvement in biopsy-proven temporal arteritis. Ann Ophthalmol 1988;20:400–2.