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Letter to the Editor (Case report)

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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, co-ordination 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

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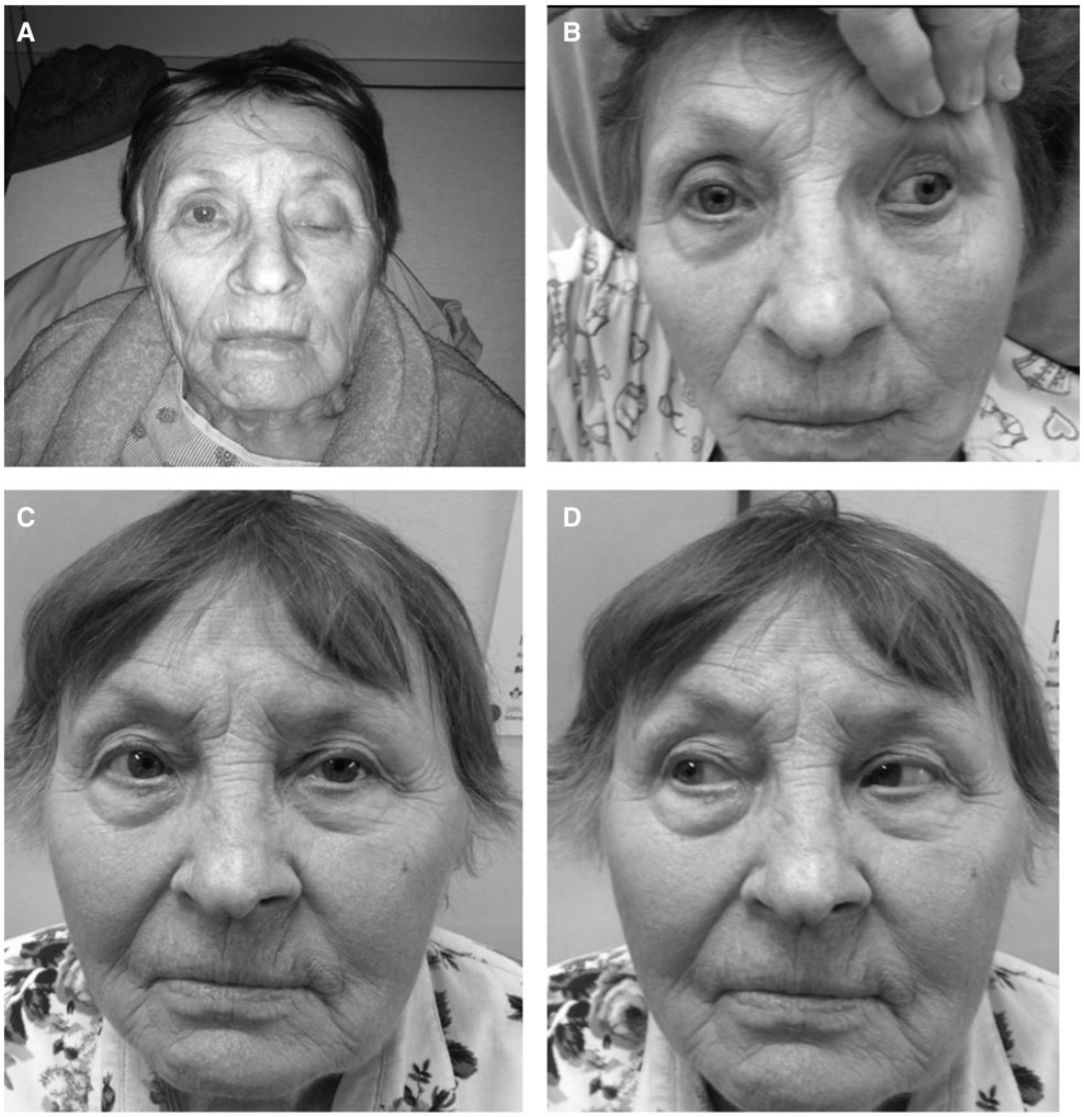
Fig. 1 Patient as presented before and after treatment



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**).

they may uncover subtle symptoms that the patient had not noticed themselves and lead to the correct diagnosis. If the patient lacks systemic features and inflammatory 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 clinician in ruling GCA in or out.

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