Giant cell arteritis presenting as painful third nerve palsy with normal erythrocyte sedimentation rate

Introduction

This case presents a man with painful diplopia and a normal erythrocyte sedimentation rate (ESR), found to be caused by giant cell arteritis (GCA). Diplopia in GCA is an important sign of impending visual loss, requiring urgent treatment.

Discussion

The incidence of diplopia in GCA is reported from 2–15% (Hayreh, 1991), but the incidence of ophthalmoplegia may be higher. In a study of patients with biopsy-proven GCA, Dimant et al (1980) described seven patients with ophthalmoplegia of whom only one had diplopia. The failure to find ophthalmoplegia in patients with diplopia may be related to the transient nature of the phenomenon. In 175 patients with GCA seen at the Mayo clinic, 22 complained of diplopia, with ophthalmoplegia only documented in 10 (Hollenhorst et al, 1960). In two of these, the particular muscle involvement changed from day to day.

The variable nature of ophthalmoplegia, together with its rapid recovery with corticosteroid therapy, has led to suggestions that extraocular muscle ischaemia, rather than oculomotor nerve ischaemia, is responsible (Barricks et al, 1977; Dimant et al, 1980). Barricks et al found severe ischaemic post-mortem changes in extraocular muscles, but not nerves, in one patient with GCA. Conversely, Sibony and Lessell (1984) found transient aberrant regeneration in another patient, which may be more consistent with a neurogenic than a myogenic cause.

Diplopia in GCA is a warning sign for impending loss of vision and should prompt urgent treatment (Hollenhorst et al, 1960). Of 22 patients in this series, 10 later lost vision in one or both eyes; two had amaurosis fugax and four, with no visual impairment, had clinical evidence of retinal ischaemia. Corticosteroid therapy generally leads to complete or partial recovery of ophthalmoplegia, sometimes within 24 hours.

An elevated ESR is often found in GCA, but up to 22.5% of patients with GCA have a normal ESR before treatment (Salvarani and Hunder, 2001). In a prospective study of 363 patients with GCA over 20 years, Hayreh et al (1997) found that the ESR of patients with biopsy-proven GCA ranged from 4–140 mm/hr (median 87.5 mm/hr). Finding a low ESR in combination with ophthalmoplegia, however, is unusual.

Conclusions

This case is unusual in both its clinical presentation and laboratory features. Ophthalmoplegia in GCA is uncommon, but well recognized, is usually associated with elevated inflammatory markers and is often a harbinger of subsequent visual loss. A low ESR should not exclude the diagnosis of GCA and a high index of suspicion is necessary in all such atypical cases. BJHM

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Figure 1. Chronic inflammatory cells infiltrate the artery wall. A giant cell is seen (arrow).

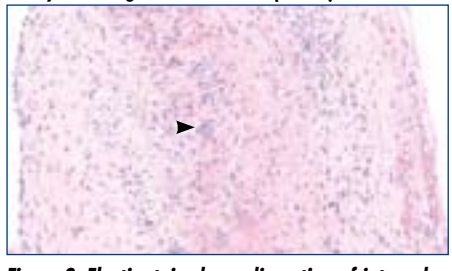
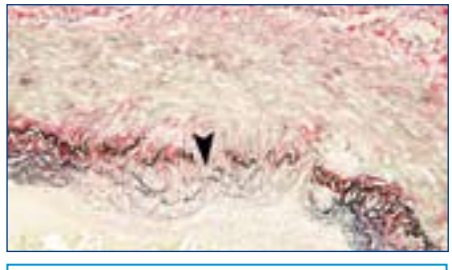


Figure 2. Elastic stain shows disruption of internal elastic lamina (arrow).



Case Report

An 80-year-old man was referred to hospital with a 2-week history of headache and 1 day of vertical diplopia, but was systemically well with no symptoms of giant cell arteritis (GCA). Past medical history included right central retinal vein occlusion, for which he had previously received panretinal photocoagulation. His full blood count and blood glucose were normal, erythrocyte sedimentation rate (ESR) 24 mm/hr and C-reactive protein (CRP) 34 mg/litre. Two days later, an ophthalmological review was requested.

The visual acuity was 6/36 right eye and 6/9 left. There was a moderate exotropia with diplopia and a left ptosis with restriction of both elevation and adduction of the left eye; depression of the left eye was normal. The left fourth nerve appeared intact. Neither optic disc appeared swollen. In view of the painful left partial third nerve palsy, urgent magnetic resonance angiography was performed but showed no significant abnormality, suggesting a microvascular cause. A Fresnel prism was fitted to his spectacles with resolution of diplopia.

Three weeks later, his symptoms had resolved. Although he remained well and had no symptoms of GCA, his left superficial temporal artery was markedly swollen, albeit non-tender. His ESR was 25 mm/hr and CRP 30 mg/litre. In light of his unusually rapid recovery, a diagnosis of atypical GCA was made and prednisolone started. Temporal artery biopsy confirmed GCA (*Figures 1* and *2*).

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