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Case Report

CASE REPORT

An interesting case of temporal arteritis that manifested as ptosis and diplopia

Anoka Martis[1,*,†], Romesa Sajjad Hassan[1], Ana Goico Alburquerque[2], Manasa Kotte[1], Anshu Martis[3] and Hassan Ahmed[4]

[1]Internal Medicine, Advocate Illinois Masonic Medical Center, Chicago, IL 60657, USA, [2]Rheumatology, University of Illinois at Chicago, Chicago, IL 60612, USA, [3]Internal Medicine, Father Muller Medical College, Mangalore 575002, India, [4]Internal Medicine, Dow Medical College, Karachi 74200, Pakistan

*Correspondence address. Internal Medicine Department, Advocate Illinois Masonic Medical Center, 836 W Wellington Avenue, Chicago, 60657, Illinois, USA. Tel: 312.383.9314; Fax: 773.296.7486; E-mail: anoka.martis@advocatehealth.com

Abstract

Giant cell arteritis (GCA) or temporal arteritis is a granulomatous vasculitis that affects medium-to-large vessels seen primarily in older Caucasian populations. Here, we describe a 67-year-old male who presented with atypical symptoms of worsening headaches associated with left-sided pupil-sparing, isolated third nerve palsy, blurry vision, diplopia and myalgias in bilateral extremities. He was immediately started on intravenous Methylprednisolone for suspected GCA. Subsequent biopsy of the temporal arteries showed panarteritis without giant cells and disruption of the internal elastic lamina. His symptoms improved in a day following treatment and he was discharged on a Prednisone taper. At the time of writing this case, there are only two cases in the literature of ptosis as a presenting symptom in GCA, thus highlighting the importance of recognizing rare red flag symptoms such as ptosis and diplopia. More study is needed in the prognostic significance of these unusual clinical features.

INTRODUCTION

Giant cell arteritis (GCA), also commonly known as temporal arteritis, is a disease with severe morbidity, largely due to the devastating permanent visual loss that can affect up to 15–25% of cases [1]. It remains a disease of older Caucasians, with the highest incidence noted in Scandinavians and populations descended from them [1]. Although most healthcare providers are aware of the common red flag symptoms of GCA such as blurriness of vision, headache and jaw claudication, lesser known manifestations such as isolated third nerve palsies (ptosis), diplopia and Horner's syndrome can also occur. In fact, there are only two reported cases in the literature of isolated third nerve palsies [2, 3], highlighting the significance of this unusual symptom in what is already a rare condition.

Here, we describe an interesting case of GCA which first presented as ptosis and diplopia.

CASE REPORT

A 67-year-old Caucasian male presented to the emergency department with progressively worsening headache that started 5 weeks prior. He described his headache as constant, sharp, localized to bilateral temporal regions with radiation to the eyes, waking him up from sleep and minimally relieved with Ibuprofen use. He denied having similar symptoms in the past. Four weeks after his headache started, he noted new left eyelid drooping with tearing, redness, double and blurry vision (Fig. 1). The double vision had resolved upon presentation. In addition,

†Anoka Martis, http://orcid.org/0000-0002-1453-093X
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Figure 1: Patient 1 week prior to presentation.

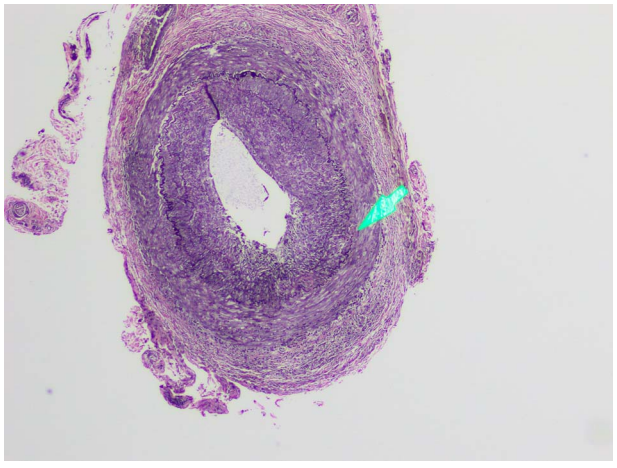

Figure 2: Elastic staining of left temporal artery showing disruption of the internal elastic lamina (green arrow).

he also described myalgias in all four extremities for ~2 weeks, limiting him from lifting his arms above his head or getting up from a squatted position. Past medical history was notable for COPD, hypertension and 80 pack-year smoking history. Physical exam was significant for trace left ptosis, conjunctival injection; visual acuity was 20/70 in the right eye and 20/50 in the left with unremarkable appearance of the optic nerves.

Laboratory studies obtained were significant for elevated C-reactive protein: 14.4 [reference range (RR): <1.0 mg/dl] and erythrocyte sedimentation rate: 54 (RR: 0–20 mm/h). Other laboratory work including thyroid stimulating hormone, creatinine kinase, vitamin D level, antinuclear antibody, rheumatoid factor and anti-citrullinated protein antibody were unremarkable. Computed tomography (CT) of the chest showed non-calcified pulmonary nodules without any concerning features. This constellation of findings was highly suspicious for temporal arteritis associated with polymyalgia rheumatica and he was immediately started on 1000 mg of intravenous Methylprednisolone given daily for 3 days. Imaging studies including CT, magnetic resonance imaging, and magnetic resonance angiogram of head and neck were noted to be unremarkable. Bilateral temporal artery biopsy was obtained, which showed panarteritis, predominantly lymphocytes admixed with few histiocytes and occasional eosinophils. Giant cells were not seen. Elastic stain highlighted the disruption of internal elastic lamina (Figs 2 and 3).

Patient reported improvement in symptoms after 2 days of steroids and he was discharged on a Prednisone taper. Two weeks after discharge, he reported feeling much better with only very mild headache and no visual symptoms at all (Fig. 4).

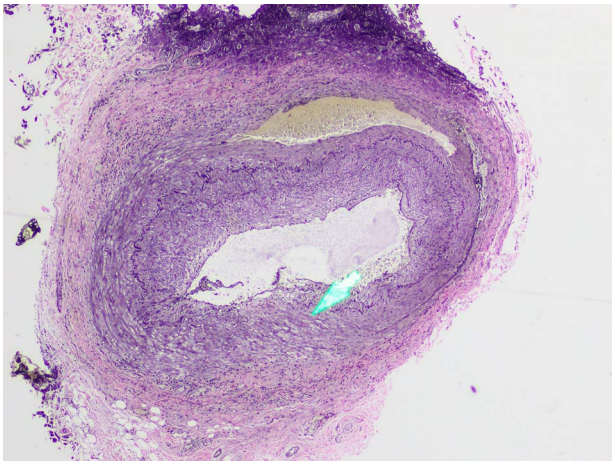

Figure 3: Elastic stain showing disruption of internal elastic lamina of right temporal artery (green arrow).

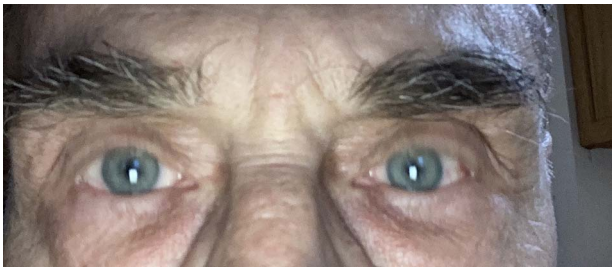

Figure 4: Patient 2 weeks after initiation of treatment.

Criteria	Description
Age at disease onset	≥ 50 years
New headache	New localized pain
Temporal artery abnormality	Tenderness to palpation or decreased palpation unrelated to atherosclerosis
Elevated ESR	≥ 50
Abnormal artery biopsy	Biopsy specimen with artery showing vasculitis characterized by a predominance of mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells

Figure 5: ACR criteria for GCA.

DISCUSSION

GCA is a granulomatous vasculitis that affects medium-to-large vessels. GCA more commonly affects females and those over the age of 50 [1]. Its effects have been known to be devastating, with permanent visual loss being one of the known complications. Early diagnosis and treatment have been shown to significantly reduce the incidence of irreversible blindness in these patients [1].

ACR has well-known criteria for the diagnosis of GCA (Fig. 5) [4] since 1990. Three out of five are considered to be positive with a high sensitivity and specificity. Currently, these criteria are undergoing possible revision with the inclusion of imaging validations. Temporal artery biopsy is considered the gold standard; however, it has a sensitivity of only 15–40% [4]. About 25% of biopsies do not show granulomas [3] as demonstrated in our patient above.

The pathogenesis of GCA remains poorly understood. One of the mechanisms appears to be the innate pathway in which there is recognition of host cells as foreign by dendritic cells, with activation of the inflammatory cascade, the key mediators being CD4+, IFN-y and IL-6 [1]. The other important mechanism is the antigen-driven response, by which vessels are affected—the arterial walls are compromised, resulting in focal ischemic changes [1].

As noted above, visual disturbances are important diagnostic symptoms, including visual loss, either partial or complete; amaurosis fugax; double vision and ocular pain. Of these, diplopia occurs rarely and is present in ~5–10% of GCA patients [5], though this is likely underreported due to its transient and retrospective nature, as it was for our patient. Diplopia is associated with higher incidence of permanent visual loss [6].

Ptosis in GCA remains even more rare. Though there have been some reports in the literature of Horner's syndrome in GCA [2, 7], pupil-sparing isolated third nerve palsies were only noted twice [8, 9] The pathogenesis behind ptosis or Horner's syndrome remains obscure, likely due to the rarity of this manifestation; however, evidence points to muscle ischemia as the cause behind the visual compromise. In an autopsy done of a patient with ophthalmoplegia secondary to GCA, the extraocular muscles were noted to be necrosed [2, 3], suggesting that GCA contributes to internal and external carotid artery ischemia and, subsequently, muscle ischemia and necrosis. There are no data on the effect of Horner's syndrome or isolated third nerve palsies on the prognosis of visual loss in GCA. This remains to be further studied.

Corticosteroids remain the first line of treatment in GCA [1]. In patients with neurological or ophthalmological manifestations, the equivalent of Prednisone at 1–2 mg/kg/day for 3–5 days is initiated after which a maintenance dose is started. In patients without visual involvement, 40–60 mg/day can be given to patients. Corticosteroids are often needed for months and it is important to give concurrent PPI as well as calcium and vitamin D supplementation.

Methotrexate has an important role in patients who are steroid-reliant by reducing the dosage required [1]. Recently, Tocilizumab (TCZ), an anti-IL-6 receptor humanized monoclonal antibody, has been used as a steroid-sparing drug with excellent remission rates [1].

GCA is a time-sensitive disease—early diagnosis and treatment are crucial in preventing vision loss. Interestingly, ptosis and diplopia happen to be very rare ophthalmic symptoms of GCA. Hence, patients presenting with isolated third nerve palsies should be carefully assessed for high-risk vasculitis such as GCA.

CONFLICT OF INTEREST STATEMENT

None declared.

FUNDING

None.

ETHICAL APPROVAL

Not applicable.

CONSENT

Written consent was obtained from the patient.

GUARANTOR

Anoka Martis.

REFERENCES

1. Baig IF, Pascoe AR, Kini A, Lee AG. Giant cell arteritis: early diagnosis is key. *Eye Brain* 2019;**11**:1–12 (Published on 17 January 2019). doi: 10.2147/EB.S170388.
2. Arunagiri G, Santhi S, Harrington T. Horner syndrome and ipsilateral abduction deficit attributed to giant cell arteritis. *J Neuroophthalmol* 2006;**26**:231–2. doi: 10.1097/01.wno.0000235562.42894.33.
3. Walters B, Lazic D, Ahmed A, Yiin G. Lessons of the month 4: giant cell arteritis with normal inflammatory markers and isolated oculomotor nerve palsy. *Clin Med (Lond)* 2020;**20**:224–6. doi: 10.7861/clinmed.2019-0504.
4. Sait MR, Lepore M, Kwasnicki R, Allington J. The 2016 revised ACR criteria for diagnosis of giant cell arteritis – our case series: can this avoid unnecessary temporal artery biopsies? *Int J Surg Open* 2017;**9**:19–23. doi: 10.1016/j.ijso.2017.09.003.
5. González-Gay MA, Blanco R, Rodríguez-Valverde V *et al*. Permanent visual loss and cerebrovascular accidents in giant cell arteritis: predictors and response to treatment. *Arthritis Rheum* 1998;**41**:1497–504. doi: 10.1002/1529-0131(199808)41:8<1497::AID-ART22>3.0.CO;2-Z.
6. Hayreh SS, Podhajsky PA, Zimmerman B. Ocular manifestations of giant cell arteritis. *Am J Ophthalmol* 1998;**125**:509–20. doi: 10.1016/s0002-9394(99)80192-5.
7. Shah A, Paul-Oddoye A, Madill S *et al*. Horner's syndrome associated with giant cell arteritis. *Eye* 2007;**21**:130–1. doi: 10.1038/sj.eye.6702454.
8. Lazaridis C, Torabi A, Cannon S. Bilateral third nerve palsy and temporal arteritis. *Arch Neurol* 2005;**62**:1766–8. doi: 10.1001/archneur.62.11.1766.
9. Mahr A, Saba M, Kambouchner M, *et al*. Temporal artery biopsy for diagnosing giant cell arteritis: the longer, the better? *Ann Rheum Dis* 2006;**65**:826–8. doi: 10.1136/ard.2005.042770.