Title
Diplopia and Ptosis in an Older Woman

Journal
JAMA Neurology, 79(9)

ISSN
2168-6149

Authors
Stallworth, Jeannette Y
Greene, J Palmer
Rasool, Nailyn

Publication Date
2022-09-01

DOI
10.1001/jamaneurol.2022.1982

Peer reviewed

JAMA Neurology Clinical Challenge

Diplopia and Ptosis in an Older Woman

Jeannette Y. Stallworth, MD; J. Palmer Greene, MD; Nailyn Rasool, MD

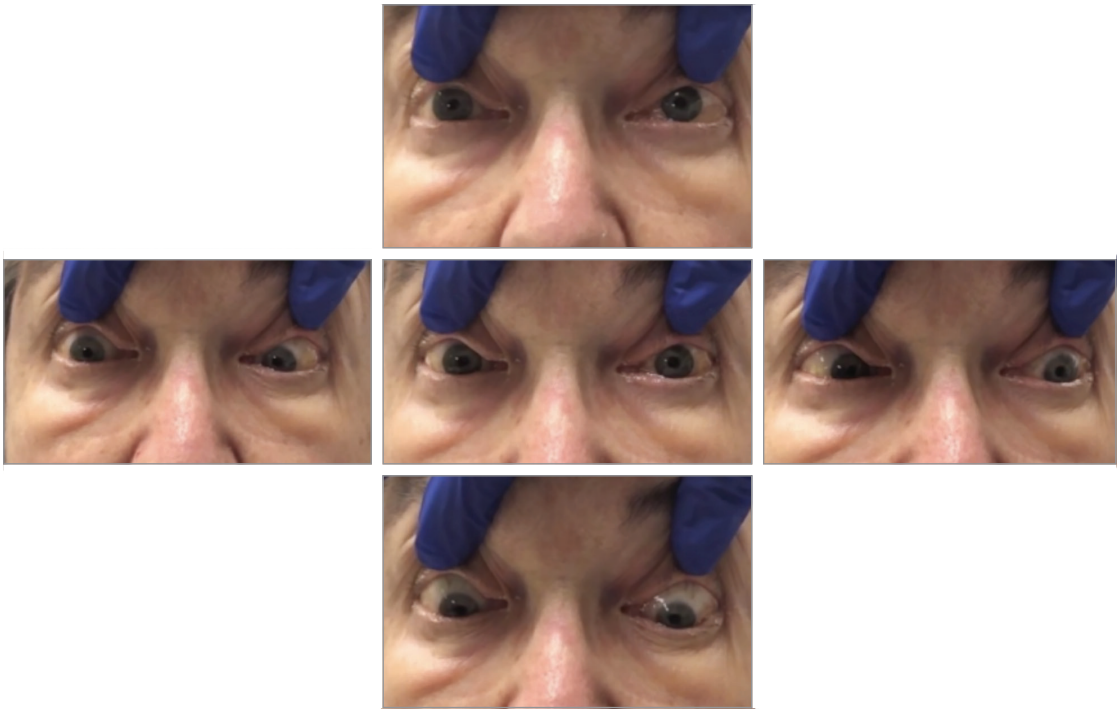

Figure 1. Extraocular movements on presentation. Examination reveals deficits in elevation and abduction of both eyes not conforming to a cranial nerve distribution.

A 75-year-old woman presented with horizontal binocular diplopia, right-sided ptosis, and a new headache that was progressive over 3 days. She reported difficulty opening her jaw, pain when chewing, and a 2.3-kg weight loss. The week prior, she experienced left-sided ptosis that persisted for 2 days and subsequently resolved. She denied vision changes, eye pain, scalp tenderness, and myalgias. Past medical history included schizophrenia, hypothyroidism, and supraventricular tachycardia.

Her examination demonstrated normal visual acuity in both eyes. Pupils were equal, round, and reactive. There was right upper-eyelid ptosis. Extraocular motility demonstrated limitation of elevation and abduction bilaterally (**Figure 1**). Results of dilated fundus examination were normal, and the remainder of her neurologic examination results were normal. Her eye movements and ptosis did not change with fatiguability, rest, or an ice-pack test. Investigations revealed a C-reactive protein level of 292 mg/L (normal, <10 mg/L; to convert to milligrams per deciliter, divide by 10) and erythrocyte sedimentation rate of 93 mm/h (normal, <30 mm/h). A lumbar puncture demonstrated normal opening pressure, cell count, and glucose and protein levels. Magnetic resonance imaging with contrast of the brain demonstrated diffuse edema and enhancement of the scalp, skull base, neck, and paraspinal soft tissues, as well as T2 hyperintensity and enhancement of the extraocular muscles bilaterally. An additional test was obtained, and a diagnosis was made.

WHAT IS YOUR DIAGNOSIS?

A. Giant cell arteritis

B. Anti–transfer RNA synthetase myositis

C. Immunoglobulin G4–related disease

D. Myasthenia gravis

Quiz at jamacmelookup.com

Diagnosis

A. Giant cell arteritis

Discussion

Facial and extraocular muscles are typically spared by the myositis associated with aminoacyl transfer RNA synthetase antibodies, and the lack of other features of antisynthetase syndrome (interstitial

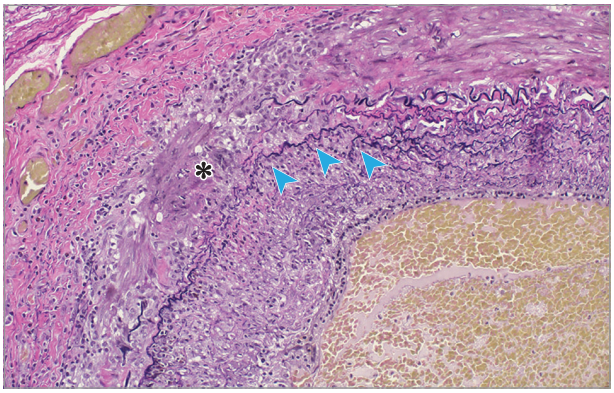

Figure 2. Histopathologic photograph of a temporal artery biopsy specimen. High-power image (original magnification ×200) shows granulomatous inflammation with giant cells (asterisk), transmural inflammation, intimal hyperplasia, and fragmentation of the internal elastic lamina (arrowheads) with elastic Verhoeff–Van Gieson stain.

lung disease, nonerosive arthritis, Raynaud phenomenon) makes this diagnosis less likely. Immunoglobulin G4–related disease can cause enlargement of the extraocular muscles, creating a restrictive pattern of motility but may also be accompanied by hypertrophic pachymeningitis and sclerosing lesions of the abdomen and lungs. Lastly, although the history of transient contralateral ptosis may be reminiscent of myasthenia gravis, the lack of fatiguability, normal ice-pack test results, and soft tissue changes on magnetic resonance imaging suggest an alternate etiology.

Temporal artery biopsy was obtained demonstrating intimal hyperplasia and transmural inflammation with giant cells (**Figure 2**), consistent with giant cell arteritis (GCA). GCA is classically thought to affect large and medium-sized arteries, although recent definitions reflect the capacity of GCA to affect vessels of any size.[1] Most patients with ocular GCA present with acute vision loss, with 80% attributable to arteritic anterior ischemic optic neuropathy owing to ischemia of the posterior ciliary arteries.[2]

Case series of GCA have demonstrated that although approximately 5% to 15% of patients report diplopia, only a subset have demonstrable oculomotor deficits on presentation, suggesting that ophthalmoplegia may be transient and underreported.[2-4] There are 2 proposed pathophysiologic mechanisms for ophthalmoplegia in GCA. The first is arteritic involvement of the vasa nervorum of the ocular motor nerves. However, ocular motility deficits in GCA often do not conform to the pattern of a cranial nerve. The second is arteritis of the vascular supply of the extraocular muscles. Despite the rich anastomotic vasculature from both the ophthalmic artery and branches of the external carotid artery, ischemia of the extraocular muscles has been observed. A histopathologic survey of the entire ocular motor apparatus in a case of GCA with severe ophthalmoparesis demonstrated extraocular muscle ischemia with preservation of cranial nerves 3, 4, and 6 along their entire course from the nerve nuclei to the orbits.[5]

Ptosis associated with ophthalmoplegia in GCA may be attributable to similar mechanisms.[4] Ptosis has also been reported in relation to Horner syndrome secondary to vasculitis affecting the vasa nervorum supplying the sympathetic plexus or direct granulomatous involvement of the sympathetic nerve fibers as they run alongside the inflamed vessel wall of the cavernous internal carotid artery.[6,7]

This patient had ocular motility deficits and ptosis that did not conform to the pattern of the ocular motor nerves and had no other signs of Horner syndrome. There was no ataxia or hemiparesis on examination, and magnetic resonance imaging ruled out a brainstem infarct. Her presentation was most suggestive of vasculitis leading to ischemia of the periorbital musculature. The patient promptly received high-dose intravenous steroids, which led to symptomatic improvement within hours.

This case demonstrates the multifaceted presentation of GCA and the importance of maintaining a high suspicion in patients with elevated inflammatory markers, headache, and uncommon neuro-ophthalmic symptoms. When ocular motility abnormalities do not correspond to a typical ocular motor nerve palsy, ischemic myopathy should be considered, and prompt treatment with corticosteroids should be initiated to prevent vision loss and complications from GCA.

ARTICLE INFORMATION

Author Affiliations: Department of Ophthalmology, University of California, San Francisco, San Francisco (Stallworth, Rasool); Department of Neurology, University of California, San Francisco, San Francisco (Greene).

Corresponding Author: Jeannette Y. Stallworth, MD, Department of Ophthalmology, University of California, San Francisco, 490 Illinois St, 5th Floor, Room 5X8, San Francisco, CA 94143 (jeannette.stallworth@ucsf.edu).

Published Online: July 25, 2022. doi:10.1001/jamaneurol.2022.1982

Conflict of Interest Disclosures: None reported.

Additional Contributions: We thank the patient for granting permission to publish this information.

REFERENCES

1. Jennette JC, Falk RJ, Bacon PA, et al. 2012 Revised international Chapel Hill consensus conference nomenclature of vasculitides. *Arthritis Rheum*. 2013;65(1):1-11. doi:10.1002/art.37715

2. De Smit E, O'Sullivan E, Mackey DA, Hewitt AW. Giant cell arteritis: ophthalmic manifestations of a systemic disease. *Graefes Arch Clin Exp Ophthalmol*. 2016;254(12):2291-2306. doi:10.1007/s00417-016-3434-7

3. González-Gay MA, García-Porrúa C, Llorca J, et al. Visual manifestations of giant cell arteritis—trends and clinical spectrum in 161 patients. *Medicine (Baltimore)*. 2000;79(5):283-292. doi:10.1097/00005792-200009000-00001

4. Dimant J, Grob D, Brunner NG. Ophthalmoplegia, ptosis, and miosis in temporal arteritis. *Neurology*. 1980;30(10):1054-1058. doi:10.1212/WNL.30.10.1054

5. Barricks ME, Traviesa DB, Glaser JS, Levy IS. Ophthalmoplegia in cranial arteritis. *Brain*. 1977;100(2):209-221. doi:10.1093/brain/100.2.209

6. Bromfield EB, Slakter JS. Horner syndrome in temporal arteritis. *Arch Neurol*. 1988;45(6):604. doi:10.1001/archneur.1988.00520300018011

7. Arunagiri G, Santhi S, Harrington T. Horner syndrome and ipsilateral abduction deficit attributed to giant cell arteritis. *J Neuroophthalmol*. 2006;26(3):231-232. doi:10.1097/01.wno.0000235562.42894.33