

Short communication

Mydriatic pupil in giant cell arteritis

Sashank Prasad^{a,*}, Jennifer Baccon^b, Steven L. Galetta^a^a Division of Neuro-ophthalmology, Department of Neurology, Hospital of the University of Pennsylvania, United States^b Neuropathology Division, Department of Pathology, Hospital of the University of Pennsylvania, United States

ARTICLE INFO

Article history:

Received 9 January 2009

Received in revised form 18 March 2009

Accepted 15 April 2009

Available online 8 May 2009

Keywords:

Mydriasis

Tonic pupil

Ciliary ganglion

Giant cell arteritis

Temporal arteritis

ABSTRACT

A mydriatic pupil has been infrequently reported as a manifestation of giant cell arteritis. We report a patient with acute, evolving pupil dilation who was diagnosed with biopsy-proven giant cell arteritis. We document the time course for the development of pupillary near-light dissociation and denervation hypersensitivity. We discuss the possible mechanisms leading to mydriasis, including 1) parasympathetic dysfunction due to ischemia of the ciliary ganglion and post-ganglionic parasympathetic fibers and 2) direct iris ischemia. Repeated episodes of pupil dilation in this patient suggested ongoing microvascular insufficiency.

© 2009 Elsevier B.V. All rights reserved.

1. Introduction

Anisocoria which accompanies giant cell arteritis (GCA) is usually due to pupil-involving third nerve ischemia [1]. We report the rare occurrence of GCA-related anisocoria without motility deficits, presumably due to microvascular ischemia of either the ciliary ganglion and post-ganglionic parasympathetic fibers or the iris sphincter itself [2–7]. Multiple recurrences of the patient's dilated pupil suggested ongoing microvascular insufficiency.

2. Case report

An 84-year-old man presented after a five minute episode of transient right monocular blurred vision. He had a history of diabetes, hyperlipidemia, dermatomyositis, and polymyalgia rheumatica, for which he took low-dose oral prednisone. He reported one month of mild headaches with temporal throbbing. He had episodes of jaw claudication, but denied myalgias, fevers, or fatigue.

Examination revealed right pupil dilatation, which was not present on recent photographs. The right pupil was 5 mm in dark, reacting to 4.5 mm in light and at near (Fig. 1, panel A). The left pupil was 4 mm in dark, reacting to 3 mm in light and at near. Slit lamp examination revealed segmental paralysis of the right pupil and loss of pupillary ruff. Thirty minutes after instilling 0.125% pilocarpine there was no change in the size of the right pupil (examined in darkness). Visual

acuity and fields were full. The optic discs were flat, without pallor. No choroidal ischemia was noted, although fluorescein angiography was not performed. Ocular motility was full. There was no ptosis or evidence of aberrant regeneration of the third nerve. Applanation ocular tension was 16 mm Hg OD and 15 mm Hg OS. The remainder of the neurological examination was normal.

The Westergren erythrocyte sedimentation rate (ESR) was 85 mm/h, which had been 35 mm/h two months earlier. The C-reactive protein was 7.8 mg/dL. Brain MRI was normal.

Empiric treatment with high-dose intravenous steroids was initiated. Bilateral temporal artery biopsy revealed giant cell arteritis (Fig. 1, panel B). He was placed on oral prednisone 60 mg daily.

Ten days after the initial presentation, the patient developed isolated, acute increased right pupil dilation (Fig. 1, panel A). The right pupil was 7 mm in dark, reacting sluggishly to 6.5 mm in light and near. The left pupil was 4 mm in dark, reacting briskly to 3 mm in light and near. With dilute pilocarpine, the dilated right pupil slightly constricted to 6 mm in darkness. Again visual acuity, fields, disc appearance, ocular motility, and lid position were normal. The change in the pupil exam raised concern for ongoing vascular inflammation and ischemia. Despite an improved ESR (27 mm/h) and CRP (0.6 mg/dL), the dose of oral prednisone was raised to 80 mg daily. Shortly thereafter, he experienced a brief episode of amaurosis in his left eye.

One month after the initial presentation, the patient demonstrated constriction of the larger right pupil to dilute pilocarpine, with reversal of anisocoria. In addition, the left pupil had become slightly larger with a small segment of poor reaction temporally. Slit lamp examination did not reveal iris transillumination. On a weaning dose of prednisone, he continued to have rare transient episodes of right pupil dilation upon awakening.

* Corresponding author. Department of Neurology, Hospital of the University of Pennsylvania, 3W Gates Bldg, 3400 Spruce St., Philadelphia, PA 19104, United States. Fax: +1 215 349 5165.

E-mail address: sashank.prasad@uphs.upenn.edu (S. Prasad).

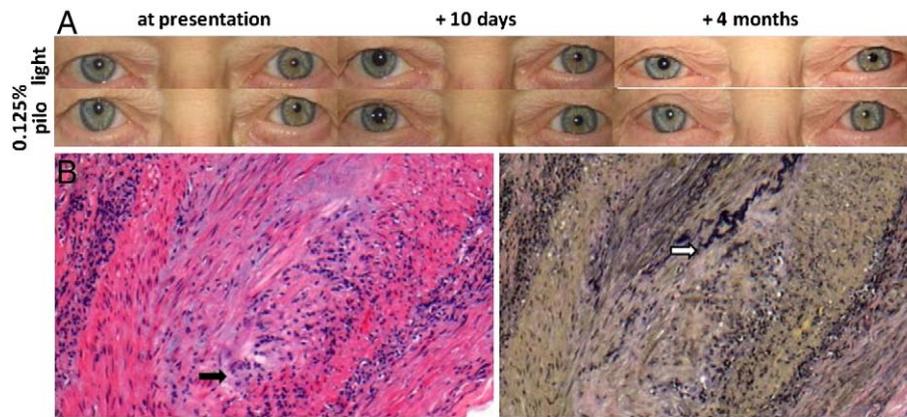


Fig. 1. Evolution of tonic pupil due to giant cell arteritis. Panel A: pupil examination in light and after 0.125% pilocarpine at presentation, after 10 days, and after 4 months. Pilocarpine was instilled in the right eye only at presentation and after 10 days; it was instilled in both eyes at 4 months. Panel B: left, marked inflammation of the vessel wall with granuloma formation (black arrow, hematoxylin and eosin stain, 10x); right, disruption of internal elastic lamina (white arrow, elastin stain, 10x).

Four months after presentation, there was even greater response to dilute pilocarpine (Fig. 1, panel A). In light, the pupils were 4.5 mm OD and 4 mm OS. With near effort, each pupil constricted by approximately 0.5 mm. After instilling a drop of dilute pilocarpine in each eye, the pupils both constricted to 1.5 mm.

3. Discussion

The pupil dilation that occurred in this case of biopsy-proven GCA was likely due to parasympathetic dysfunction from ciliary ganglion ischemia or less likely direct iris ischemia. Isolated pupil dilation, as occurs with a tonic pupil, is usually a benign condition; in this case, however, the patient's advanced age and his associated transient blurred vision, headaches, and jaw claudication raised concern for a systemic illness. Pupil dilation in GCA is commonly due to preganglionic third nerve involvement, but that mechanism was unlikely in this case given the absence of other signs of third nerve dysfunction. Ocular ischemia could cause a dilated, nonreactive pupil, but would be associated with conjunctival injection and hypotony, which were absent (1). Furthermore, choroidal ischemia was not observed (although fluorescein angiography was not performed). Direct iris ischemia was one potential explanation for this patient's mydriasis, and could account for the absent response to dilute pilocarpine within the first ten days [7]. However, the absence of iris transillumination defects makes this explanation less likely. Furthermore, an ischemic iris would not be expected to show the near-light dissociation that was observed by four months.

We considered the most likely cause of pupil dilation to be disrupted parasympathetic innervation from isolated ciliary ganglion ischemia. The initial pupil responses to near and to dilute pilocarpine were minimal, suggesting that denervation hypersensitivity and aberrant regeneration had not yet occurred. The pupil dilation that occurred 10 days later was likely due to ongoing or recurrent ischemia to the ciliary ganglion. By 1 month, denervation hypersensitivity had occurred in the right pupil, as evidenced by the response to dilute pilocarpine testing.

Supersensitivity testing of the iris sphincter to dilute pilocarpine is often difficult to interpret, and testing both eyes at the onset may have provided additional information by allowing a direct control for comparison. Even in normal individuals, the iris sphincter has been shown to react to dilute pilocarpine in select cases, presumably from

substantive drug penetration across the cornea. In our patient the pupillary responses in both eyes may have been abnormal, further complicating the interpretation of pilocarpine testing, and limiting the utility of the fellow eye's responses as a negative control. Nonetheless, it may have been valuable to follow dilute pilocarpine testing with concentrated pilocarpine testing to further evaluate the possibility of direct iris injury.

Tonic pupil is a rare complication of GCA, presumably because the anastomotic blood supply to the ciliary ganglion reduces its vulnerability to ischemia [8]. The pathophysiology of GCA is penetrating immune-mediated inflammation of the arterial wall, leading to destruction of elastic lamina and intimal layer hyperplasia with luminal occlusion. This occurs predominantly in large and medium caliber extracranial vessels, presumably following antigen recognition by adventitial T cells [9]. The normal ciliary ganglion blood supply has been shown to arise from up to four arteries (the posterior lateral ciliary and lateral muscular arteries, followed by the ophthalmic and central retinal arteries); these contain elastic lamina and are therefore susceptible in GCA [8,10].

Our patient highlights the importance of recognizing a mydriatic pupil as an early manifestation of GCA.

References

- [1] Hayreh SS, Podhajsky PA, Zimmerman B. Ocular manifestations of giant cell arteritis. *Am J Ophthalmol* Apr 1998;125(4):509–20.
- [2] Davis RH, Daroff RB, Hoyt WF. Tonic pupil after temporal arteritis. *Lancet* Apr 13 1968;1(7546):822.
- [3] Currie J, Lessell S. Tonic pupil with giant cell arteritis. *Br J Ophthalmol* Feb 1984;68(2):135–8.
- [4] Foroozan R, Buono LM, Savino PJ, Sergott RC. Tonic pupils from giant cell arteritis. *Br J Ophthalmol* Apr 2003;87(4):510–2.
- [5] Wilhelm H. Tonic pupil caused by ischemia. *Fortschr Ophthalmol* 1989;86(4):380–2.
- [6] Coppeto JR, Greco T. Mydriasis in giant-cell arteritis. *J Clin Neuroophthalmol* Dec 1989;9(4):267–9.
- [7] McKillop E, Tejwani D, Weir C, Jay J. Anterior segment ischaemia with giant cell arteritis. *Can J Ophthalmol* Apr 2006;41(2):201–3.
- [8] Eliskova M. Blood vessels of the ciliary ganglion in man. *Br J Ophthalmol* Oct 1973;57(10):766–72.
- [9] Weyand CM, Goronzy JJ. Pathogenic mechanisms in giant cell arteritis. *Cleve Clin J Med* 2002;69(Suppl 2):S128–32.
- [10] Aristodemou P, Stanford M. Therapy insight: the recognition and treatment of retinal manifestations of systemic vasculitis. *Nat Clin Pract Rheumatol* Aug 2006;2(8):443–51.