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Amaurosis Fugax, Anterior Ischemic Optic Neuropathy and Cilioretinal Artery Occlusion Secondary to Giant Cell Arteritis

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Permanent visual loss due to giant cell arteritis (GCA) may be prevented by early diagnosis and treatment. Transient visual loss in one eye (amaurosis fugax) may be the first sign of GCA [1]. We report the rare case of a patient who presented with the combination of anterior ischemic optic neuropathy and cilioretinal artery occlusion resulting in permanent visual loss in her left eye. The patient reported having had several episodes of transient visual loss prior to that. Temporal artery biopsy confirmed the diagnosis of GCA.

PATIENT DESCRIPTION

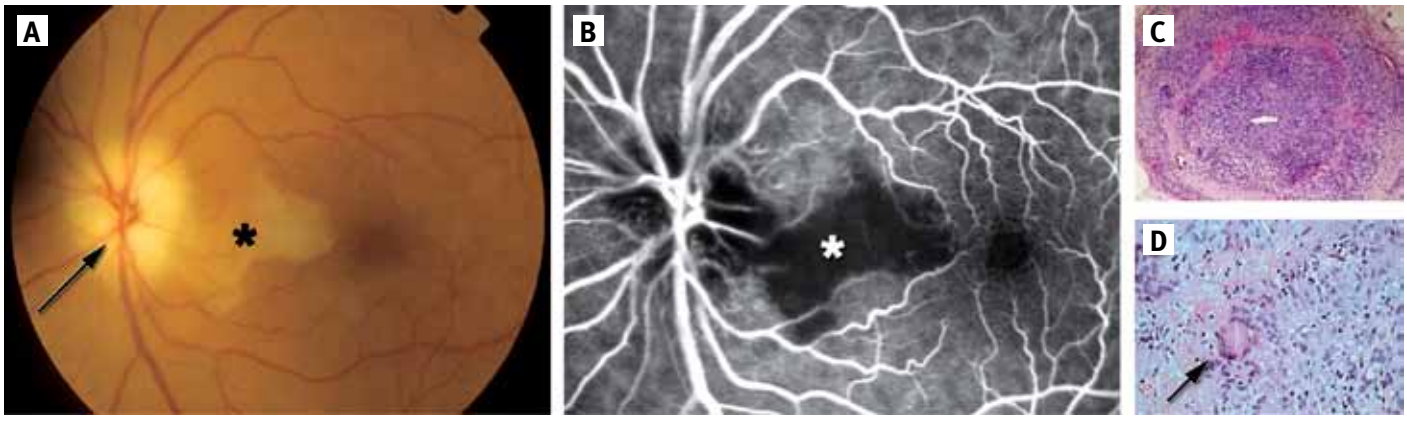
A 70 year old woman presented with sudden visual loss in her left eye. Visual acuity was no light perception and there was a 3+ relative afferent pupillary defect in the left eye. Funduscopy revealed a pale and edematous optic disk [Figure A, arrow] and ischemia of the papillomacular bundle area [Figure A, asterisk], which was also noticed on fluorescein angiography [Figure B, asterisk]. She reported having experienced several episodes of transient loss of vision in her left eye during the previous few weeks which resolved spontaneously. According to the patient she was examined by an ophthalmologist who concluded her eye examination as normal. Review of systems revealed that she had had temporal headaches and jaw claudication for a few weeks. Left scalp tenderness with nodular and pulseless temporal artery was noted. She did not show weight loss or systemic fever.

Laboratory tests revealed erythrocyte sedimentation rate of 40 mm/hr and C-reactive protein level of 42 mg/L. Complete blood count including platelets was within normal limits. Treatment with 60 mg (1 mg/kg/day) oral prednisone was immediately started.

An emergent biopsy of her left temporal artery demonstrated a thickened-wall artery with an inflammatory response that included giant cells [Figures C and D, arrow]. This confirmed the diagnosis of arteritic anterior ischemic optic neuropathy combined with cilioretinal artery occlusion secondary to GCA. In the following weeks the left optic nerve and retinal edema slowly resolved but the optic nerve was pale and visual acuity was no light perception.

Follow-up was continued in conjunction with a rheumatologist and during the next few months the corticosteroid dosage was gradually tapered by 10 mg every month. Six months after her initial presentation

[A] Edematous optic disk (arrow) and ischemia of the papillomacular bundle area (asterisk). **[B]** Fluorescein angiography shows papillomacular bundle ischemia (asterisk). **[C]** Thickened-wall temporal artery with an inflammatory response. **[D]** Giant cell (arrow)



the patient was on 10 mg prednisone. The corticosteroids were then tapered down by 2.5 mg decrements each year. The patient was left on a prednisone maintenance dose of 2.5 mg. Additional ocular or systemic manifestations of GCA did not occur during 4 years of follow-up.

COMMENT

Prompt diagnosis of GCA is critical to prevent further vision loss in the affected eye or any involvement of the second eye. This can be achieved by early treatment with systemic corticosteroids, given intravenously or orally. GCA occurs in 2.2 per 10,000 patient-years and a general practitioner may expect to see one new case every 1–2 years [2]. The reported incidence of visual loss and ocular involvement in giant cell arteritis varies greatly, between 6% and 70% [3].

The most common ophthalmic complication is anterior ischemic optic neuropathy followed by central retinal artery occlusion, cilioretinal artery occlusion, and posterior ischemic optic neuropathy [3]. Other ocular symptoms are diplopia and non-specific pain. Amaurosis fugax, which is believed to be due to optic nerve head ischemia, is a presenting symptom in 7–36% of cases [4]. It often precedes permanent visual loss and may be the key to early diagnosis and prevention in many cases [1]. The differential diagnosis of ameurosis fugax or painless transient monocular visual loss includes ocular pathology such as tear film abnormality or transient elevation of intraocular pressure, optic nerve disorders such as compressive lesion or demyelinating disease, vascular disease such as carotid or cardiac emboli, migraine or vasculitis and non-organic visual loss.

The episodes of transient loss of vision for several weeks should have alerted the patient's local ophthalmologist to the possibility of GCA. An earlier treatment with systemic corticosteroids might have prevented the additional ischemic events resulting in no light perception in the affected eye. The combination of anterior ischemic optic neuropathy and cilioretinal artery occlusion is a rare clinical manifestation of giant cell arteritis. It results from concomitant thrombotic occlusion of the short posterior ciliary arteries and the cilioretinal artery. The recurrent episodes of amaurosis fugax in the affected eye constitute a rare case of three ocular manifestations of GCA in one patient.

There is universal agreement among clinicians that corticosteroids are the mainstay of treatment for GCA and should be initiated immediately, even if the diagnosis is not yet confirmed [5]. However, there has been controversy about whether intravenous or oral corticosteroid therapy is required to treat acute visual loss in GCA. Some physicians claim that intravenous corticosteroid therapy is no more effective than oral therapy in preventing visual deterioration in GCA [5]. Others will always recommend intravenous corticosteroids (methylprednisolone at a dose of 500–1000 mg per day for 3 days) followed by 1 mg/kg/day of oral prednisone and admission of patients with acute visual loss and presumed GCA [5]. Hayreh and Biousse [5] advised oral prednisone treatment, except in the following three situations: i) a history of amaurosis fugax but no visual loss (amaurosis fugax is an ominous sign of impending visual loss), ii) complete or marked loss of vision in one eye, and iii) early signs of involvement of the second eye. In these conditions, he suggested one dose of intravenous corticosteroid (as outpatients), followed by the oral corticosteroid regimen, starting with 80 mg prednisone daily [5]. In the current case, as the patient presented 2 days after she had a complete vision loss (no light perception), she was treated with oral steroids only, in order to prevent involvement of the second eye. Other acceptable approaches would have been starting a single intravenous steroid dose followed by oral treatment or intravenous methylprednisolone for 3 days followed by oral prednisone. There is no consensus on the optimal way to taper the corticosteroid; however, it should be done slowly over a period of at least 1 year. Patients may stop treatment by 2 years but some will continue treatment further on.

GCA in the setting of amaurosis fugax can be easily missed by both ophthalmologists and general practitioners. In their recent article, Hassan et al. [2] reviewed the symptoms and signs of GCA, including jaw claudication, temporal headache, diplopia, abnormality on palpation of the temporal artery, elevated erythrocyte sedimentation rate, and anemia. This article was criticized because amaurosis fugax was not mentioned as one of the symptoms [1]. GCA should always be sought in a patient presenting with amaurosis fugax, as delay in recognition may lead to irreversible loss of vision. The occurrence of amaurosis fugax, anterior ischemic optic neuropathy and cilioretinal artery occlusion in one patient is a rare complication of GCA.

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"War will exist until that distant day when the conscientious objector enjoys the same reputation and prestige that the warrior does today"

John F. Kennedy (1917-1963), 35th U.S. president