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SHORT COMMUNICATION

Case report

Bilateral glaucomatous optic neuropathy in Takayasu's disease without cervical arterial stenosis

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PURPOSE. *Although significant decrease in retinal perfusion is usually not observed before all of the cervical arteries became markedly narrowed in patients with Takayasu's disease (TD), we present bilateral glaucomatous optic neuropathy in a patient with TD without any cervical arterial stenosis.*

METHODS. *Ophthalmoscopic examination disclosed glaucomatous optic neuropathy in both eyes with 7/10-cup/disc ratio in the right eye and 9/10 in the left eye. Left subclavian selective arteriographic examination demonstrated segmental high-grade stenosis, namely 90 percent stenosis in the mid portion of the left subclavian artery. Arteriography, digital subtraction angiography (DSA), magnetic resonance angiography (MRA) and color Doppler sonography revealed patent cervical, carotid interna, ophthalmic, retinal and posterior ciliary arteries.*

RESULTS. *Patient was followed up for 48 months with frequent intervals and there was no deterioration of visual acuity, visual field and optic neuropathy without any antiglaucomatous treatment.*

CONCLUSIONS. *Although it is a known fact that classical ophthalmic manifestations of the TD occur only when major cervical arteries are occluded, no occlusion was observed in this patient with bilateral optic atrophy. The optic nerve damage is caused by various factors, but these factors require much elucidation before the optic neuropathy can be understood. (Eur J Ophthalmol 2001; 11: 93-6)*

KEY WORDS. *Takayasu's disease, Glaucomatous optic neuropathy, Low tension glaucoma*

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INTRODUCTION

Takayasu's disease (TD) is a chronic inflammatory arteriopathy of unknown origin that leads to stenotic changes in the arterial lumen (1). Reduced perfusion of the eye leads to well-known ophthalmic manifestations, which occur late in the course of the disease (1-5). A significant decrease in retinal perfusion does not occur until all of the cervical arteries become markedly narrowed in patients with TD (6). We report a unique case of bilateral glaucomatous optic neuropathy in TD without cervical arterial stenosis.

Case report

A 29-year-old male presented to SSK Istanbul Hospital Ophthalmology Department in January 1994 with a three-month history of blurred vision in his left eye. He had been seen in the intensive care unit with a complaint of fever, weight loss, ventricular arrhythmia and unconsciousness for further investigation 15 months before. He had responded well to the antiarrhythmic treatment, but no diagnosis could be established at that time. He had been complaining of left arm claudication for one year.

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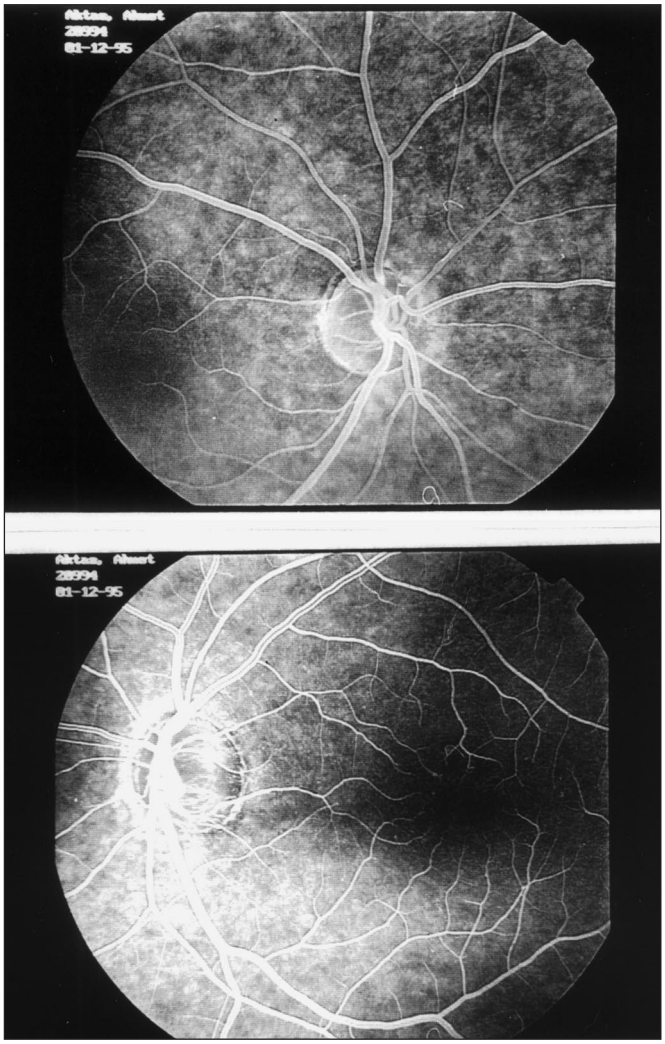

Fig. 1 - *Fluorescein angiography shows normal retinal circulation of the right and left eye.*

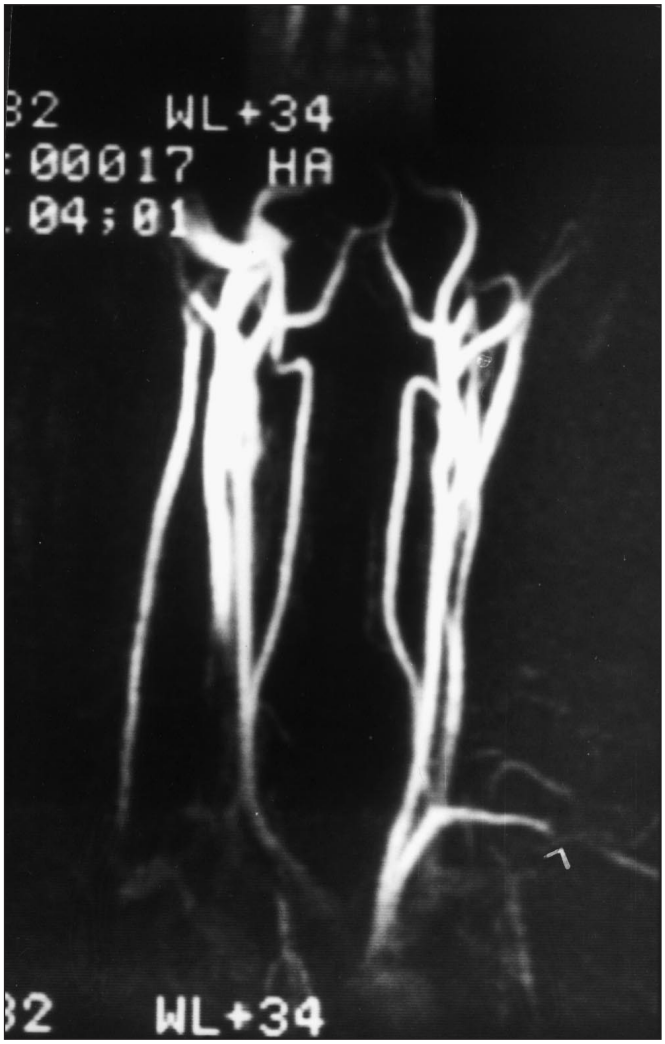

Fig. 2 - *MR angiography shows a severe stenotic subclavian artery segment, 2 cm long, distal to the vertebral artery origin (white arrow).*

Ophthalmic examination: Visual acuity was 10/10 in the right and 7/10 in the left eye. There was a left-sided afferent pupillary defect and ophthalmoscopic examination disclosed glaucomatous optic neuropathy in both eyes. There was a 7/10 cup/disc ratio in the right eye and 9/10 in the left eye. There was baring of the lamina cribrosa and diffuse thinning of the disc rim in both eyes. Intraocular pressure (IOP) was 10 mm Hg bilaterally. Diurnal variations in IOP did not exceed 15 mmHg. Retinal arterial pressure was 125/65 mmHg in both eyes. The fundus fluorescein angiography was completely normal in both eyes and arm-to-retina circulation time was within the normal limits (Fig. 1). ICG videoangiography showed no abnormality in the choroideal circulation. Visual field examination revealed double arcuate scotoma in both eyes and the left eye was in the end stage with central and temporal islands.

Physical examination: Blood pressure at the right and left arms was 150/110 and 120/100 mmHg. There was a moderate systolic murmur in the left supraclavicular area. The left radial and brachial artery pulses were decreased. Bilateral carotid and other artery pulses were normal. Lungs were clear. Cardiac examination was normal. There was no abdominal bruit or murmur. The skin was normal. Behçet's disease was eluded.

Laboratory findings: CBC, urinalysis, renal and liver function tests, total cholesterol and triglyceride levels were normal. ESR was 30 mm/h (Westergren method); anticardiolipin antibody, antinuclear antibody, antineutrophil cytoplasmic antibody, rheumatoid factor were negative. ECG was normal. HLA B-5 and HLA B-51 were negative. Toxoplasma, rubella, CMV, HSV Ig M were negative (TORCH). TPHA and VDRL were also negative.

Radiologic findings: Lung radiograph was normal. Left subclavian selective arteriographic examination showed segmental high-grade stenosis, 90 percent diameter stenosis in the mid-portion of the left subclavian artery, distal to the vertebral artery branch, with extensive collateral flash. Aortoperipheral angiographic examination showed a normal right subclavian artery, thoracic, abdominal aorta and peripheral branches.

DSA showed patent ophthalmic arteries bilaterally. There was neither stenosis nor obstruction of the carotid arteries. The aortorenal DSA examination revealed normal main renal arteries and intrarenal branches.

On MRA examination of the cervical region, CCA, carotid bulb, and extracranial ICA were normal on both sides. On the left side, however, the examination revealed a severely stenotic subclavian artery segment 2 cm long, distal to the vertebral artery origin (Fig. 2). Intracranial arterial structures did not present any abnormality on cranial MRA examination. Intracranial and intraorbital MR images were normal.

The color-Doppler examination of the bilateral ophthalmic, central retinal and short posterior ciliary arteries gave blood flow velocities within normal ranges and anterograde flow in the ophthalmic arteries, bilaterally. Peak systolic velocity in the right and left ophthalmic artery measured 0.34 m/sec and 0.30 m/sec, respectively, and 0.10 m/sec in both posterior ciliary arteries.

The patient was initially treated with prednisone 1 mg/kg body weight. Treatment was maintained at this level for two weeks, then the dosage was tapered to 0.1 mg/kg gradually. He underwent percutanous transluminal angioplasty for the left arm claudication. There was initial improvement in his symptoms within two months, but the claudication recurred.

The patient was followed up for 48 months at frequent intervals and there was no deterioration of visual acuity, visual field and optic neuropathy, without any antiglaucomatous treatment.

DISCUSSION

A significant decrease in retinal arterial pressure does not occur until all the cervical arteries become markedly narrowed in patients with TD (4-6). These lesions do however, lead to a poor prognosis for life-span and/or the eyes, regardless of treatment. The chronic nature of the disease produces progressive dilatation of the retinal capillaries leading to the formation of fusiform or saccular microaneurysms (5). Exudate and hemorrhage are rare in pulseless disease, since the intraluminal pressure in these dilated vessels is very low. Progressive retinal hypoxia causes arteriovenous communications. These begin to form in the peripheral retina and then progress to surround the disc in a wholrlike arrangement as detailed by Takayasu (3). Late ophthalmological complications of TD can include vitreous hemorrhage, secondary cataract, rubeosis iridis with neovascular glaucoma and anterior segment ischemia which can become fulminant to the point of mydriasis, anterior uveitis, orbital pain, profound loss of vision, hypotony and phthisis bulbi (6-11).

In this patient, various radiological exainations detected left subclavian stenosis. Cervical arteries and intracranial vessels that could affect ophthalmic blood supply were not occluded. Bilateral optic neuropathy did not occur due to major cervical arterial stenosis in this patient.

In order to explain this unusual clinical entity we could suggest two different mechanisms. Firstly, during the initial period of disease the patient had a very severe inflammatory phase with fever, cardiac arrhythmia and unconsciousness. Transient perfusion deficiency due to circulation disturbance may led to non-progressive glaucomatous optic neuropathy. This theory correlates well with the non-progression of optic atrophy without any antiglaucomatous medication. As a second possibility, in individuals with an inclination to autoimmunity, immunopathogens may inflict damage on the optic nerve or its vessels. Cartwright and co-workers observed an association between glaucomatous damage and many immune-related diseases (12). An autoimmune disorder is often proposed in TD (12-15). The favourable response to corticosteroid therapy in most patients supports an autoimmune etiology.

In this patient, the optic nerve damage was caused by various factors, but they all await clarification before the optic neuropathy can be undestood (16-18).

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