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CASE REPORT

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A case of polyarteritis nodosa complicated by left central retinal artery occlusion, ischemic optic neuropathy, and retinal vasculitis

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Abstract A 23-year-old single female patient developed constitutional manifestations in the form of fever, weight loss, anorexia, malaise, fatigue, and generalized aches in January 1995, 2 weeks after an attack of German measles. This was followed by painful, reddish, macular skin lesions over both legs which healed by dark pigmentation (leucocytoclastic vasculitis), mononeuritis multiplex, and Raynaud's phenomena of both hands and feet. Angiography of lower limbs was done to visualize the arterial tree of both lower limbs and revealed typical beading of distal arterial branches, a diagnosis compatible with polyarteritis nodosa (PAN). At that time, the patient received prednisone (45 mg/day) and azatioprin (100 mg/day) and responded well to treatment. In a second presentation in June 2005, the patient developed sudden attack of loss of vision in her left eye. Ophthalmological examination of the patient revealed evidence of left central retinal artery occlusion, ischemic optic neuropathy. The patient received methyl prednisolone, 1 g IV infusion, daily infusion for three consecutive days followed by oral prednisolone, 30 mg/day. The patient received pulse cyclophosphamide IV infusion (0.6 g/m^2) on the fourth day. One week after receiving therapy, the patient progressed from having light perception to counting of fingers from a distance of 1 m.

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Keywords Central retinal artery occlusion · Polyarteritis nodosa

Polyarteritis nodosa (PAN) is a disease of small- and medium-sized arteries. Although any organ can be affected, PAN most commonly involves the skin, joints, peripheral nerves, gastrointestinal (GI) tract, and kidneys. Untreated polyarteritis nodosa has a very poor prognosis. Treatment with steroids increases the 5-year survival to 48%, whereas addition of cytotoxic immunosuppressive treatment improves the outcome dramatically [1]. Central retinal artery occlusion is not a well-recognized complication of polyarteritis nodosa.

Case report

A 23-year-old single female patient developed fever, weight loss, anorexia, and generalized aches in January 1995, 2 weeks after an attack of German measles. This was followed by painful, reddish, macular skin lesions over both legs which healed by dark pigmentation (leucocytoclastic vasculitis), mononeuritis multiplex, and Raynaud's phenomena of both hands and feet. Angiography of lower limbs revealed typical beading of distal arterial branches, a diagnosis compatible with polyarteritis nodosa. At that time, the patient received prednisone, 45 mg/day and was advised to receive pulse cyclophosphamide therapy on a monthly basis, but the patient refused at that time and received immuran (100 mg/day), instead. She responded well to treatment.

A second presentation in June 2005, after 4 months of stopping her treatment, the patient developed sudden attack of loss of vision in her left eye with no preceding attacks of amaurosis fugax. Ophthalmological examination of the patient revealed evidence of left central retinal artery occlusion, ischemic optic neuropathy, and beading of arterial retinal branches (see Fig. 1). On examination, the patient showed normal vital signs and peripheral pulsations were equally felt in both lower limbs, with no abdominal

tenderness or rigidity. Her laboratory results showed erythrocyte sedimentation rate (ESR) of 5 mm first hour, C-reactive protein (CRP), 1.5 mg/dl, normal CBC, liver and kidney function tests normal, urine analysis was free, C3–C4 showed normal levels, antinuclear antibody (ANA)-DNA were negative, c-ANCA and p-ANCA were negative, and hepatitis B surface antigen (HBsAg), hepatitis C virus (HCV) were negative.

Abdominal and brain computed tomography (CT) angiography was done for the patient to visualize the abdominal and brain vessels and to assess the disease activity and its extent as well. CT angiography for the abdomen revealed beading of small terminal branches of the superior mesenteric vessels (see Fig. 2). CT angiography of the brain revealed attenuation of the medium and small brain vessels (see Fig. 3). The patient received methyl prednisolone, 1 g IV infusion daily for three consecutive days followed by oral prednisolone, 30 mg/day. The patient received pulse cyclophosphamide IV infusion (0.6 g/m^2) on the fourth day. One week after receiving therapy, the patient progressed from having light perception to counting of fingers from a distance of 1 m.

Discussion Up to our knowledge, there are few case reports in the literature about central retinal artery occlusion in patients suffering from PAN. Central retinal artery occlusion in our case, in the absence of disease activity elsewhere and in the absence of elevated ESR and CRP levels is interesting.

Pathophysiology The central retinal artery is the first intraorbital branch of the ophthalmic artery, which enters the optic nerve 8–15 mm behind the globe to supply the retina. Short posterior ciliary arteries branch distally from the ophthalmic artery and supply the choroid. Anatomical variants include cilioretinal branches from the short posterior ciliary artery, which gives additional supply to the macula from the choroidal circulation. A cilioretinal artery occurs in approximately 14% of the population.

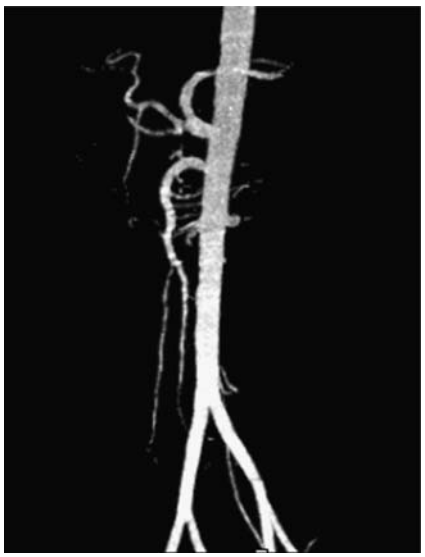

Fig. 2 Beading of the superior mesenteric arterial branches

Approximately 14% of the general population has cilioretinal arteries and 25% of eyes with acute central retinal artery occlusion (CRAO) have cilioretinal artery involvement. The cilioretinal artery supplies part or all of papillomacular bundle. In 10% of eyes, the cilioretinal artery supplies some or all of the foveola. In such an eye, the visual acuity generally returns to 20/50 or better in 80% of eyes over a 2-week period.

Solomon and Solomon [2] reported a case of bilateral central retinal artery occlusion associated with polyarteritis nodosa. Akova et al. [3] reviewed five cases of polyarteritis nodosa with a spectrum of ocular findings which preceded and contributed to its diagnosis. The observed ophthalmic manifestations of polyarteritis nodosa in these patients included scleritis, peripheral ulcerative keratitis, nongranulomatous uveitis, retinal vasculitis, pseudotumor of the orbit, and central retinal artery occlusion. Hsu et al. [4] reported a 70-year-old woman who developed hand and

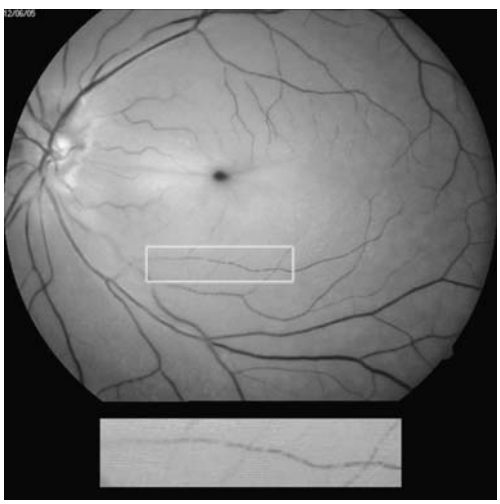

Fig. 1 Optic disc edema, cherry red spot, and beading of the arterial branches

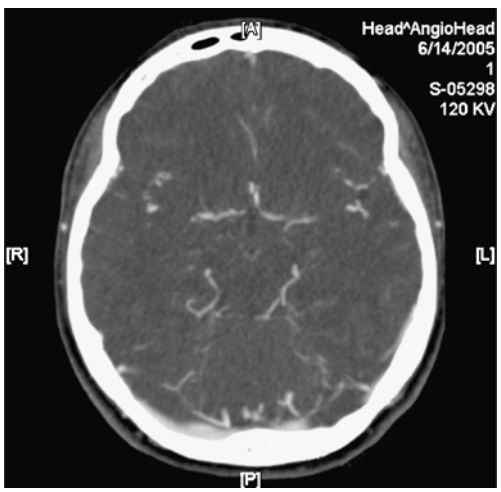

Fig. 3 Attenuation of the middle cerebral arteries with subtle intimal irregularities

foot numbness followed by intermittent blurred vision and binocular horizontal diplopia. Two weeks later, she suddenly lost vision in the right eye from a central retinal artery occlusion and then developed a left anterior ischemic optic neuropathy and bilateral triangular choroidal abnormalities consistent with infarction. They concluded that polyarteritis nodosa can produce ischemia of a variety of ocular structures, including the retina, choroid, and optic nerve. Schmidt et al. [5] reported unusual ocular findings in three patients suffering from PAN with acute visual disturbance:

Case 1.
A 40-year-old man initially presented with papilledema together with partial optic atrophy in the eyes, later polyneuropathy, gangrene of the toes, and myalgic pains. Under treatment with cyclophosphamide and prednisone, no relapse occurred during a follow-up of 2 years.

Case 2.
In a 67-year-old man who suffered from arterial hypertension and coronary heart disease, central retinal artery occlusion occurred, at first in the left and then later in the right eye. During a follow-up of 4 years, including treatment with prednisone and cyclophosphamide, no relapse occurred.

Case 3.
A 16-year-old adolescent, diagnosed with PAN, presented with Raynaud's disease, cachexia, arterial hypertension, and visual disturbance due to inflammation of the choroidal vessels of the right eye.

The authors concluded in their report that PAN should be considered in the differential diagnosis in patients with acute inflammatory signs of the optic nerve head, the choroid, and/or the retina together with the general signs of the disease.

Conclusion

Polyarteritis nodosa should be considered in the differential diagnosis of patients with acute inflammatory signs of the optic nerve head, central retinal artery occlusion, and/or retinal vasculitis together with the general symptoms and signs of the disease. Presentation of this case is intended to emphasize the fact that ocular inflammation should be considered in patients suffering from polyarteritis nodosa and early institution of aggressive therapy for central retinal artery occlusion in patients with PAN is mandatory in spite of normal ESR and CRP levels.

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