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Sir,

Retinal Vascular Abnormalities in Aortic Coarctation

In the elderly population central retinal vein occlusion (CRVO) is often associated with systemic conditions such as arteriosclerosis, hypertension, diabetes mellitus, hyperviscosity and hypercoagulability states, and connective tissue disorders.[1] The prevalence of hypertension in patients with CRVO is reported as approximately 60%, which is twice as common as for an age-matched population.[1–5] CRVO is uncommon in the younger age group but has important associations including head injury, use of oestrogen-containing compounds, connective tissue disorders, hyperlipidaemia, hyperviscosity and cryofibrinogenaemia.[1,6,7] In young adults, hypertension is a less frequent association of retinal vein occlusion and is often secondary to renal disease, endocrine disorders, drugs, toxaemia of pregnancy and collagen disorders. Coarctation of the aorta is an uncommon cause of hypertension in adults. It is frequently accompanied by retinal vascular abnormalities,[8] which together with chronic hypertension can contribute to increased risk of retinal vascular occlusion.

A case of a previously healthy 23-year-old man with bilateral retinal vascular tortuosity, left CRVO and hypertension secondary to aortic coarctation is reported.

Case Report

A 23-year-old male office worker presented to the emergency eye clinic with a 3 day history of painless loss of vision in the left eye. He had been healthy in the past and had suffered no significant systemic or eye conditions. Corrected visual acuity was 6/6 right eye and 6/60 left eye. Anterior segment examination was unremarkable and the intraocular pressures were 17 mmHg in each eye. The right retinal vessels were tortuous (Fig. 1) and there was a left CRVO with marked oedema and superficial haemorrhages (Fig. 2). Left brachial arterial pressure was 170/100 mmHg, femoral pulses were diminished and labora-

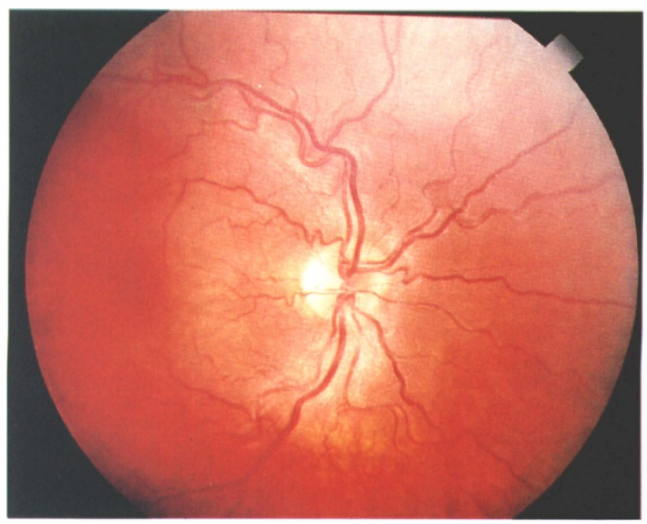

Fig. 1. *Right retinal arteriolar tortuosity associated with aortic coarctation.*

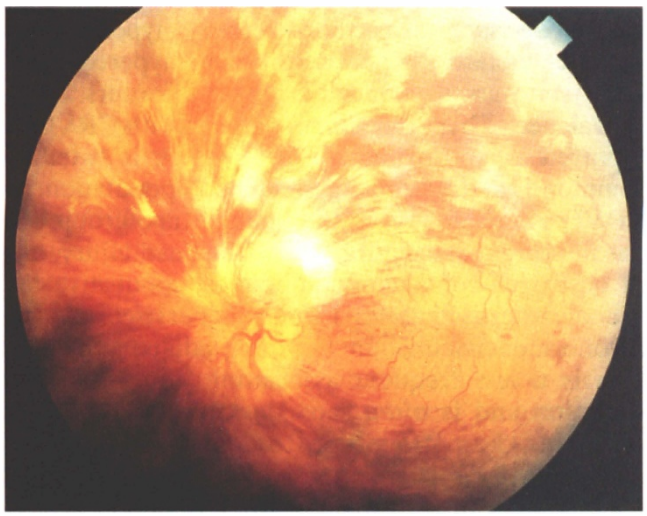

Fig. 2. *Left central retinal vein occlusion and retinal arteriolar tortuosity.*

tory investigations including full blood count, erythrocyte sedimentation rate, lipids, clotting studies and serum proteins were normal.

Chest radiography demonstrated rib notching, and aortic angiography confirmed the diagnosis of coarctation of the aorta distal to the origin of the left subclavian artery (Fig. 3). The patient was referred for early surgical correction of the coarctation. Four years following successful surgery the visual acuity was 6/6 in the right eye and 6/60 in the left; the left fundus had a central macular scar with areas of pigment atrophy and no evidence of neovascularisation. The retinal arteriolar tortuosity remained unchanged in both eyes (Fig. 4).

Discussion

Aortic coarctation is an uncommon cause of hyper-

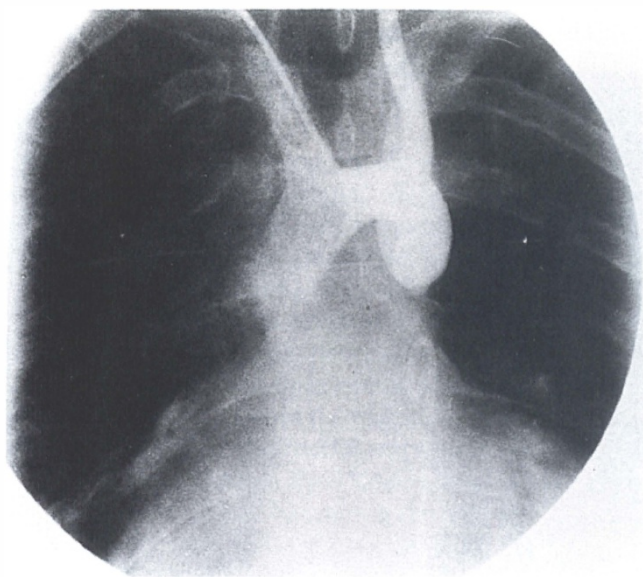

Fig. 3. *Aortic angiogram demonstrating coarctation distal to the origin of the left subclavian artery.*

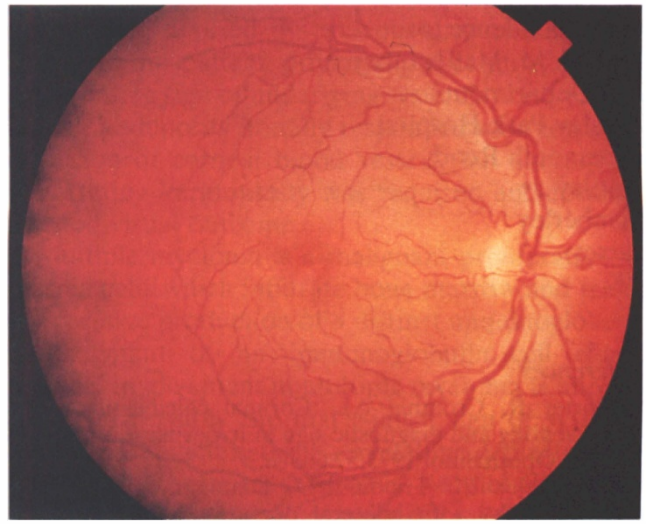

Fig. 4. *Retinal arteriolar tortuosity persisted 4 years after successful surgical correction of the aortic coarctation and a normal blood pressure.*

tension in adults. It is a congenital maldevelopment of the aorta that occurs in 1 in 2000 of the population. The narrowing of the aortic lumen may occur at isolated or multiple sites with the commonest site of obstruction being just distal to the origin of the left subclavian artery. Upper body hypertension is associated with symptoms such as recurrent epistaxis and throbbing headaches, while leg fatigue, claudications and cold feet result from poor blood perfusion. Adults may present with infective endocarditis, rupture of an aortic aneurysm or cerebrovascular accident. Early surgical correction is indicated before secondary and irreversible pulmonary and cardiovascular damage develops. If the condition is untreated, death occurs by the age of 40 years.[9]

Ophthalmological manifestations of aortic coarctation are not well described and include those of hypertension such as arteriovenous crossing abnormalities and altered vascular light reflex. More pronounced changes such as exudates, haemorrhages, oedema and cotton wool spots are rare. Granstrom[8] reported corkscrew tortuosity of the retinal arteries in 50% of his series of patients with aortic coarctation, which he believed to be characteristic of the condition. Others have also reported tortuosity, focal or generalised narrowing and pulsation of retinal arteries.[10–13] Other developmental anomalies such as iris and choroidal coloboma, corneoscleral dermoid and scleral staphyloma have also been reported in young children with various forms of neurocutaneous syndromes involving coarctation of the aorta.[14–16]

This case is of interest for two reasons. First, the occurrence of CRVO in aortic coarctation has not, to the author's knowledge, been reported previously. Second, persistence of retinal arteriolar tortuosity following normalisation of blood pressure by surgical correction is documented.

In the case reported, thrombosis of the central retinal vein may have been related to the vascular abnormality, or to the effects of chronic hypertension or both. Alternatively it may have been a coincidental event. However, CRVO is uncommon in young adults, and when it does occur it is commonly mild with good visual recovery. This makes this coincidence an unlikely event. In this patient, associated arterial abnormality may have contributed to the severity of vascular decompensation.

Pathophysiological factors that contribute to central retinal vein thrombosis include inflammatory or degenerative disease of the venous wall, compression by an adjacent sclerotic artery, haemodynamic alterations predisposing to stagnation and clotting abnormalities. Arteriosclerosis and hypertension can affect any of these factors, and indeed they are the most common systemic associations of CRVO. Vascular developmental anomalies can also promote thrombosis further. Walters and Spalton,[17] in a study of 17 young patients with CRVO, suggested that inflammation of the venous wall is an unlikely cause and that an alternative explanation for the development of CRVO in young patients is a congenital anomaly of the central retinal vein.

In conclusion, a rare case of CRVO and retinal arteriolar tortuosity associated with coarctation of the aorta is reported. The vascular tortuosity persisted 4 years after successful treatment.

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Sir,

Pseudohypopyon due to Synchisis Scintillans

Synchisis scintillans (cholesterolosis bulbi) is a rare ocular condition in which cholesterol crystals form in the vitreous, usually following a traumatic vitreous haemorrhage. The clinical appearance is of multiple crystalline flakes, golden or coloured when illuminated, in a fluid vitreous. Most of the crystals settle in the lower vitreous, to be briefly resuspended when the eye moves. If the integrity of the lens or zonule is disrupted, vitreous with synchisis scintillans may enter the anterior chamber and present a striking appearance. In the case documented here, many of the crystals settled to the bottom of the anterior chamber, giving the appearance of a pseudohypopyon.

Case Report

A woman with Down's syndrome (trisomy 21), had a history of self-injury, in that she had habitually punched her own eyes since childhood. At age 24 years she was found to have early cataracts, but no other ocular abnormality. At age 30 years vision had deteriorated significantly, and a dense cataract was removed from the left eye. Because of posterior synechiae and calcification of the lens capsule, the procedure was technically difficult, resulting in the complete removal of the lens.

Post-operatively, the anterior chamber was found to contain vitreous, with a suspension of crystalline bodies which sparkled pink, green and gold in the slit-lamp beam (Fig. 1). Some of the crystals remained suspended, but most settled to the bottom of the anterior chamber, forming a 2 mm pseudohypopyon. With saccadic eye movements, many crystals became resuspended, and swirled for few seconds before resettling.