neurosensory retinal thickening, with deeper dark shadows consistent with calcification (Figure 1d and e). Retinal oedema in the macula was confirmed on OCT (Figure 1f). B-scan ultrasound (Figure 2d) revealed a solid dome-shaped mass with high focal reflectivity due to the calcification. The left eye was within normal limits and systemic examination was negative for tuberous sclerosis. The tumour remained stable after 1 year and we obtained a fundus photograph from 9 years previously that confirmed no change (Figure 1a).

Comment

Our final diagnosis was sporadic retinal astrocytic hamartoma with recurrent vitreous haemorrhage. Vitreous haemorrhage in retinal astrocytic hamartoma is unusual and has been reported previously only in the context of tuberous sclerosis.[2–6] Along with the case of Giles *et al*,[1] our case suggests a vascular component exists in sporadic retinal astrocytic hamartoma.

Retinal astrocytic hamartomas of tuberous sclerosis have been classified into three types by angiographic features: all three types block choroidal fluorescence to some extent and show late leakage on FA, but on ICG type 1 has subtle blockage of choroidal fluorescence, type 2 has total blockage (from calcification) and type 3 has total blockage only in the centre.[7] Though our case is sporadic, it has features of type 1 on FA and type 2 on ICG, suggesting incomplete calcification. OCT features are also consistent with retinal astrocytic hamartoma.[8]

We believe that sporadic retinal astrocytic hamartoma should now be included in the differential diagnosis of recurrent vitreous haemorrhage – sporadic cases can exhibit vasculopathic features.

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Sir,

Protein S deficiency manifesting simultaneously as central retinal artery occlusion, oculomotor nerve palsy, and systemic arterial occlusive diseases

A 28-year-old Asian gentleman presented with sudden onset of left eye ptosis and visual loss owing to central

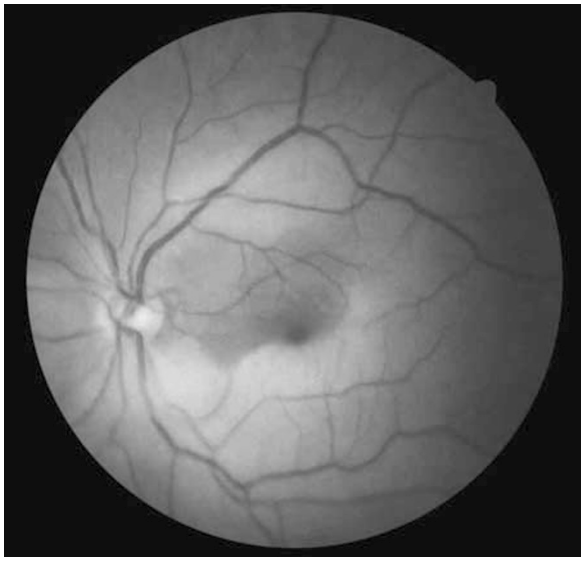

Figure 1 Left fundus photo. Central retinal artery occlusion with presence of cilioretinal circulation.

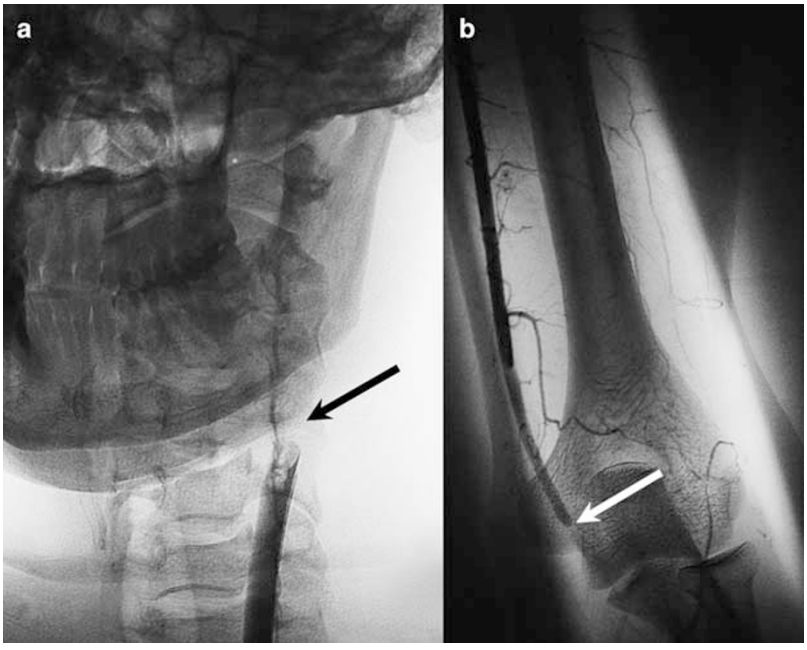

Figure 2 (a) Angiography of left common carotid artery. Occlusion of left common carotid artery as indicated by black arrow, (b) Angiography of left brachial artery. Occlusion of left brachial artery as indicated by white arrow.

retinal artery occlusion, ischaemic oculomotor nerve palsy with simultaneous systemic manifestations of left common carotid artery, and brachial artery occlusions secondary to congenital protein S deficiency syndrome.

Congenital protein S deficiency is a rare, autosomal-dominant coagulation disorder characterized by arterial or venous thrombosis in young people.[1]

We report a case of protein S deficiency with simultaneous manifestations of central retinal arterial occlusion, oculomotor nerve palsy, common carotid artery, and brachial artery occlusion.

A 28-year-old Asian gentleman presented with sudden onset of no light perception in the left eye, associated with partial ptosis. Examination revealed left oculomotor nerve palsy, central retinal artery occlusion with sparing of the cilioretinal circulation (Figure 1), and an ischaemic left hand. Angiography showed occlusions of the left common carotid artery (Figure 2a) and brachial artery (Figure 2b). Blood investigations revealed protein S deficiency. Anticoagulation with intravenous heparin was commenced followed by embolectomy of the left brachial artery. His vision improved to 20/20 with resolution of the nerve palsy and re-establishment of the circulation to the left hand within 3 days.

Protein S deficiency has been associated with systemic thrombosis involving cerebral, illiofemoral, mesenteric, and renal vessels, and ocular manifestations such as central retinal artery occlusion, branch retinal artery, retinal vein occlusion, anterior ischaemic optic neuropathy, and ischaemic abducens nerve palsy.[2–6]

To our knowledge, the simultaneous manifestations of central retinal artery occlusion, oculomotor nerve palsy, common carotid artery, and brachial artery occlusion have not been reported previously.

The acute loss of vision with ophthalmoplegia suggested a thromboembolic phenomenon affecting the arterial supply to the retina and the oculomotor nerve.

A young patient presenting with central retinal artery occlusion may have other ocular manifestations such as ischaemic oculomotor nerve palsy and systemic vascular occlusion secondary to protein S deficiency. Prompt diagnosis with institution of anticoagulation treatment as demonstrated in our case can lead to rapid resolution of symptoms with good systemic and visual outcome.

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