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Sir,

Choroidal telangiectasia in a patient with hereditary hemorrhagic telangiectasia

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Hereditary hemorrhagic telangiectasia, previous known as Rendu–Oslar–Weber disease, is an autosomal dominant inherited disease characterized by telangiectasia of skin, mucous membrane and various vascular beds in the body.[1] These vascular lesions are composed of multiple dilatation of capillaries and venules that frequently have thinned vessel walls, which makes them friable and more prone to hemorrhage. The clinical manifestations of this disease are variable according to both location of vascular lesions and severity of organ dysfunction secondary to these lesions. The most common systemic symptoms are epistaxis, gastrointestinal bleeding, and hemoptysis.[1]

Ocular involvement has been reported in patients with hereditary hemorrhagic telangiectasia. Conjunctival telangiectasia is the most common manifestation.[1] Intraocular vascular lesions, including retinal telangiectasia, arteriovenous malformation in the retina, were reported only in a few cases.[1–5] Herein we describe a patient with hereditary hemorrhagic telangiectasia who had abnormal choroidal vascular change which has never been reported in the literature, to our knowledge.

Case report

A 73-year-old male patient, a documented case of hereditary hemorrhagic telangiectasia, suffered from blurred vision of the left eye in May 1999. At that time, he was admitted to ENT ward with a chief complaint of epistaxis off and on for more than 20 years. He received CO_2 laser cauterization of multiple telangiectasia over nasal and oral mucosa. In addition, other lesions including multiple conjunctival telangiectasia, skin hemangioma and hepatic hemangioma were also present. Family history revealed a high incidence of frequent nosebleeding among the family members.

Ophthalmic examination revealed that best-corrected visual acuity was 20/40 in the right eye and 20/50 in the left eye. Slit lamp biomicroscopy disclosed several patches of telangiectasic vessel over bilateral palpebral conjunctiva. Mild nuclear sclerosis was noted in both lens. No retinal telangiectasia was found in either eye. There was a serous neurosensory detachment over the central foveal region in the left eye. A retinal pigment epithelial detachment (RPED) with early pooling of fluorescein dye was also found inferiotemporal to the fovea (Figure 1). Meanwhile two hot spots at the notch of RPED were found, which showed a mild degree of dye leakage at the late phase. Besides, two segments of prominently dilated choroidal vessels were distributed superio-temporal to the fovea. These dilated choroidal vessels were surrounded with blocked fluorescence. In the right eye, there was a window defect over the macular area. Indocyanine green (ICG) angiography demonstrated marked dilatation and tortuosity of choroidal vasculature at the early phase in both eyes. These ectactic vessels were distributed from the peripapillary area to the periphery, and appeared to be more pronounced in the left eye (Figure 2). The

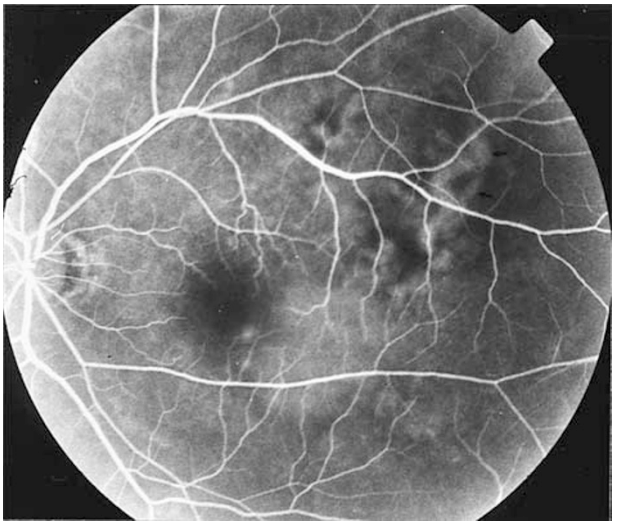

Figure 1 Fluorescein angiogram of the left eye showing an area of faint pooling, compatible with serous RPED. Note the prominent segments of choroidal vessels (arrows) beneath the macula and the superior temporal arcade.

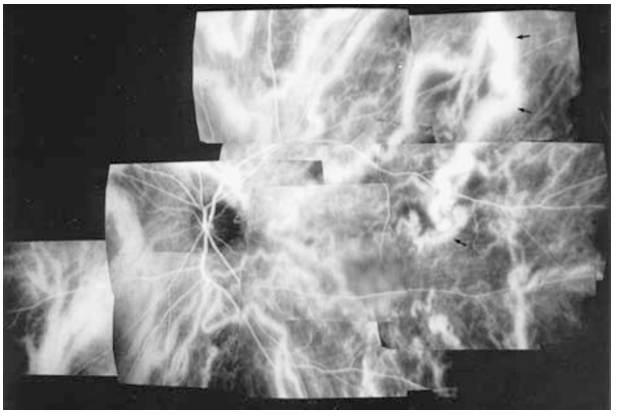

Figure 2 SLO with ICG videoangiography of the left eye demonstrated astonishing dilatation and tortuosity of choroidal vasculature (arrows) in the early phase.

diameter of those ectactic choroidal vessels was larger than one disc diameter. They seemed to drain directly to the vortex vein. The serous RPED appeared as a dome-shaped hypofluorescent area both at the early and late stage. No corresponding dye leakage of the hot spots could be found on the ICG angiography.

This serous RPED subsided spontaneously about 2 months later. Vision improved to 20/30 in the left eye. Only some RPE mottling was found in the previous lesion site of RPED in the left eye.

Comment

Ocular involvement is not uncommon in hereditary hemorrhagic telangiectasia. According to previous studies, between 45% and 65% of patients had ocular abnormalities, of which the most common lesion was conjunctival telangiectasia.[1] Intraocular involvement in hereditary hemorrhagic telangiectasia is relatively rare. It has been estimated that its incidence would be less than 1%, when based on 1500 reported cases of hereditary hemorrhagic telangiectasia.[2] Since Francois first described intraocular involvement in 1938, reports of fundus abnormalities were exclusively vascular lesions of retina or optic disc.[1–4] More specific lesions are dilated and tortuous veins and tortuous arterioles of retina.[1] Retinal telangiectasia, localized arteriovenous malformation, vitreous hemorrhage, and newly formed vessels in the retina or on the disc have also been reported.[1,3,4]

In our patient, dilatation and tortuosity of choroidal vessels was disclosed with ICG angiography. These ectactic choroidal vessels were found to exist from the peripapillary area to the periphery. They seemed to drain directly to the vortex vein in each quadrant. These ectactic choroidal vessels showed intense early hyperfluorescence with surrounding hypofluorescence. There was no leakage of dye throughout the study. Brant *et al*[1] also described a patient with a network of telangiectatic vessels in the retina that did not show any leakage in the fluorescein angiography (FAG). It seems that these intraocular vascular anomalies in hereditary hemorrhagic telangiectasia have an intact barrier function and do not leak. However, the surrounding hypofluorescence of the ectactic vessels is due to the pigmentary disturbance of the overlying retina. This pigmentary disturbance implies that the ectactic choroidal vessel induces degenerative change of RPE, and that the possibility of micro-exudation from ectactic vessels cannot be dismissed.

Decreased vision in the left eye of our patient resulted from the serous RPED. Though we found two hot spots near the RPED in the FAG, there was neither choroidal neovascular membrane nor dye pooling found in the ICG angiography. The serous RPED showed hypofluorescence surrounded by tortuous ectactic choroidal vessels at the early phase, and remained hypofluorescent till the late phase. Giovannini *et al* reported that choroidal venous dilatation could be found in 33.3% of patients with idiopathic serous RPED.[5] The dilated choroidal vein would be at the site or within an area of one disk diameter from the RPED. We followed this patient carefully and the RPED subsided spontaneously. We thought that the ectactic choroidal vessels might cause a degenerative change of both overlying Bruch's membrane and RPE. The serous RPED might result from the ectactic choroidal vessels and the degenerative change of Bruch's membrane. In conclusion, the choroidal vascular alteration represents a rare intraocular manifestation of hereditary hemorrhagic telangiectasia.

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Sir,

Retained fragments in the anterior segment following phacoemulsification surgery

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Phacoemulsification is the procedure of choice for most surgeons performing cataract surgery in the United Kingdom. The sequelae and management of retained nuclear fragments in the vitreous following cataract surgery have been well described in the literature.[1–3] However, retention of material in the anterior segment is less well documented. We report a case of a retained fragment in the anterior segment following routine phacoemulsification surgery.

Case report

An 88-year-old patient with nuclear sclerosis underwent routine phacoemulsification under local anaesthetic. The phaco priming and tuning had proceeded uneventfully. As soon as phacoemulsification commenced, a shower of small white particles was noticed in the anterior chamber. These appeared to be emanating from the lower end of the sleeve. The instrument was immediately withdrawn from the anterior chamber. The anterior segment was examined under the microscope where numerous small white particles were seen on the iris and floating in the anterior chamber. The sleeve was then removed from the phacoemusification needle and inspected under the microscope. The sleeve lining did not appear to be completely smooth and was lined with small white fragments which appeared to be floating away from the sleeve. No abnormalities of the phacoemulsification tip were noted nor of the irrigating fluid or tubing. Both the sleeve and tip were changed, the machine was reset and surgery then proceeded uneventfully.

At the time of surgery all visible particles were washed out of the anterior chamber. However on the first day postoperatively, a small white particle was seen on the surface of the iris at about the 12 o'clock position (Figure 1). The eye was otherwise quiet with a clear cornea, only the occasional cell in the anterior chamber and no flare. The patient had an unaided visual acuity of 6/9. In view of these findings it was decided to start the routine postoperative drops used by our department, ie G. Dexamethasone q.i.d. and G. Chloramphenicol q.i.d. and to observe the patient closely for any adverse effects of the retained foreign body in the anterior segment.

The patient's postoperative course was completely uneventful and her drops were tailed off and stopped after 4 weeks. Her visual acuity throughout was 6/9 unaided in the operated eye. There were no abnormal particles in the vitreous and the fundus showed no abnormality apart from retinal pigment epithelial changes at the macula which predated the surgery.

It is now one year post surgery, the white fragment is still visible in the same position on the iris and the patient has not experienced any adverse effects from its presence in her eye. The eye remains quiet with unaided visual acuity of 6/9.

Comment

There have been several reports of retained foreign bodies in the anterior chamber following cataract surgery. These have mainly concerned lens fragments[4,5] or metallic foreign bodies.[6,8]

Our case is unusual in that the source of the foreign bodies was the inner lining of the sleeve, which covered the phacoemulsification tip. On close

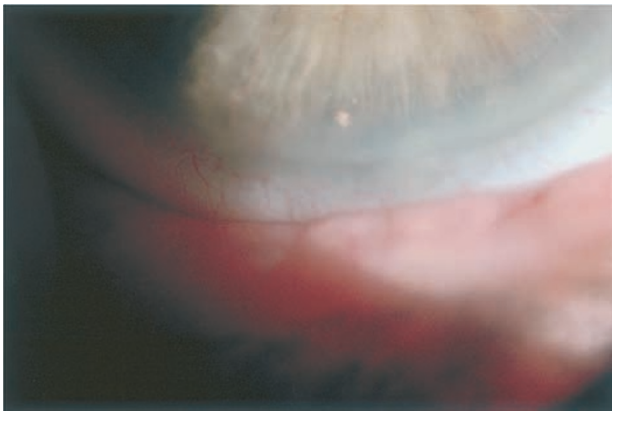

Figure 1 Small white particle on the surface of the iris.