primary germ cell tumour, as is the testis, the possibility that 'double' tumours could arise in these areas has to be considered.

Intracranial germ cell tumour associated with gonadal germ cell tumour is apparently rare, and to our knowledge only three such cases have been reported.

The remarkable similarity of the histopathological appearances of these tumours from different locations suggested that they originated from the same cell type. It is not clear whether these cells correspond to the primordial germs cells embryologically or to certain stages of differentiation of the primordial germ cells. Sano *et al.*[5] proposed that germ cell tumours may not originate from only one type of cell (primordial germ cells), except for the germinoma. He suggested that these intracranial tumours may be explained by the misinvolvement–misenfoldment hypothesis, in which the primordial germ cells during their physiological migration to the gonadal ridge may become involved in a group of laterally moving mesoderm cells and migrate to the cranial area to be enfolded into the neuraxis. Chaganti *et al.*[6] have proposed a new model for the genetic origin of all germ cell tumours in the male, based on the cytogenic analysis of carcinoma *in situ*, primary mediastinal and gonadal germ cell tumours, embryonal migration of primordial germ cells and spermatogenesis. They suggested that the increase in chromosome 12p copy numbers in germ cell tumours leads to increased expression of a gene or genes located on this chromosomal arm which, in turn, triggers neoplastic transformation. Because of the identical cytogenetics of mediastinal and gonadal germ cell tumours, they suggested that these tumours arise from dissemination of early gonadal lesions which recapitulate embryonal memory and reverse-migrate to thymus and pineal where receptive environments permit their establishment as primary tumours. These hypotheses could help to explain the occurrence of synchronous primary germ cell tumours at different sites.

We consider that it is important for the ophthalmologist or other clinician investigating a patient with an overt germ cell tumour to bear in mind the possible coexistence of a similar, perhaps symptomless neoplasm at another site. If a testicular tumour of this type is present, it would also be worthwhile considering needle biopsy of the other testis. All patients with germ cell tumours should be monitored using a panel of markers.

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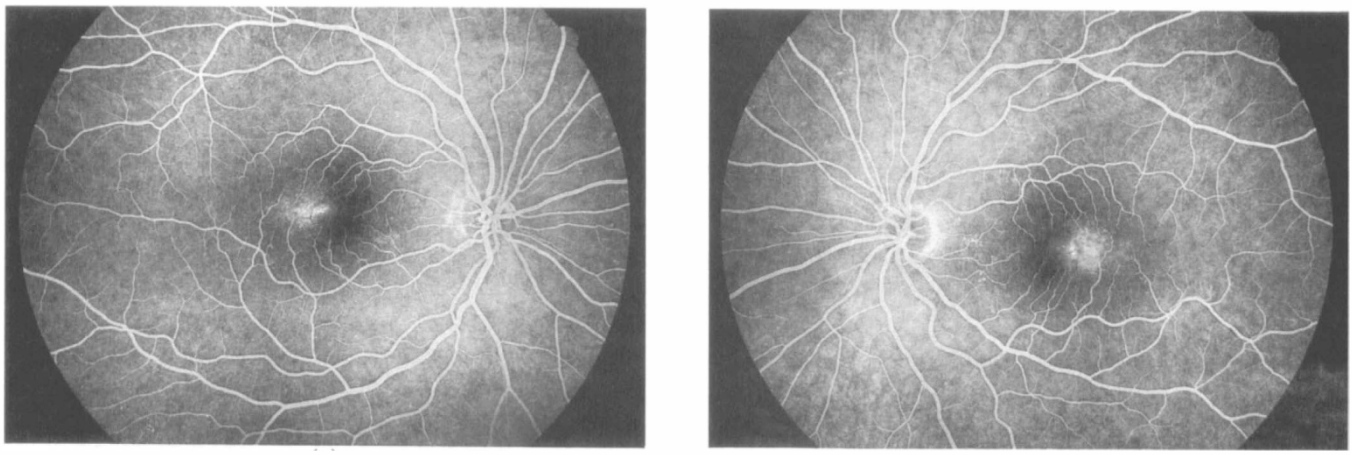
Sir,

Familial Idiopathic Juxtafoveolar Retinal Telangiectasis

Idiopathic juxtafoveolar retinal telangiectasis has been reported only rarely as a familial condition, and there are no well-documented cases in the literature of bilateral involvement in siblings. We describe the clinical and fluorescein angiographic findings over a period of 8 years in two sisters who presented with this disorder in middle age.

Case Reports

Case 1. A 49-year-old woman presented in March 1987 complaining of reduced vision in her left eye for more than 1 year. There was no past ophthalmic history and her general health was good. Visual acuity was 6/9 in each eye with correction, and slit lamp examination was unremarkable. Examination of both maculae showed subtle changes of symmetrical telangiectasia affecting capillaries at the temporal margin of the perifoveal arcade in each eye. The peripheral retinal vasculature appeared normal. Fluorescein angiography in the right eye showed dilatation of perifoveal capillaries, with early filling and then marked late staining of microaneurysms immediately temporal to the fovea (Fig. 1a). Similar



(a) (b)

Fig. 1. *Case 1. Fundus fluorescein angiograms. (a) Right eye. Venous phase, showing focal hyperfluorescence in the temporal perifoveal region due to dilatation and leakage from perifoveal capillaries, and filling followed by staining of microaneurysms (b) Left eye. Venous phase. Similar changes are present, affecting a slightly larger area than in the right eye.*

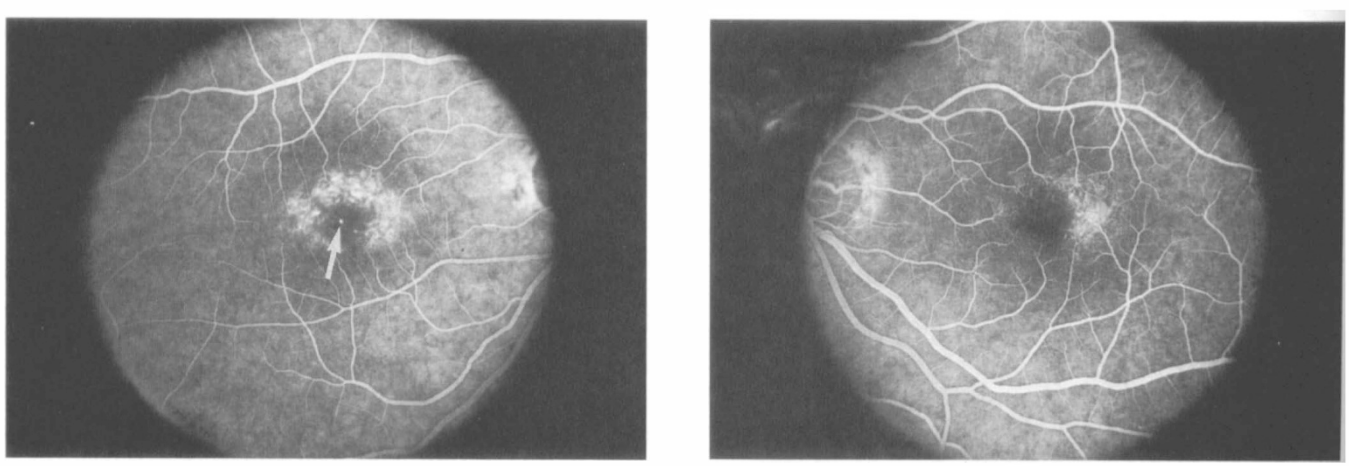
angiographic features were present in the left eye, but affecting a slightly larger area (Fig. 1b). No treatment was offered.

In August 1991 the patient was re-referred complaining of a loss of vision in her right eye for several months. Visual acuity was now 6/24 in her right eye, and 6/9 in her left. Repeated fundus fluorescein angiography revealed little change in the pattern of telangiectatic capillaries in each eye, but an increase in the extent of the transmission defects within the pigment epithelium was observed. In the later stages of the angiogram these defects showed progressive staining.

Case 2. A 48-year-old woman, sister to case 1, was referred in August 1989 complaining of progressive blurring of distance vision for 6 months. There was no significant past ophthalmic history, and she was in good general health. Corrected visual acuity was 6/15 in her right eye and 6/12 in her left. Fundoscopy showed retinal changes temporal to fixation in both eyes. The paramacular area was thickened and greyish in colour, and there was dilatation and engorgement of perifoveal capillaries. The peripheral retina appeared normal. Fluorescein angiography in the right eye demonstrated a transmission defect in the pigment epithelium (Fig. 2a), and confirmed the presence of dilated perifoveal capillaries which progressively leaked fluorescein into the intraretinal space in the later stages of the angiogram. Similar, though less extensive changes were present in the left eye (Fig. 2b).

Discussion

The pathogenesis of juxtafoveolar retinal telangiectasis in the two cases reported here is unknown. Abnormal dilatation of the parafoveal capillary



(a) (b)

Fig. 2. *Case 2. Fundus fluorescein angiograms. (a) Right eye. Late venous phase, showing multiple dilated perifoveal capillaries leaking fluorescein. A small central focus of hyperfluorescence corresponding to a retinal pigment epithelial 'window defect' is arrowed. (b) Left eye. Early venous phase showing similar, though less extensive, perifoveal capillary changes.*

network may be seen in ocular inflammatory conditions, after retinal venous occlusion, in association with diabetes, hypertension or carotid artery occlusion, or after ocular or periocular radiation.[1–3] Less commonly, telangiectasis involving vessels of the posterior pole has been described in association with generalised conditions including fascioscapulohumeral muscular dystrophy[4,5] and incontinentia pigmenti.[6] Posterior pole involvement has also been recorded in familial exudative vitreoretinopathy.[7,8] Idiopathic cases of juxtafoveolar retinal telangiectasis were described in 1968 by Gass,[9] and a classification of the disorder was subsequently proposed.[10] This has recently been updated, based on the biomicroscopic and fluorescein angiographic findings in 140 cases.[11] Three major groups of patients are now recognised. Group 1 comprises young, predominantly male patients with unilateral, easily visible parafoveal telangiectatic blood vessels and retinal exudation. Group 2 comprises patients with occult or barely visible telangiectasis, minimal exudation, superficial retinal crystalline deposits, and right-angled venules; the telangiectasis is acquired during middle age, is bilateral, and frequently precedes the development of foveolar atrophy, intraretinal pigment plaques, and subretinal neovascularisation. Group 3 comprises patients with bilateral, easily visible telangiectasis, minimal exudation, and parafoveolar capillary occlusion. Laser photocoagulation may be beneficial for some patients in group 1, but not for those in other groups, in the absence of subretinal neovascularisation.[11]

The two cases reported here show bilateral involvement, with occult telangiectasis affecting predominantly the temporal parafoveal region, transmission defects in the adjacent retinal pigment epithelium, and no exudates. The cases appear to fall into group 2 of the classification of Gass and Blodi, although two of the characteristic findings in this group, namely superficial retinal crystalline deposits and right-angle venules, are absent in our patients.

The cause of this localised form of retinal telangiectasis, affecting initially the temporal parafoveal region, remains unknown. Gass and Oyakawa[10] have suggested that the temporal parafoveal retina is affected primarily in this condition due to chronic venous congestion caused by obstruction of retinal veins as they cross retinal arterioles on either side of the horizontal raphe. Several authors have suggested that the condition resembles diabetic retinopathy.[3,12] Retinal microvascular changes, including telangiectasia of the parafoveal capillaries and parafoveal capillary non-perfusion, are well-recognised manifestations of diabetic retinopathy,[13] and Green *et al.*[14] noted the similarity between the electron microscopic appearance of the retinal capillaries in idiopathic telangiectasia and those observed in diabetic retinopathy. Millay *et al.*[15] performed glucose tolerance testing on 28 patients with parafoveal telangiectasia and normal fasting blood glucose levels, and found results consistent with diabetes in five cases. They suggest that glucose tolerance testing should be performed in all patients who have bilateral parafoveal telangiectasia.[15] Each of our patients has been followed for at least 5 years without the development of overt diabetes, and repeated measurements of fasting serum glucose and glycosylated haemoglobin to November 1995 have been normal. Formal glucose tolerance testing was declined by both patients.

The occurrence of idiopathic retinal telangiectasis in siblings has been previously reported. Hutton *et al.*[16] described two sisters with the disorder, although in one case retinal signs were unilateral. In the large series collected by Gass and Blodi,[11] three patients from group 2, comprising 92 cases, were noted to be siblings, but the clinical features of these individual cases are not described. Putteman *et al.*[17] have reported two brothers with idiopathic telangiectasia localised to the temporal aspect of the parafoveal region, although again, telangiectasia was present in only one eye in one of their patients. We have examined the offspring of one of our patients, two daughters of case 2, aged 32 and 37 years. Each is asymptomatic, visual acuity is normal in both eyes, and fundus examination and fluorescein angiography have revealed no evidence of telangiectasis or other abnormality in either patient. No other family members are available for examination.

We believe that our report is the first to describe in detail the occurrence of bilateral idiopathic retinal telangiectasis in two siblings, and may reflect a genetic predisposition toward the development of juxtafoveolar telangiectasis in some cases.

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Sir,

Cavernous Sinus Syndrome Associated with Neurocysticercosis

We present the case of a 26-year-old Brazilian man who presented with cavernous sinus syndrome associated with neurocysticercosis. The patient was treated with albendazole and prednisolone; he made a full recovery. Neurocysticercosis should be added to the differential diagnosis of cavernous sinus pathology, especially in patients from endemic areas.

Case Report

A 26-year-old Brazilian man resident in the UK for 3 years presented with a 7 day history of left retro-orbital pain and diplopia in all positions of gaze. For 4 days he had been vomiting and feeling generally unwell.

On examination visual acuities were 6/6 in the right eye and 6/9 in the left. Goldmann fields were full. Colour vision by Ishihara plates was normal. Examination of extraocular muscle movements showed left IIIrd and left VIth nerve palsies. The IVth cranial nerve on the left was intact as demonstrated by intorsion on downgaze. He had a left ptosis; pupil reactions were normal. Corneal sensation on the left was reduced. The remaining cranial nerves and peripheral nervous system were all normal.

A full blood count revealed a mild neutrophilia count of 8.6×10^9/l (normal range 1.5–7.5 $\times 10^9$/l); the ESR was not raised. A CT scan showed multiple small round calcified lesions scattered throughout the brain parenchyma. In the frontal lobes there were two large cystic lesions (Fig. 1). These findings are typical of neurocysticercosis.[1] Lumbar puncture revealed elevated cerebrospinal fluid protein (0.59 g/l; normal range 0.1–0.4 g/l). Glucose levels and cytology were normal. Serum immunoflourescent antibody titres (IFAT) and enzyme-linked immunosorbent assay (ELISA) were negative. The CSF IFAT was negative but the CSF ELISA was positive at an optical density of 0.504 (normal <0.250). MRI scans of brain and orbits showed changes consistent with neurocysticercosis; however, no lesion could be identified in the cavernous sinus.

Cavernous sinus syndrome was diagnosed on the clinical findings. Radiological and serological evidence supported a diagnosis of neurocysticercosis. The patient was commenced on albendazole 400 mg t.d.s. with prednisolone 30 mg o.d. Symptoms improved within 48 hours; he went on to make a full recovery.

Discussion

Cysticercosis may cause ocular motor disorders due to direct muscle infiltration[2] or isolated cranial nerve palsy due to raised intracranial pressure. We are

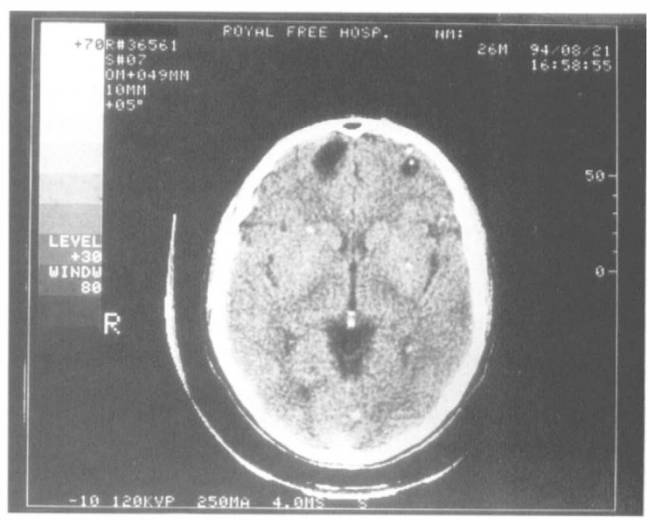

Fig. 1. *CT scan showing multiple calcified lesions scattered throughout the brain parenchyma. Note two cystic lesions in the frontal lobes.*