

pattern. Haemangiopericytoma of the orbit is composed of oval to fusiform cells surrounded by scanty clear cytoplasm, basal lamina material and the cells are orientated around branching vessels with a 'staghorn' configuration. There are no specific immunohistochemical makers for fibrous histiocytoma, haemangiopericytoma and fibrosarcoma. These diagnoses are made by exclusion of other specific diagnoses. The consistent CD34 and 013 reactivity is a helpful clue to its diagnosis. Malignant nerve sheath tumours, synovial sarcoma and leiomyosarcoma can be differentiated by their characteristic histological pattern and by immunohistochemical staining for S100, EMA and smooth muscle actin respectively.

This report adds to the evidence that SFT does occur in the orbit and is the first reported case to show locally aggressively behaviour and dedifferentiation to fibrosarcoma occurring in an SFT of the orbit. It emphasises the necessity for complete excision of these tumours.

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Sir,

#### **Eccentric disciform lesions: a marker of treponemal disease?**

Although the use of penicillin has resulted in a steady decline in both endemic and sexually transmitted treponemal disease, there has been an increase in the number of reported cases over the last two decades. Most ophthalmologists rarely encounter treponemal disease,

but patients with endemic disease may arrive in western countries and pose diagnostic difficulties. The two subspecies of *Treponema pallidum* causing venereal syphilis and yaws are morphologically identical and cannot be differentiated.

We report an unusual ocular manifestation of infection with *T. pallidum* in the form of haemorrhagic disciform retinal detachment. Although subretinal neovascularisation has been reported as a rare finding in acquired ocular syphilis, it may be the first to be reported as an initial manifestation of infection with *T. p. pertenue* (yaws).

We report on four patients, all of whom originated in a common endemic area of the West Indies.

#### Case reports

*Case 1.* A 54-year-old West Indian man presented with sudden onset of blurred vision in the right eye. On examination his visual acuity was 6/6 in each eye. Anterior chamber and vitreous were quiet. Fundoscopy showed extensive drusen in his left eye. There was an area of chorioretinal scarring with a halo of pigmentation in the supratemporal periphery of the right eye. There was a retrohyaloid haemorrhage overlying most of the inferotemporal retina which broke through into the vitreous over the next few days. As the vitreous haemorrhage cleared, it was possible to see an area of three disc diameters of haemorrhagic serous retinal detachment supratemporally adjacent to another similar yellow subretinal lesion. Fluorescein angiography showed early hyperfluorescence which increased in intensity compatible with pigment epithelial detachment. The area of serous detachment was masked by haemorrhage and subretinal neovascular membrane was not obvious.

The patient tested positive for treponemal serology and was treated with 2.4 megaunits intramuscularly of benzathine penicillin on three occasions. Vitreous haemorrhage cleared and the area of serous detachment resolved into a round area of chorioretinal scarring with an improvement in final visual acuity to 6/9 over the next few months. It was later confirmed that he was treated for a positive serological test for syphilis in 1989 with three injections of spectinomycin, despite giving a clear history of yaws in childhood and clinical evidence of old scarring on his left lateral calf consistent with the infection.

*Case 2.* A 64-year-old diabetic West Indian man presented with reduced vision in his left eye. On examination, his vision was 6/60 in the left eye and 6/9 in the right eye. Fundoscopy showed bilateral pigmented chorioretinal scars and left vitreous haemorrhage. There was an area of subretinal fibrosis adjacent to a peripheral solid haemorrhagic retinal detachment. Ultrasound showed an area of shallow solid retinal detachment corresponding to the haemorrhagic area. No definitive diagnosis was reached.



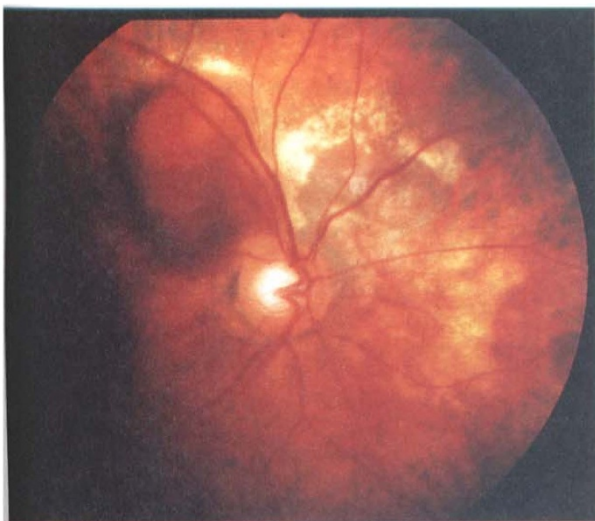
**Fig. 1.** Case 3. Left fundus view showing a haemorrhagic serous elevation.

Three years later he presented with multiple haemorrhagic elevated lesions in the peripheral retina of his right eye. Anterior chamber and vitreous were quiet. The lesions resolved into patches of generalised chorioretinal atrophy with subretinal fibrosis. His treponemal serology was positive compatible with active disease and he was treated with 2.4 megaunits of benzathine penicillin intramuscularly. His final visual acuity remained unchanged. The patient denied sexually transmitted disease or childhood infection with yaws.

*Case 3.* A 69-year-old West Indian man presented with unilateral visual loss. On examination he was found to have an extensive haemorrhagic disciform lesion in the macular area of the right eye with a vision of counting fingers. Visual acuity in the left eye was 6/18. There was an area of haemorrhagic serous elevation of the retina in the temporal periphery of his left eye (Fig. 1). There were patches of geographical chorioretinal atrophy at the periphery of both retinæ. The anterior chamber and vitreous were quiet. His treponemal serology was

positive. The patient denied any venereal contact but recalled a childhood history of yaws. There was a thin wrinkled patch on his calf consistent with yaws skin. He appeared to have poor short-term memory and it was only after a few consultations that it became clear he had been under the care of neurologists with loss of memory, epilepsy and ischaemic cerebral lesions. CT scan showed areas of translucence compatible with infarcts. He was treated with 2.4 megaunits of benzathine penicillin intramuscularly. His final visual acuity remained unchanged.

*Case 4.* A 78-year-old West Indian man presented with a 2 month history of reduced vision in the left eye. His visual acuity was 6/24 in the right eye and counting fingers in the left. He was noted to have bilateral cataracts and a subfoveal neovascular membrane on the left. He underwent an uneventful right phacoemulsification with a posterior chamber implant. At his first post-operative visit at 2 weeks, his right vision was 6/24 unaided improving to 6/12 with a pinhole. It was during this visit that he was noted to have a juxtapapillary and peripheral disciform lesion in the right eye (Fig. 2a). There was chorioretinal scarring adjacent to the optic disc nasally. He had post-operative anterior uveitis but showed no signs of vitreous activity. A fluorescein angiogram showed early leakage with increasing intensity being partly masked by overlying haemorrhage consistent with a subretinal neovascular membrane (Fig. 2b). His treponemal serology was positive and he was treated with doxycycline 100 mg b.d. for 1 month. His final visual acuity was unchanged in the left eye and improved to 6/18 in the right over the next few months. His medical history dating back 10 years included an indurated ulcer on the right calf associated with numerous fairly well demarcated soft cutaneous lesions over both his arms consistent with yaws. The main serological findings in the four cases are summarised in Table 1.



(a)



(b)

**Fig. 2.** Case 4. (a) Right fundus view showing a juxtapapillary subretinal neovascular membrane. (b) Fluorescein angiography showing leakage partly masked by overlying haemorrhage, confirming findings of (a).

**Table 1.** Serological findings

Serological tests	Case 1	Case 2	Case 3	Case 4
1 TPHA	Positive	Positive	Positive	Positive
2 FTA-Abs	Positive	Positive	Positive	Positive
3 VDRL/RPR	Positive 1:4	Positive 1:2	Positive 1:4	Positive 1:2
4 CSF <sup>a</sup>				
(i) Protein (normal = <0.4 g/l)	Raised (0.43 g/l)	Raised (0.71 g/l)	Raised	
(ii) Glucose (normal = >2.2 mmol/l)	Normal	Normal	Normal	
(iii) TPHA	Negative	Positive	Negative	
(iv) FTA-Abs	Negative	Positive	Negative	
(v) VDRL/RPR	Negative	Negative	Negative	

<sup>a</sup>Not done in in case 4.

*Comment*

Treponemes that cause disease in humans are *Treponema pallidum* (three subspecies) and *Treponema carateum*. *T. carateum* causes pinta, *T. pallidum pallidum* causes venereal syphilis, *T. pallidum pertenue* causes yaws, and *T. pallidum endemicum* causes bejel (endemic syphilis). All diseases result in host serological responses indistinguishable from one another. The causative organisms are morphologically identical and cannot readily be cultured. Standard tests for syphilis are unable to differentiate between the *T. pallidum* subspecies and between *T. pallidum* and *T. carateum*. Newer techniques such as Western blotting assays and most molecular approaches such as DNA sequencing, DNA probes and PCR techniques have also failed to distinguish the pathogenic treponemes.<sup>1</sup> Current diagnosis of these diseases as separate entities<sup>1-4</sup> is based on the clinical appearance of the lesions formed by the various treponemes, anatomical location of the lesion, mode of transmission, age and geographic location of the individual.

The non-venereal treponematoses are a group of contagious diseases endemic among rural populations in tropical and subtropical countries. All are transmitted primarily by direct contact among children and congenital transmission has not been reported. They are characterised by cutaneous, mucous membrane and osseous involvement. Ocular and neuro-ophthalmological abnormalities in the form of moderate disc pallor, light-near dissociation of the pupils, peripheral perivascular sheathing and pigmentation do occur in patients with late yaws<sup>5</sup> but subretinal neovascularisation has not been reported.<sup>6,7</sup> Choroidal neovascular membrane formation is extremely rare and has been reported in cases of acquired ocular syphilis<sup>8-11</sup> all of which were associated with uveitis which is the most common mode of presentation<sup>12</sup> (68%).

The inflammatory reaction associated with treponemal infection causes occlusion of choroidal vessels<sup>11</sup> and the structural destruction that follows may occur at the level of Bruch's membrane<sup>13</sup> and the retinal pigment epithelium, which could activate choroidal or subretinal neovascular proliferation resulting in an elevated scar (Figs. 1, 2). Differential diagnoses of inflammatory conditions<sup>8</sup> of the choroid and retina associated with choroidal neovascularisation include

histoplasmosis, toxoplasmosis, sarcoidosis, chronic uveitis, Behçet's syndrome, serpiginous choroidopathy and tuberculosis.<sup>13</sup>

All four patients came from areas endemic<sup>4</sup> for yaws and all denied a past history of venereal syphilis or mucous membrane involvement. A definite history of yaws and evidence of cutaneous involvement in cases 1, 3 and 4; positive treponemal tests in all four cases; positive cerebrospinal fluid serology in one and raised protein in three of the cases give reasonable cause to suspect a treponemal infection. Our cases are interesting in that none had evidence of ocular inflammation in the form of anterior chamber or vitreous activity or retinal vasculitis. This was probably the reason for delay in performing treponemal tests.

Therefore, we recommend testing for treponemal serology in patients with subretinal neovascularisation in the periphery, juxtapapillary, paramacular and macular areas, especially when they originate from countries where yaws is endemic. In patients with yaws, superimposed venereal infection cannot be ruled out, so it is advisable to administer a course of systemic penicillin or other appropriate antibiotic.

We would like to thank Mr Richard Smith and Dr Luzzi for their advice.

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Sir,

**Late-onset capsular block syndrome: an occult cause of decreased vision in a 63-year-old pseudophakic Asian woman**

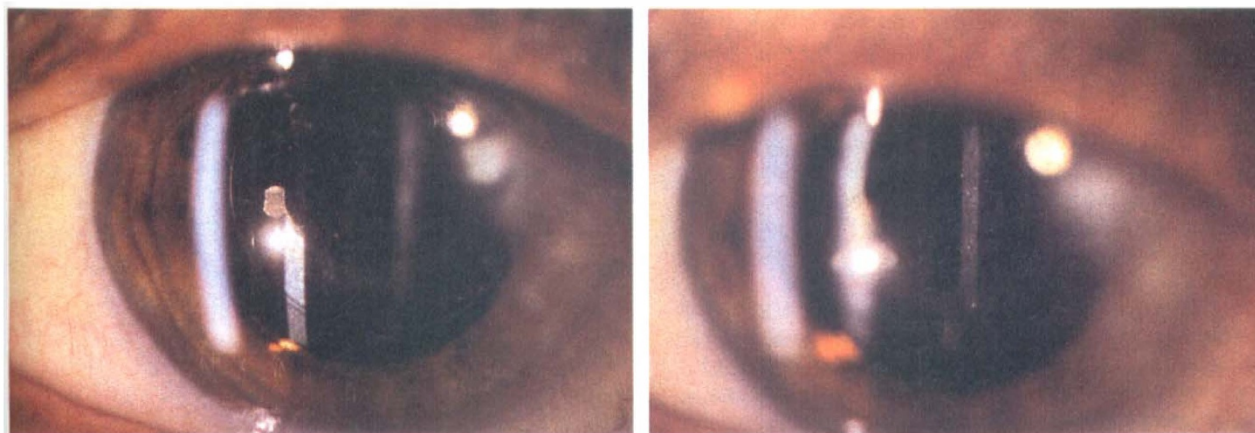
A 63-year-old Indian woman underwent uneventful right phacoemulsification with intraocular lens implantation. Pre-operative assessment showed an unaided right visual acuity of 6/60, improving to 6/18 with pinhole testing. Fundoscopy was difficult due to poor mydriasis but showed normal fundi.

Operation details included a temporal clear corneal two-step incision, continuous curvilinear capsulorhexis aided with Provisc (Alcon) viscoelastic substance and phacoemulsification via the 'divide and conquer' technique. An Acrysof (Alcon) acrylic foldable intraocular lens was implanted and the viscoelastic substance aspirated.

The patient was discharged with an unaided right visual acuity of 6/12, improving to 6/9 with spectacle correction (OD -0.50/-0.50 × 180).

A year later, she reattended with decreased right visual acuity of 6/60, improving to 6/18 with pinhole. Anterior segment examination showed quiet pseudophakia with an anteriorly displaced intraocular lens such that the anterior surface of the intraocular lens implant optic was in apposition with the edge of the pupil. Fundoscopy was again difficult due to poor mydriasis but was normal. This was confirmed by intravenous fluorescein angiography.

Only after 30 min of intensive cyclopentolate and phenylephrine drop instillation could satisfactory slit-lamp examination of the anterior vitreous cavity be performed, showing a posteriorly displaced fibrotic posterior capsule with anteriorly located lens implant (Fig. 1). Retinoscopy demonstrated a myopic shift (OD -2.50/-0.50 × 178 6/18). A diagnosis of late-onset capsular block syndrome was made and Nd:YAG laser posterior capsulotomy performed (Fig. 2). The lens implant returned to its normal position, with the unaided right visual acuity improving to 6/12 (-0.50/-0.75 × 173 6/9).



**Fig. 1.** Digital photographs of the anterior segment prior to Nd:YAG laser capsulotomy. The depth of field is equivalent to the distance between the cornea and the intraocular lens (IOL). Left: The posterior capsule is out of focus when focusing on the cornea. Right: The cornea is out of focus when focusing on the IOL.