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Sir,

**Malignant solitary fibrous tumour of the orbit: report of a case with 8 years follow-up**

Solitary fibrous tumour (SFT) is a rare spindle cell neoplasm of the pleura of adults. It has been recently described in the orbit[1] among other extrapleural sites.[2,3] A small percentage of this type of tumour has malignant histological features and may manifest aggressive clinical behaviour, including recurrence, invasion of adjacent tissues and distant metastases.[4] No cases of the malignant variant have been reported in the upper respiratory tract and orbit.[1] We report the first case of a malignant orbital SFT with several recurrences, and showing progressive histological dedifferentiation and local invasion.

*Case report*

*Clinical findings.* A 64-year-old man presented unilateral exophthalmos due to a palpable mass in his left upper orbit. The lesion had been resected twice in the past 3 years since presentation. The mass is now radiologically well defined, with a medium signal on T1, in the upper external orbit, and pushed slightly on the optic nerve (Fig. 1, left).

A 3.3 cm macroscopically well-circumscribed mass was excised. It showed micoscopic involvement of the margins by the neoplasm. Therefore, an exenteration was performed. There was no evidence of residual tumour by macroscopic or microscopic examination of the specimen. Thirty-eight months later the patient had a recurrence of the tumour on the base of the orbit. It extended through the lateral wall, temporal muscle fibres and subcutaneous tissue. A new resection of various neoplastic nodules measuring < 3 cm in total was performed. However, the lesions recurred again 13 months later. A mass of 5 cm was found invading through the lateral wall of the orbit, temporal muscle and the maxillary region (Fig. 1, right). The patient died with a cervical abscess shortly after the surgical resection and 8 years after the onset of the orbital symptoms.

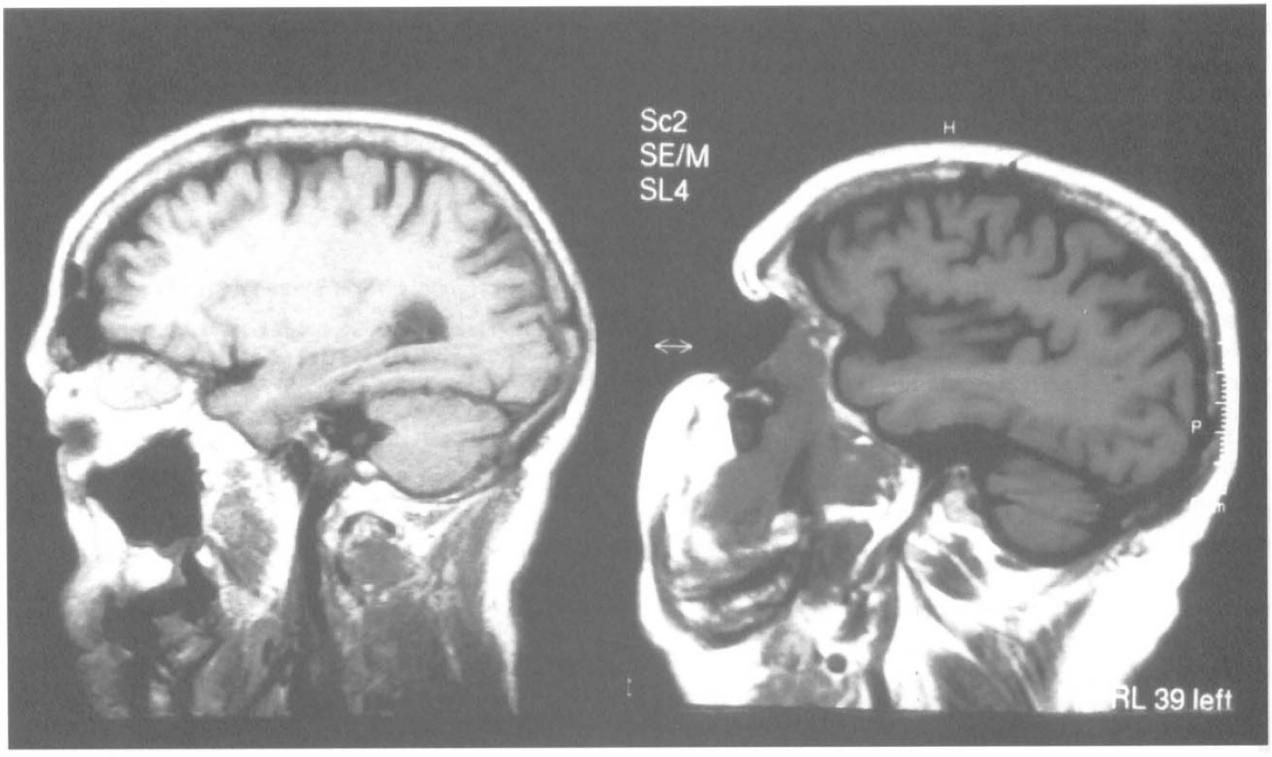


**Fig. 1.** *CT scan performed before exenteration (left) and extension of the tumour 38 months later (right).*

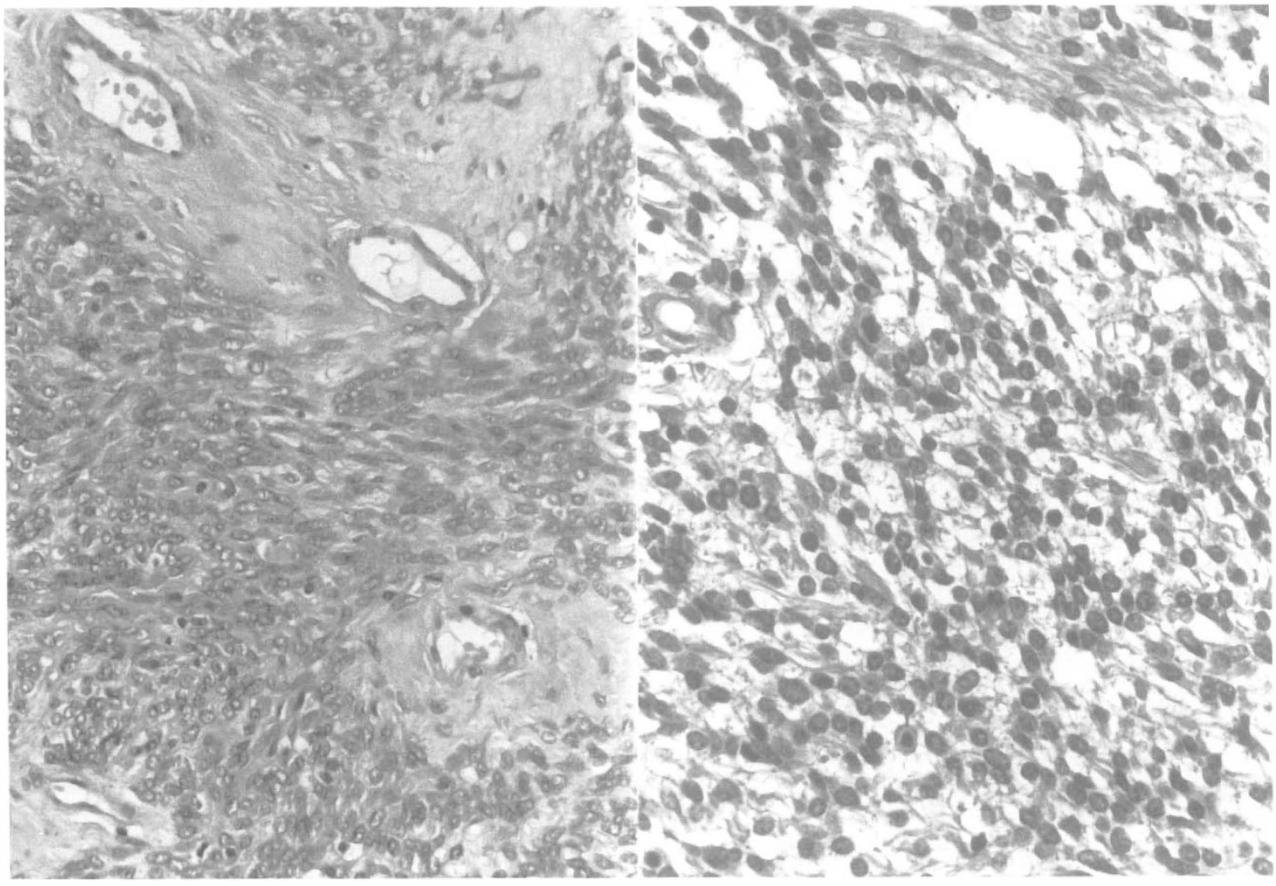

**Fig. 2.** *Microscopic appearance of the first specimen examined (left) and the dyscohesive appearance of the tumour obtained from the last resection (right) (HE, ×40).*

*Histological examination.* The first specimen consisted of a fascicular tumour composed of spindle cells with finely dispersed nuclear chromatin and inconspicuous nucleoli alternating with less cellular sclerotic areas (Fig. 2, left). The mitotic count was 6 or 7 mitoses per square millimetre. There was no cellular pleomorphism, giant cells or confluent necrosis. Immunohistochemical examination revealed negatively for EMA (an epithelial marker). The tumour showed immunoreactivity for CD34, a vascular marker known to be positive in SFT. Morphology of the last recurrence (Fig. 2, right) showed dyscohesive tumour cells with focal single cell necrosis and no sclerotic area. The tumour involved bone marrow. Ki 67, which is a marker of cell proliferation, showed increased staining in the tumour, indicating an increased proliferative rate. The diagnosis was SFT of the orbit with histological evidence of progressive dedifferentiation to fibrosarcoma.

*Comment*

SFT is an uncommon neoplasm that was originally thought to occur exclusively in the intrathoracic region. SFT has been described more recently in extraserosal and extrathoracic sites. Classically the tumour is composed of bland spindle cells with alternating hyper- and hypocellular areas, keloid-like hyalinisation (described as a 'patternless pattern') and a frequently prominent vasculature which resembles that of a haemangiopericytoma.[2] Complete local excision is curative in most cases but recurrence or metastasis following complete resection has been described after more than 5 years. Recurrence and distant metastases occur in approximately 10–15% of cases. Twenty-three per cent of patients die of their tumour. Few extrathoracic malignant cases have been described to date.[4] Histological features associated with local or distant recurrence of intrathoracic SFT include high cellularity, mitotic activity, nuclear pleomorphism and necrosis. The primary tumour is usually of a lower histological grade than the recurrence. This pattern of progression was found in our case and is the first report of malignant behaviour in this location. Histogenesis of this neoplasm, originally thought to be of mesothelial origin, is now considered of mesenchymal, possibly fibroblastic origin.[5] Morphological and immunohistochemical studies have identified the SFT cells as relatively undifferentiated mesenchymal cells. CD34 and O13[4] are the only immunohistochemical markers frequently found in these cells.

The differential diagnosis outside the subserosal location includes fibrosarcoma, fibrous histiocytoma, haemangiopericytoma and other spindle tumours such as malignant nerve sheet tumour and meningioma. The so-called patternless pattern of SFT may focally mimic other tumours such as monophasic synovial sarcoma and leiomyosarcoma. Fibrous histiocytoma includes varying proportions of histiocytic cells, some of them multinucleated and having a predominantly storiform

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 sheet 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.