hypoxic factors prevailing within the retina. Some modification of fluid dynamics relating to inner retinal and pigment epithelial function was achieved using acetazolamide, and BMT consolidated this benefit. This suggests that definitive treatment of the leukaemia has removed the stimulus for CMO and allowed recovery of vision, despite persisting features of blood–retinal barrier disruption.

S. E. Horgan[1]
S. G. Fraser[2]
P. F. Ferrante[1]
A. Spencer[3]
M. Beaconsfield[1]

[1]Department of Ophthalmology
Oldchurch Hospital
Romford
Essex, UK
[2]Department of Ophthalmology
Whipps Cross Hospital
London, UK
[3]LRF Adult Leukaemia Unit
Hammersmith Hospital
London, UK

Correspondence to:
Mr Simon Horgan, FRCOphth
Moorfields Eye Hospital
City Road
London EC1V 2PD, UK

References

1. Kincaid MC, Green WR. Ocular and orbital involvement in leukaemia. Surv Ophthalmol 1983;27:211–32.
2. Holt JM, Gordon-Smith EC. Retinal abnormalities in diseases of the blood. Br J Ophthalmol 1969;53:145–60.
3. Duke JR, Wilkinson CP, Sigelman S. Retinal microaneurysms in leukaemia. Br J Ophthalmol 1968;52: 368–74.
4. Morse PH, McCready JL. Peripheral retinal neovascularisation with chronic myelocytic leukaemia. Am J Ophthalmol 1971;72:975–8.
5. Kincaid MC, Green WR, Kelley JS. Acute ocular leukaemia. Am J Ophthalmol 1979;87:698–702.
6. Gass JD, Anderson DR, Davis EB. A clinical, fluorescein angiographic, and electron microscopic correlation of cystoid macular edema. Am J Ophthalmol 1985;100:82–6.
7. Cox SN, Hay EH, Bird AC. Treatment of chronic macular oedema with acetazolamide. Arch Ophthalmol 1988;106:1190–5.
8. Spalton DJ, Bird AC, Cleary PE. Retinitis pigmentosa and retinal oedema. Br J Ophthalmol 1978;62:174–82.
9. Tso M. Animal modelling of human cystoid macular edema. Surv Ophthalmol 1984;28(Suppl):512–9.
10. Tso M. Pathology of cystoid macular edema. Ophthalmology 1982;89:902–14.
11. Tsuboi S. Fluid movement across the blood–retinal barrier: a review of studies by vitreous flourophotometry. Jpn J Ophthalmol 1990;34:133–41.
12. Tsuboi S, Pederson JE. Acetazolamide effect on the inward permeability of the blood–retinal barrier. Invest Ophthalmol Vis Sci 1987;28:92–5.

Sir,

Solitary Fibrous Tumour: An Atypical Presentation Within the Orbit

Solitary fibrous tumour is a rare neoplasm which more commonly arises in the pleura.[1–7] Extrapleural sites are recognised including mediastinum,[5,8,9] pericardium,[4] upper respiratory tract, nasal cavities,[10–12] lung parenchyma,[13] peritoneum[4,14] tunica vaginalis of the testis,[15] liver, thyroid and parotid glands.[16] Recently, orbital involvement has been recognised.[17,18]

Case Report

A 23-year-old woman, who gave a 2 year history of a watery left eye, presented with a painless swelling over the left medial canthus which had slowly enlarged over 6 months. Clinical examination revealed a large, rubbery mass with no evidence of proptosis. Ultrasonography of the orbit demonstrated a well-circumscribed lesion medially, of fairly low reflectivity. This was confirmed on CT scanning (Fig. 1).

Exploration and surgical excision of the lesion was performed under general anaesthesia. The upper pole of the lesion was visible just below the medial palpebral ligament, and was adherent to the periosteum and the lacrimal sac. Probes were placed in the upper and lower canaliculus, and the tumour was carefully dissected. Anterior and posterior dacryocystorhinostomy flaps were fashioned, O'Donoghue tubes were inserted, and the wound closed.

Pathology. A bosselated, smooth, grey-brown mass measuring 2.2 × 1.3 × 1 cm was removed from the left orbit. It was firm and showed a whorled, pale grey cut surface. Microscopically, it was composed of

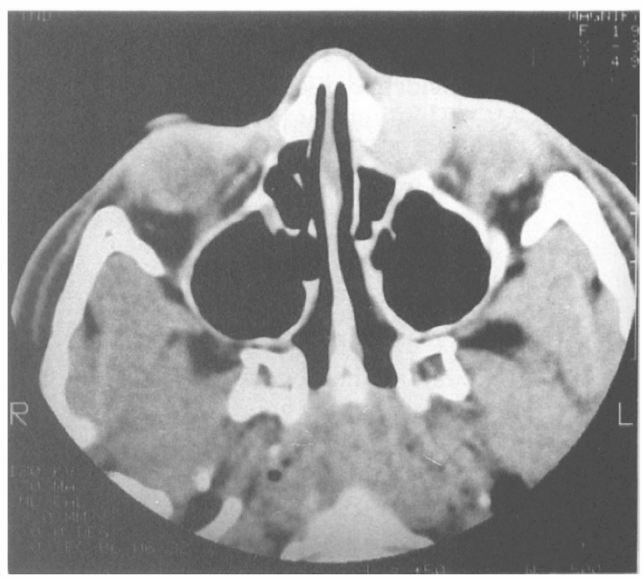


Fig. 1. *CT scan showing lesion in left medial canthal region.*

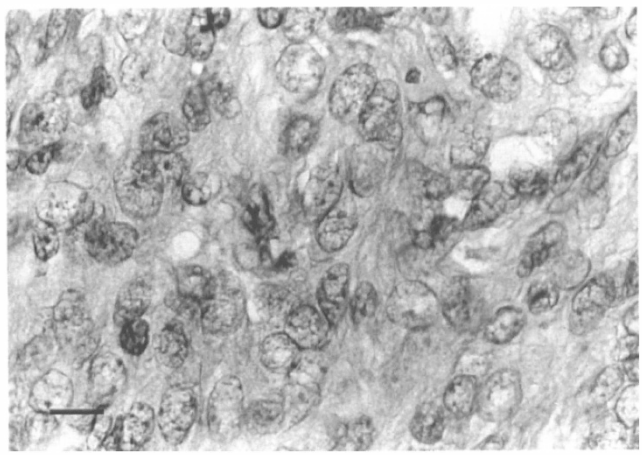


Fig. 2. *Cohesive polygonal cells from part of the solitary fibrous tumour (haematoxylin & eosin). Scale bar represents 10 μm.*

polygonal (Fig. 2) and spindle-shaped cells (Fig. 3) with ill-defined cytoplasm and oval vesicular nuclei. There was no evidence of mitotic activity or necrosis. Abundant hyalinised collagen was interspersed between clusters of cells and isolated cells. The tumour showed a prominent vascular pattern with focally dilated staghorn-like vascular channels, resembling a haemangiopericytoma. The mass showed a predominant expansile growth pattern, and extended from the lining of the nasolacrimal duct to surrounding skeletal muscle. An additional and distinct neoplasm located just beneath the nasolacrimal duct was identified. It was composed of naevus cells which contained melanin granules, and these naevus cells interdigitated with the spindle-shaped cells of the solitary fibrous tumour. These appearances indicated a collision of two distinct neoplasms.

Immunohistochemistry. The neoplastic cells from the bulk of the tumour exhibited vimentin positivity and

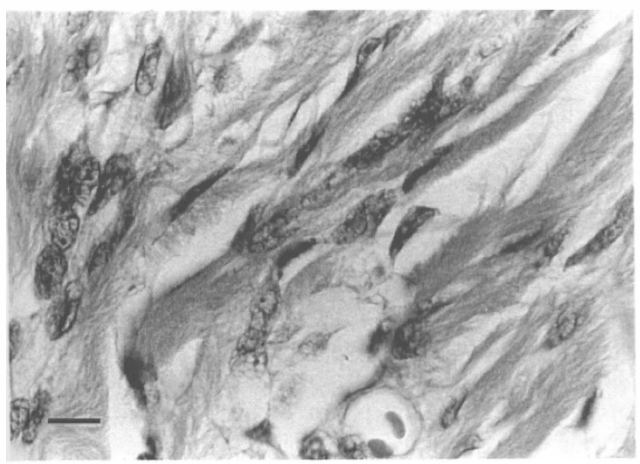
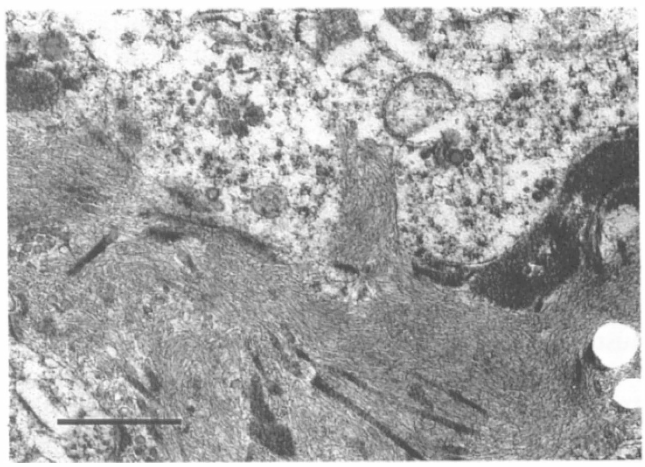
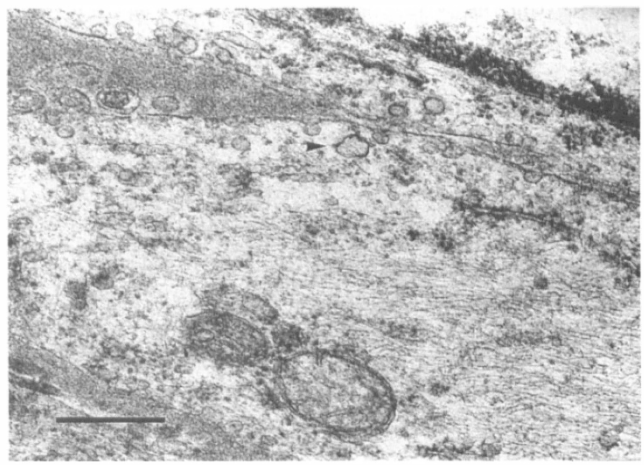


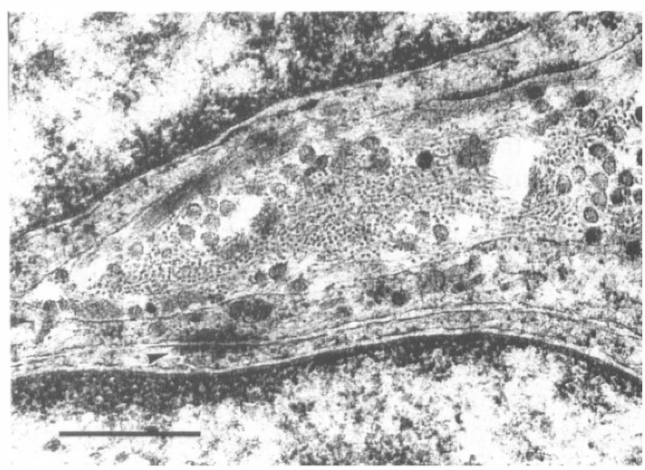
Fig. 3. *Solitary fibrous tumour showing spindle cells separated by dense collagen bundles (haematoxylin & eosin). Scale bar represents 10 μm.*



(a)



(b)



(c)

Fig. 4. *Montage of electron micrographs of solitary fibrous tumour. (a) A cell containing mitochondria and ribosomes surrounded by extracellular collagen fibrils and glycoprotein filaments. Scale bar represents 1 μm. (b) Intermediate filaments and plasmalemmal caveolae (arrowhead). Scale bar represents 500 nm. (c) Collagen fibres and hollow glycoprotein filaments in transverse section. The cytoplasm and nuclei of two cells lie above and below. A subplasmalemmal linear density (arrowhead) joins the cytoplasm of two cells. Scale bar represents 500 nm.*

strong CD34 positivity, but were negative for S100, CAM 5.2, EMA and smooth muscle actin (SMA). The melanised cells just beneath the nasolacrimal duct were positive for S100 after bleaching, whereas the cells of the solitary fibrous tumour were negative.

Electron Microscopy. Electron microscopy of the neoplastic cells showed a high nuclear/cytoplasmic ratio with plump polygonal and spindle-shaped cells. Various areas of the neoplasm showed both cohesive clusters of cells and cells separated from one another by a collagen and glycoprotein filament containing stroma (Fig. 4a). The cytoplasm of these neoplastic cells contained scanty mitochondria, rough endoplasmic reticulum and some free ribosomes. The rough endoplasmic reticulum was often dilated. Cytoplasmic intermediate filaments (Fig. 4b) were present but scanty and several centrioles were identified. The nuclei were oval but showed focally crenellated nuclear membranes and many contained a prominent nucleolus. There were plasmalemmal caveolae (Fig. 4b) and the cells in contact with one another showed subplasmalemmal linear densities (Fig. 4c). Basal laminae were present only in association with endothelial cells and pericytes. The melanised cells from just beneath the nasolacrimal duct were not included in the electron microscopic sample, but the neoplastic cells from the bulk of the lesion showed no melanosomes. The overall appearances of the neoplastic cells were consistent with mesenchymal differentiation.

Discussion

Solitary fibrous tumour was originally described in the pleura by Klemperer and Rabin in 1931,[1] who suggested an origin from subpleural mesenchyme. However, the subsequently postulated origin from mesothelium has caused confusion and has been shown to be incorrect by both immunohistochemical and electron microscopical techniques.[4,10] Only in recent years has the tumour's occurrence in extrapleural sites been appreciated,[9–18] which is an additional observation against a mesothelial origin.

The clinical behaviour is variable. Most solitary fibrous tumours are benign, but local invasion or recurrence of the lesion has been demonstrated and, rarely, distant metastases have been reported.[3,5,8] Long-term follow-up of these patients is therefore essential. Complete excision of pleural solitary fibrous tumour provides excellent results.[5] A proportion of those tumours which show local invasion have responded well to excision alone or to excision with chemotherapy and radiotherapy.[5] Seven such patients survived a median of 32 months.[5]

Orbital solitary fibrous tumour is thought to behave in a non-aggressive manner, similar to solitary fibrous tumour occurring at other sites in the head and neck.[10,11] However, because of the small number of reported cases, it is difficult to confirm that tumours of extrapleural sites behave differently from those of the pleura. Three of the five reported cases of solitary fibrous tumour of the orbit presented with gradually worsening proptosis.[17] The other two cases presented with orbital masses.[18] In one, the tumour recurred 4 years after the initial excision, and recurred again after another 2 years. In our case, full macroscopic excision was achieved, which was confirmed microscopically. In addition, features of aggressive behaviour or malignancy, such as high cellularity, mitotic activity, anaplasia and necrosis, were absent. Proptosis was not a feature and the patient was noted to be significantly younger than the previously reported age range of 44–69 years at diagnosis.

Mesenchymal neoplasms, which feature in the differential diagnosis at this location, include fibrous histiocytoma, haemangiopericytoma, tumours of nerve sheath origin and meningioma.

Solitary fibrous tumours are thought to arise from mesenchymal cells or fibroblasts,[17] and show spindle and sclerosing patterns of growth often with areas resembling haemangiopericytoma. Immunohistochemistry shows negativity for a wide range of markers. This includes epithelial markers such as keratin, epithelial membrane antigen (EMA), neuroectodermal markers such as S100, markers of muscle differentiation such as desmin and smooth muscle actin, and endothelial markers such as factor VIII related antigen (FVIII Ag). However, the cells are positive for vimentin (which may be used as a marker of mesenchymal cells) and are strongly positive for CD34, which is a single chain transmembrane glycoprotein and a marker of endothelial cells and haemopoietic progenitor cells. Strong positivity for CD34 is characteristic for solitary fibrous tumour.[17,18] Ultrastructurally, the cells of solitary fibrous tumours are rather bland and are surrounded by extracellular matrix indicating mesenchymal differentiation. As expected, there are no immunohistochemical or ultrastructural features of mesothelial or epithelial differentiation. Our findings are in keeping with the current view that solitary fibrous tumours of the orbit are derived from mesenchyme.[8–10,17–19]

Our case also differs from previously reported cases in the presence of a naevocytic neoplasm, best described pathologically as a blue naevus, which interdigitated with the superficial parts of the solitary fibrous tumour. This is an example of a collision tumour whereby two independent tumours arising in a similar location blend together as they grow. Increased awareness of the widespread occurrence of solitary fibrous tumour has led to an increase in

diagnosis in recent years, and recognition of examples in the orbit in only the past 2 years.

We thank Professor J. Rosai at Memorial Sloan-Kettering Cancer Centre, New York, for confirming the diagnosis, Mr B. Eyden, Christie Hospital NHS Trust, Manchester, for advice on the electron microscopy, and Mrs P. Tarpey, Department of Histopathology, Manchester Royal Infirmary, for technical assistance.

A. M. McElvanney
J. L. Noble
Manchester Royal Eye Hospital
Oxford Road
Manchester M13 9WH
UK

D. G. O'Donovan
R. E. Bonshek
Department of Pathological Sciences
University of Manchester
Manchester M13 9PT
UK

S. S. Banerjee
Department of Histopathology
Christie Hospital NHS Trust
Manchester M20 9BX
UK

References

1. Klemperer P, Rabin CB. Primary neoplasms of the pleura: a report of 5 cases. Arch Pathol 1931;11: 385–412.
2. Dalton WT, Zolliker AS, McCaughey WTE, Jacques J, Kannerstein M. Localised primary tumours of the pleura: an analysis of 40 cases. Cancer 1979;44:1465–75.
3. Briselli M, Mark EJ, Dickersin GR. Solitary fibrous tumors of the pleura: eight new cases and review of 360 cases in the literature. Cancer 1981;47:2678–89.
4. El-Naggar AK, Ro JY, Ayala AG, Ward R, Ordonez NG. Localised fibrous tumor of the serosal cavities: immunohistochemical, electron-microscopic, and flow-cytometric DNA study. Am J Clin Pathol 1989;92: 561–5.
5. England DM, Hochholzer L, McCarthy MJ. Localised benign and malignant fibrous tumors of the pleura: a clinicopathologic review of 223 cases. Am J Surg Pathol 1989;13:640–58.
6. Moran CA, Suster S, Koss MN. The spectrum of histologic growth patterns in benign and malignant fibrous tumours of the pleura. Semin Diagn Pathol 1992;9:169–80.
7. Van de Rijn M, Lombard CM, Rouse RV. Expression of CD34 by solitary fibrous tumors of the pleura, mediastinum and lung. Am J Surg Pathol 1994;18: 814–20.
8. Witkin GB, Rosai J. Solitary fibrous tumor of the mediastinum: a report of 14 cases. Am J Surg Pathol 1989;13:547–57.
9. Goodlad JR, Fletcher CDM. Solitary fibrous tumour arising at unusual sites: analysis of a series. Histopathology 1991;19:515–22.
10. Zukerberg LR, Rosenberg AE, Randolph G, Pilch BZ, Goodman ML. Solitary fibrous tumor of the nasal cavity and paranasal sinuses. Am J Surg Pathol 1991;15:126–30.
11. Witkin GB, Rosai J. Solitary fibrous tumor of the upper respiratory tract: a report of six cases. Am J Surg Pathol 1991;15:842–8.
12. Batsakis JG, Hybels RD, El-Naggar AK. Pathology consultation: solitary fibrous tumor. Ann Otol Rhinol Laryngol 1993;102:74–6.
13. Yousem SA, Flynn SD. Intrapulmonary localised fibrous tumor: intraparenchymal so-called localized fibrous mesothelioma. Am J Clin Pathol 1988;89:365–9.
14. Young RH, Clement PB, McCaughey WTE. Solitary fibrous tumors ('fibrous mesotheliomas') of the peritoneum: a report of three cases and a review of the literature. Arch Pathol Lab Med 1990;114:493–5.
15. Parveen T, Fleischmann J, Petrelli M. Benign fibrous tumor of the tunica vaginalis testis: report of a case with light, electron microscopic, and immunocytochemical study, and review of the literature. Arch Pathol Lab Med 1992;116:277–80.
16. Sobrinho-Simoes M, Fonseca E. Recently described tumours of the thyroid. In: Anthony PP, MacSween RNM, editors. Recent advances in histopathology, vol. 16. Edinburgh: Churchill Livingstone, 1994:213–29.
17. Dorfman DM, To K, Dickersin GR, Rosenberg AE, Pilch BZ. Solitary fibrous tumor of the orbit. Am J Surg Pathol 1994;18:281–7.
18. Westra WH, Gerald WL, Rosai J. Solitary fibrous tumor: consistent CD34 immunoreactivity and occurrence in the orbit. Am J Surg Pathol 1994;18:992–8.
19. Dervan PA, Tobin B, O'Connor M. Solitary (localized) fibrous mesothelioma: evidence against mesothelial cell origin. Histopathology 1986;10:867–75.

Sir,

Cryoglobulinaemia Masquerading as Rheumatoid Vasculitis: The Retina Provides the Clue

We present a case of a severe systemic illness in which retinal observation helped prompt further investigations leading to the diagnosis of cryoglobulinaemia. The ocular complications of cryoglobulinaemia have rarely been documented.

Case Report

A 72-year-old Caucasian man with severe erosive rheumatoid arthritis presented with painful and swollen lower legs, with necrotic ulcers on three of his toes and over both medial malleoli. Both feet were warm and his peripheral pulses were evident. An erythematous petechial rash with macules and telangiectasia was noted below the knees; this later spread to his forearms and face (Figs. 1, 2). Skin biopsy showed IgM and C3 deposits with occasional IgG consistent with an immune complex vasculitis. A diagnosis of rheumatoid vasculitis was made and he was admitted for systemic immunosuppression with intravenous pulses of cyclophosphamide and methylprednisolone.

He then complained of a severe reduction in vision. Visual acuities were 6/36 in the right and 6/60 in the left. There was no relative afferent pupillary defect. Anterior segment and vitreal examination of