The Journal of Laryngology and Otology
July 1994, Vol. **108**, pp. 613–616

Pathology in Focus

Inflammatory pseudotumour of the trachea: report of a case in an eight-year-old child

FARRUKH AIJAZ, M.R.C.PATH.*, ANWAR US SALAM, F.R.C.S.†, SUHAIL MUZAFFAR, M.B.B.S.*, YASMIN AKBANI, M.R.C.P.†, SHEEMA H. HASAN, F.R.C.PATH.*

Abstract
A polypoid inflammatory pseudotumour was diagnosed in the trachea of an eight-year-old child who presented with asthmatic symptoms. The tumour showed 80 per cent blockage of the lower trachea and consisted of proliferating spindly fibroblastic cells admixed with a variable number of inflammatory cells. The literature on childhood inflammatory pseudotumours is reviewed together with the differential diagnosis of other polypoid mesenchymal tumours of the trachea.

Key words: Trachea; Granuloma, plasma cell

Introduction

Primary tracheal tumours are uncommon lesions and overwhelmingly the majority of these are malignant (Pearson *et al.*, 1984). Benign mesenchymal tumours of the trachea are exceedingly rare but quite a variety of these have been reported. Inflammatory pseudotumour and its synonyms such as plasma cell granuloma and inflammatory myofibrohistiocytic proliferations, etc., are most likely to be reactive lesions that may simulate sarcomas. They constitute an important differential diagnostic consideration in the evaluation of spindle cell lesions in children especially at certain sites such as the lung (Hartman and Shochat, 1983). We have found no report of an inflammatory pseudotumour of the trachea in children in the English literature. We report a case of inflammatory pseudotumour in the trachea of an eight-year-old child with a brief review of this lesion and other benign tracheal mesenchymal tumours.

Case report

An eight-year-old child presented to the emergency room with acute respiratory distress. There was a history of a variable degree of breathlessness throughout the previous eight months. He was admitted and treated for 'status asthmaticus' without any singnificant improvement. Physical and laboratory examination at the time of his initial presentation were unremarkable except for mild stridor and rhonchi. Bronchoscopy was performed to exclude a foreign body and revealed a pink polypoidal mass obstructing almost 80 per cent of the tracheal lumen. The mass has a smooth surface, was attached by a broad base to the anterior, right lateral and posterior walls of the trachea and was located 1 cm above the carina. The tumour was excised piecemeal, but completely, and mild local bleeding was controlled. The proximal trachea and major bronchi were unremarkable. There was significant clinical improvement after the removal of the tumour and the child was discharged on the fourth day. The patient was in an excellent condition at follow-up two months after surgery.

Microscopic features of the polypoidal mass

Multiple fragments of the lesion showed a cellular lesion consisting predominantly of relatively uniform spindle cells arranged more or less in a fascicular fashion (Figures 1 and 2). The overlying epithelium was relatively intact with focal ulceration. The individual cells had elongated to oval vesicular nuclei with open chromatin pattern and occasionally prominent nucleoli. Some cells showed elongated, tapering, biopolar processes. These spindle cells were variably accompanied by plasma cells. Histiocytes, foamy macrophages, neutrophils and some eosinophils were also present focally. Thin walled capillaries and oedematous myxoid areas together with extravasated red blood cells reminiscent of nodular fasciitis were noted (Figure 3). Mitoses were present. They were typical and two mitotic figures occurred per ten high power fields. Necrosis, nuclear atypia, rhabdomyoblasts and subepithelial cambium layer were absent. The tumour was uniformly positive for vimentin but only rare cells showed positivity for desmin. Cytokeratin and S100 were negative.

Discussion

Inflammatory pseudotumour has many synonyms including inflammatory myofibroblastic tumour (Pettinato *et al.*, 1990), inflammatory myofibrohistiocytic proliferation (Tang *et al.*, 1990), fibroxanthoma (Dubilier *et al.*, 1968), histiocytoma (Bates and Hull, 1958) and plasma cell granuloma (Bahadori and Liebow, 1973). Most of these lesions have been described in the lung (Bahadori and Liebow, 1973), and also in the urinary tract (Fish and Brodey, 1976; Nochomovitz and Orenstein, 1985),

From the Department of Pathology*, The Aga Khan University Hospital and the Departments of ENT† and Paediatrics, Abbasi Shaheed Hospital, Karachi, Pakistan.
Accepted for publication: 7 January 1993.

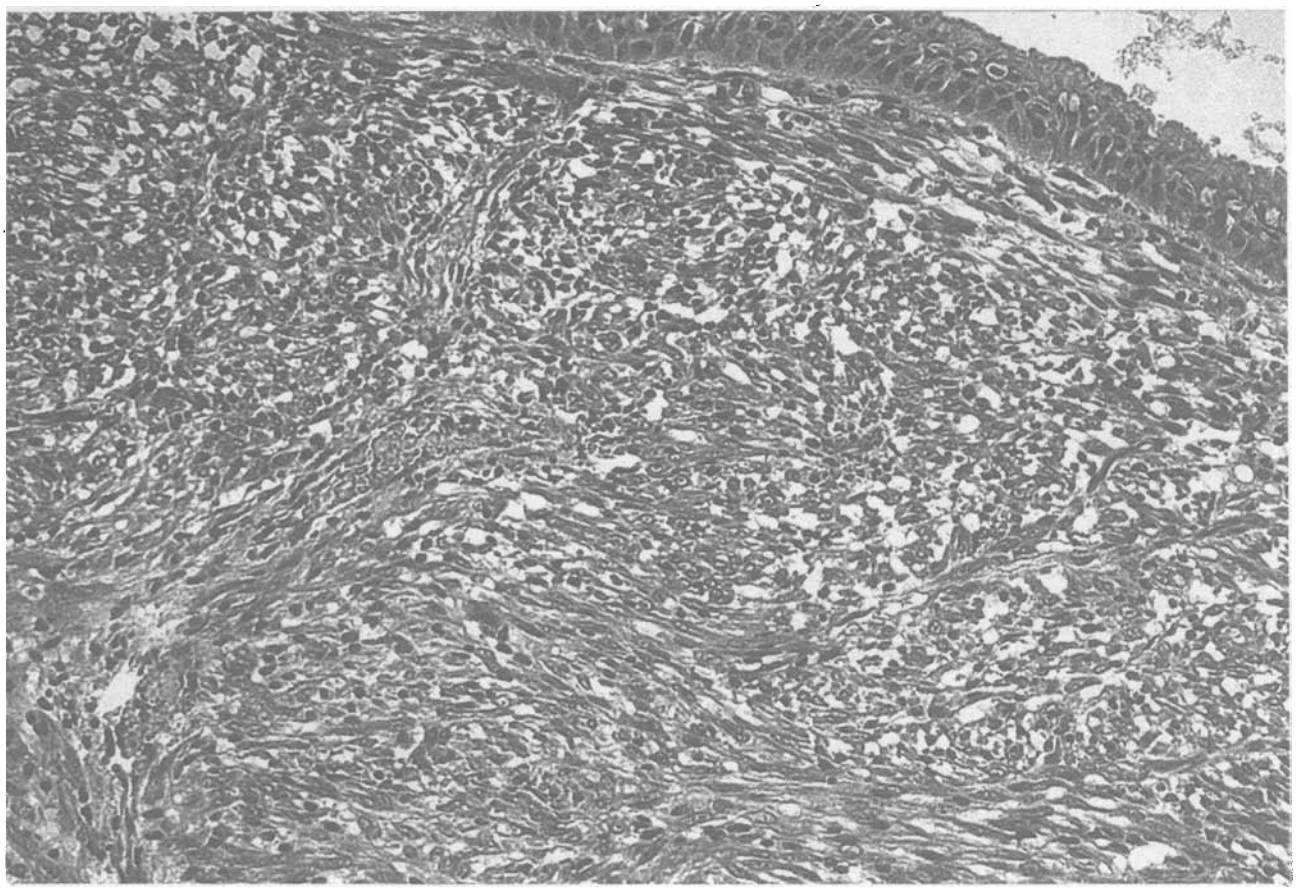

FIG. 1
Inflammatory cells intermingled with spindly fibroblastic cells. Note intact epithelial lining and absence of cambium layer. (H&E; ×100).

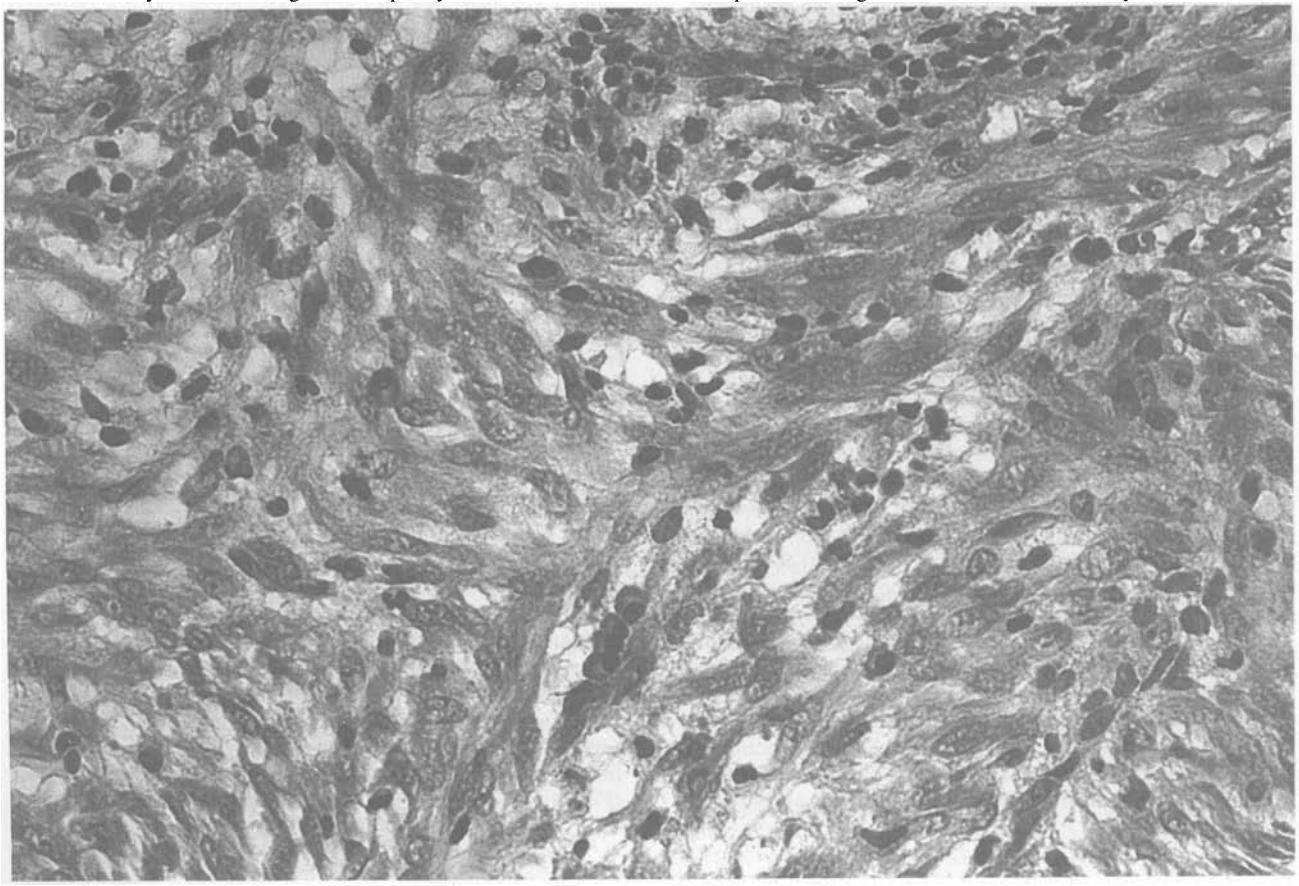

FIG. 2
High power view showing plasma cells, other inflammatory cells and spindly fibroblastic cells. (H&E; ×400).

https://doi.org/10.1017/S0022215100127604 Published online by Cambridge University Press

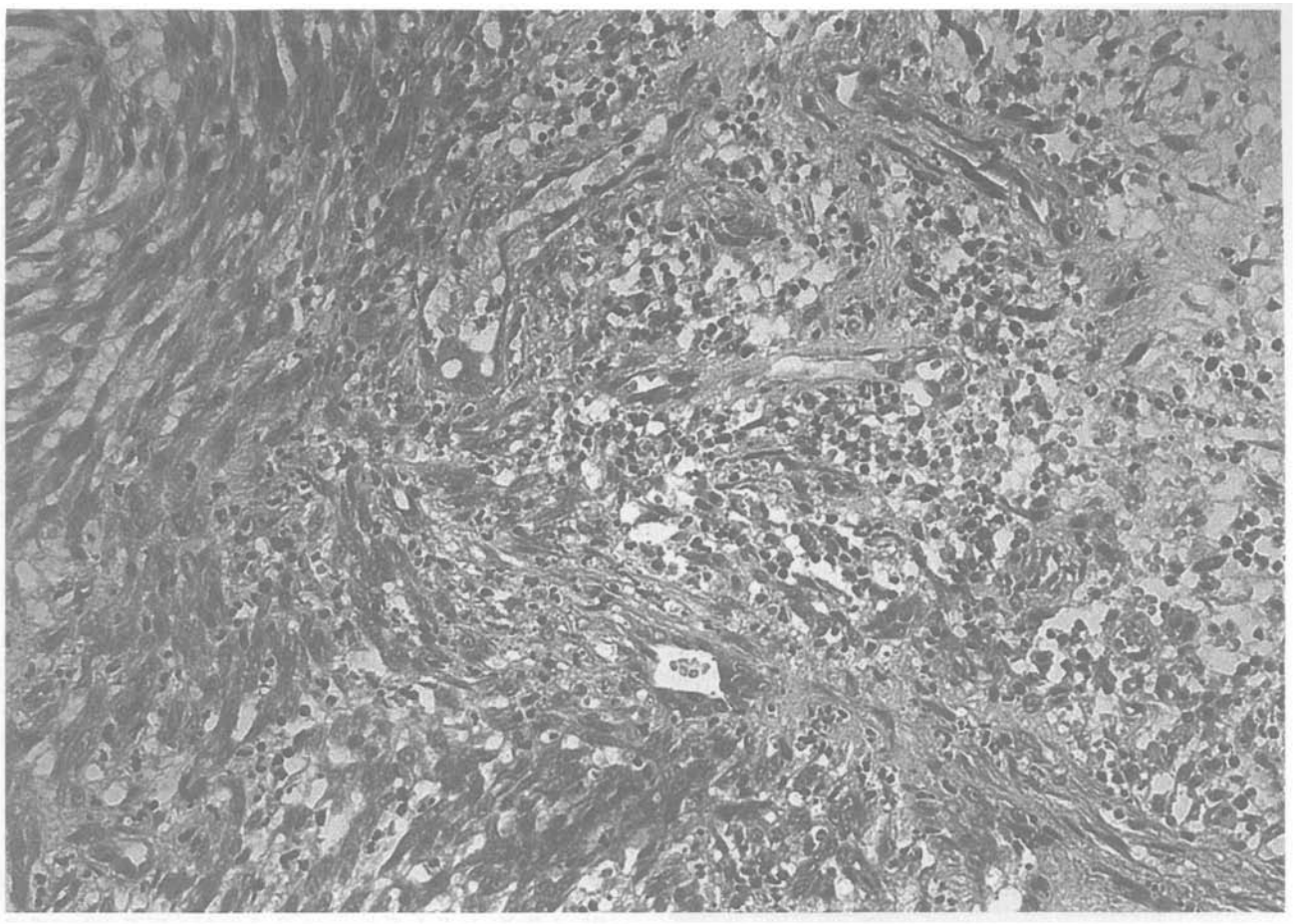

FIG. 3
Myxoid inflammatory areas with thin walled capillaries. (H&E; ×200).

intra-abdominal organs (Soga *et al.*, 1970; Wu *et al.*, 1973; Anthony and Telesinghe, 1986; Day *et al.*, 1986), lymphoid tissues (Cotelingam and Jaffe, 1984; Perrone *et al.*, 1988), central nervous system (Eimoto *et al.*, 1978; West *et al.*, 1980) etc. The basic morphological theme of these lesions consists of spindle cell proliferation admixed with a variable number and population of inflammatory cells including plasma cells, lymphocytes, histiocytes and foamy macrophages along with a minor component of neutrophils and eosinophils. There could be marked variation in different tumours depending upon the quantitative differences of various elements composing a given tumour. This variation can also be seen in different areas of a single tumour. These tumours have been described in all age groups but their incidence in children is significant. In one series these lesions constituted 20 per cent of all primary lung tumours and 57 per cent of all benign lung tumours (Hartman and Shochat, 1983). The spindle cells which normally form an important component of the lesion have been shown to be of fibroblastic and myofibroblastic nature on the basis of ultrastructural and immunohistochemical findings (Pettinato *et al.*, 1990). The true nature of these lesions is still not known but the general opinion is in favour of a reactive process. However, in the majority of cases no history of previous trauma or infectious process has been documented. Some seemingly totally removed lesions have recurred while some incompletely excised lesions have shown total resolution (Mandelbaum *et al.*, 1981). Most of the information regarding these tumours has originated from pulmonary lesions because they are the commonest.

Primary tracheal tumours whether epithelial or mesenchymal are rare. The majority of the primary tracheal tumours are malignant (Pearson *et al.*, 1984) and most of these are squamous and adenoid cystic carcinomas (Olmedo *et al.*, 1982). Among the benign mesenchymal tumours the reported entities have included fibromas (Gilbert *et al.*, 1953; MacLachlan, 1968), fibromyxoma (Pollak *et al.*, 1985), fibrous histiocytoma (Sandstorm *et al.*, 1978), granular cell tumour (Mikaelian *et al.*, 1984), leiomyoma (Harris *et al.*, 1967), and neurilemmoma (Horovitz *et al.*, 1983). Paraganglioma (Liew *et al.*, 1981), haemangiopericytoma (Ballard and Yarington, 1981) and hamartoma (Engelking, 1959) are also exceedingly rare lesions. One case of plasma cell granuloma (Satomi *et al.*, 1991) has been reported recently in the subglottic region of a 55-year-old woman and perhaps is closely related to the present case. Fibromas and fibromyxomas reported earlier may have some similarity to the present case because of the common morphological features of fibroblastic proliferation and myxoid areas also seen in inflammatory pseudotumour, however, the inflammatory component is not a major feature of fibromas and fibromyxomas.

This is the first case report to our knowledge of an inflammatory pseudotumour of the trachea in a child. This entity should be kept in mind during the differential diagnosis and evaluation of rare mesenchymal tracheal tumours. In difficult cases immunohistochemistry combined with careful morphological assessment may be exceedingly helpful in excluding other entities such as sarcomatoid carcinoma, (which is usually cytokeratin positive) and myxoid leiomyosarcoma (which will usually have a fully developed myoid differentiation exhibiting strong reactivity for desmin and smooth muscle actin and longitudinal cross striations). An inflammatory pseudotumour will tend to have a distinct fine vascularity, variable cellularity and a prominent inflammatory component. A rhabdomyosarcoma can be excluded in addition to the immunohistochemical finding by the lack of rhabdomyoblasts and a cambium layer. These are usually seen in embryonal rhabdomyosarcoma.

https://doi.org/10.1017/S0022215100127604 Published online by Cambridge University Press

Acknowledgement

We are grateful to Ms Shazleen Sadruddin for her secretarial assistance.

References

Anthony, P. P., Telesinghe, P. U. (1986) Inflammatory pseudotumour of the liver. *Journal of Clinical Pathology* **39:** 761–768.

Bahadori, M., Liebow, A. A. (1973) Plasma cell granulomas of the lung. *Cancer* **31:** 191–208.

Ballard, R. W., Yarington, C. T. (1981) Hemangiopericytoma of the tracheal wall. *Archives of Otolaryngology* **107:** 558–560.

Bates, T., Hull, O. H. (1958) Histiocytoma of bronchus: report of a case in a 6-year-old child. *American Journal of Diseases of Children* **95:** 53–56.

Cotelingam, J. D., Jaffe, E. S. (1984) Inflammatory pseudotumour of the spleen. *American Journal of Surgical Pathology* **8:** 375–380.

Day, D. L., Sane, S., Dehner, L. P. (1986) Inflammatory pseudotumour of the mesentery and small intestine. *Pediatric Radiology* **16:** 210–215.

Dubilier, L. D., Bryant, L. R., Danielson, G. K. (1968) Histiocytoma (fibrous xanthoma) of the lung. *American Journal of Surgery* **115:** 240–246.

Eimoto, T., Yanaka, M., Kurosawa, M., Ikeya, F. (1978) Plasma cell granuloma (inflammatory pseudotumour) of the spinal cord meninges. *Cancer* **41:** 1929–1936.

Engelking, C. F. (1959) Hamartoma of the trachea: report of a case, with a review of the literature of benign tracheal neoplasms. *Laryngoscope* **69:** 1278–1286.

Fish, A. E., Brodey, P. A. (1976) Plasma cell granuloma of the kidney. *Urology* **8:** 89–91.

Gilbert, J. G., Mazzarella, L. A., Feit, L. J. (1953) Primary tracheal tumours in the infant and adult. *Archives of Otolaryngology* **58:** 19.

Harris, P. F., Maness, G. M., Ward, P. H. (1967) Leiomyoma of the larynx and trachea: case reports. *Southern Medical Journal* **60:** 1223–1227.

Hartman, G. E., Shochat, S. J. (1983) Primary pulmonary neoplasms of childhood: a review. *Annals of Thoracic surgery* **36:** 108–119.

Horovitz, A. G., Khalil, K. G., Verani, R. R., Guthrie, A. M., Cowan, D. F. (1983) Primary intratracheal neurilemmoma. *Journal of Thoracic Cardiovascular Surgery* **85:** 313–320.

Liew, S. H., Leong, A. S. Y., Tang, H. M. K. (1981) Tracheal paraganglioma: a case report with review of literature. *Cancer* **47:** 1387–1393.

MacLachlan, R. F. (1968) Tracheal fibroma in a child. *Journal of Laryngology and Otology* **82:** 565–570.

Mandelbaum, I., Brashear, R. E., Hull, M. T. (1981) Surgical treatment and course of pulmonary pseudotumour (plasma cell granuloma). *Journal of Thoracic and Cardiovascular Surgery* **82:** 77–82.

Mikaelian, D. O., Cohn, H., Israel, H., Jabourian, Z. (1984) Granular cell tumour of the trachea. *Annals of Otology, Rhinology and Laryngology* **93:** 457–459.

Nochomovitz, L. E., Orenstein, J. M. (1985) Inflammatory pseudotumour of the urinary bladder. Possible relationship to nodular fasciitis. *American Journal of Surgical Pathology* **9:** 366–373.

Olmedo, G., Rosenberg, M., Fonseca, R. (1982) Primary tumours of trachea: clincoco-pathologic features and surgical results. *Chest* **81:** 6, 701–706.

Pearson, F. G., Todd, T. R. J., Cooper, J. D. (1984) Experience with primary neoplasms of the trachea and carina. *Journal of Thoracic and Cardiovascular Surgery* **88:** 511–518.

Perrone, T., de Wolf-Peeters, C., Frizzera, G. (1988) Inflammatory pseudotumour of lymph nodes. A distinctive pattern of nodal reaction. *American Journal of Surgical Pathology* **12:** 351–361.

Pettinato, G., Manivel, C., Rosa, N. D., Dehna, L.P. (1990) Inflammatory myofibroblastic tumour (plasma cell granuloma). Clinicopathologic study of 20 cases with immunohistochemical and ultrastructural observations. *American Journal of Clinical Pathology* **94:** 538–546.

Pollak, E. R., Naunheim, K. S., Little, A. G. (1985) Fibromyxoma of the trachea. A review of benign tracheal tumours. *Archives of Pathology and Laboratory Medicine* **109:** 926–929.

Sandstrom, R. E., Proppe, K. H., Trelstad, R. L. (1978) Fibrous histiocytoma of the trachea. *American Journal of Clinical Pathology* **70:** 429–433.

Satomi, F., Mori, H., Ogasawara, H., Kumoi, T., Uematsu, K. (1991) Subglottic plasma cell granuloma: report of a case. *Auris-Nasus-Larynx* **18(4):** 391–399.

Soga, J., Saito, K., Suzuki, N., Sakai, T. (1970) Plasma cell granuloma of the stomach. A report of a case and review of the literature. *Cancer* **25:** 618–625.

Tang, T. T., Segura, A. D., Oechler, H. W., Harb, J. M., Adair, S. E., Gregg, D. C., Camitta, B. M., Franciosi, R. A. (1990) Inflammatory myofibrohistiocytic proliferation simulating sarcoma in children. *Cancer* **65:** 1626–1634.

West, S. G., Pittman, D. L., Coggin, J. T. (1980) Intracranial plasma cell granuloma. *Cancer* **46:** 330–335.

Wu, J. P., Yunis, E. J., Fettenman, G., Jaeschke, W. F., Gibert, E. F. (1973) Inflammatory pseudotumour of the abdomen: plasma cell granulomas. *Journal of Clinical Pathology* **26:** 943–948.

Address for correspondence:
Dr Farrukh Aijaz, M.D., M.R.C.Path.,
Department of Pathology,
The Aga Khan University Hospital,
PO Box 3500,
Karachi 74700,
Pakistan.

Fax: 4934294