FIBRO-INFLAMMATORY PSEUDOTUMOUR IN THE MAXILLARY SINUS

T H Foo, W T Poh

ABSTRACT

We describe a rare case of fibro-inflammatory pseudotumour in the right maxillary sinus of a 13-year-old Chinese girl who presented with proptosis and radiological appearance suspicious of malignancy but was histologically benign. She underwent Caldwel-Luc operation and received prolonged steroid therapy. She is now free of the disease both clinically and radiologically 33 months after cessation of therapy.

Keywords: *reactive fibroinflammatory pseudotumour, systemic corticosteroid, head and neck.*

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INTRODUCTION

Benign or reactive fibroinflammatory disease that simulate malignancy clinically have been described in the genito-urinary tract and other sites[1-3]. During the last two decades, there have been increasing reports of similar lesions affecting the head and neck. To date, the only large series with 12 of such cases was reported in 1986 by Olsen et al[4]. We report the first local case of such a condition affecting the maxillary sinus.

CASE REPORT

LHH, a 13-year-old Chinese school girl first presented in June 1987 with proptosis of the right eye and difficulty in reading for one month. She was seen by an ophthalmologist in Singapore General Hospital. The ophthalmic findings were right unilateral exophthalmos and normal fundoscopy of both eyes. No bruit was audible over the affected eye. The visual acuity was 6/6 in the right eye and 6/9 in the left eye. Plain sinus X-ray showed complete opacification of the right maxillary antrum and thinning of the antral walls (Fig 1).

The patient was referred to an otolaryngologist. Further history revealed that she had occasional greenish, mucoid rhinorrhea from her left nostril but there was no complaint of facial pain or fever. There was no history of irradiation to the head and neck. Otolaryngologic examination with the aid of the flexible fibreoptic nasopharyngoscope showed edematous nasal mucosa and turbinates in her right nasal passage. No mucopus or nasal polyps were seen in the right middle meatus. The nasopharynx and left nasal passage were clear. Her dentition and roof of the oral cavity were normal. There was no right facial swelling or tenderness. The right maxillary antrum failed to transilluminate.

The axial and coronal CT scans of the right orbit and maxillary antrum showed a large, expansile soft tissue mass filling the right maxillary antrum and involving the adjacent ethmoidal air cells. The sphenoidal sinus was uninvolved. The mass had eroded through the medial and anterolateral walls of the antrum (Fig 2a). The coronal view showed extension of the tumour superiorly through the orbital floor (Fig 2b).

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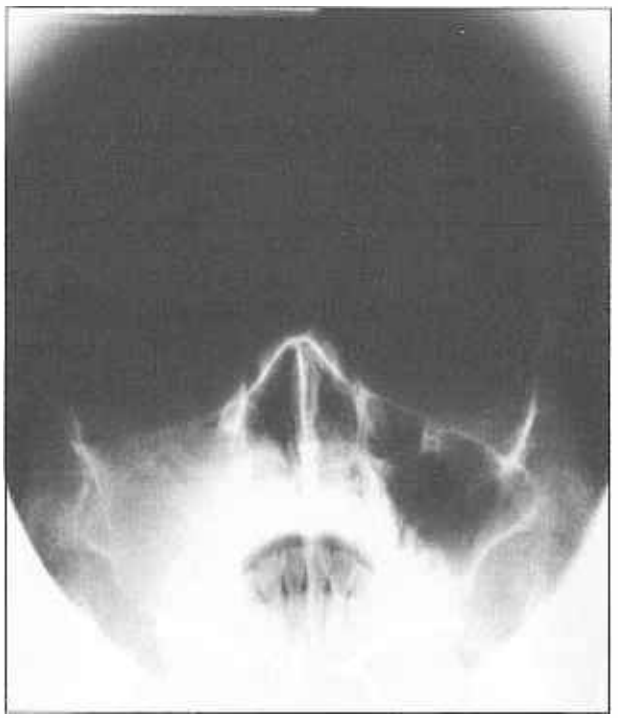
An exploratory right Caldwel-Luc operation was done. The friable mass was removed from the right maxillary antrum and adjacent ethmoidal cells. Antrostomy for drainage was performed.

Fig 1 – Plain sinus X-ray showing complete opacification of the right maxillary antrum and thinning out of its bony walls at presentation (June 1987)



Pathologic Findings

The gross specimen comprised of multiple pieces of soft white tissue with foci of haemorrhages. Microscopically, it was composed of sheets and fascicles of spindle-shaped fibroblasts with interspersed infiltrate of plasma cells and lymphocytes (Fig 3a). There was no storiform pattern. Most of the fibroblasts had bland cytologic features but some showed cytologic atypia with enlarged nulcei, prominent nucleoli and occasional multinucleation (Fig 3b).

These features taken in conjunction with the clinical and radiological features raised the possibility of a malignant soft tissue tumour. However, the generally bland cytology, absence of mitosis, the overall pattern and the heavy inflammatory infiltration were consistent with a reactive inflammatory fibroblastic proliferation. The fibroblastic cells were

immunoreactive for vimentin and negative for desmin, myosin and myoglobin. The Gomori Methenamine Silver, Ziehl Neelsen and Gram stain failed to demonstrate fungi, acid fast bacilli and other organisms. The provisional histological diagnosis was a reactive fibroinflammatory lesion. In view of the diagnostic problems, the slides were sent in consultation to Dr Dennis Heffner (Armed Forces Institute of Pathology, Washington DC) and Dr Richard Kempson (Stanford University Medical Center, California). Both of them agreed that it is an atypical reactive lesion and not neoplastic.

Fig 2a – CT scan (axial) showing a large expansile tumour in the right maxillary antrum eroding its medial and anterolateral walls at presentation (June 1987)

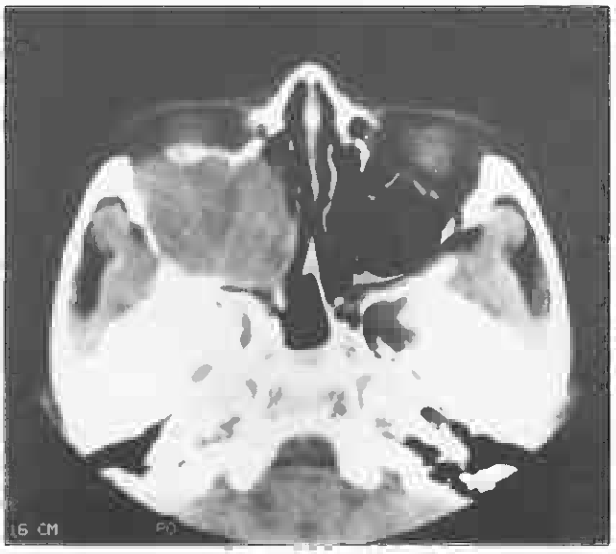


Fig 2b – CT scan (coronal) showing the extension of of tumour superiorly into the right orbit (June 1987)

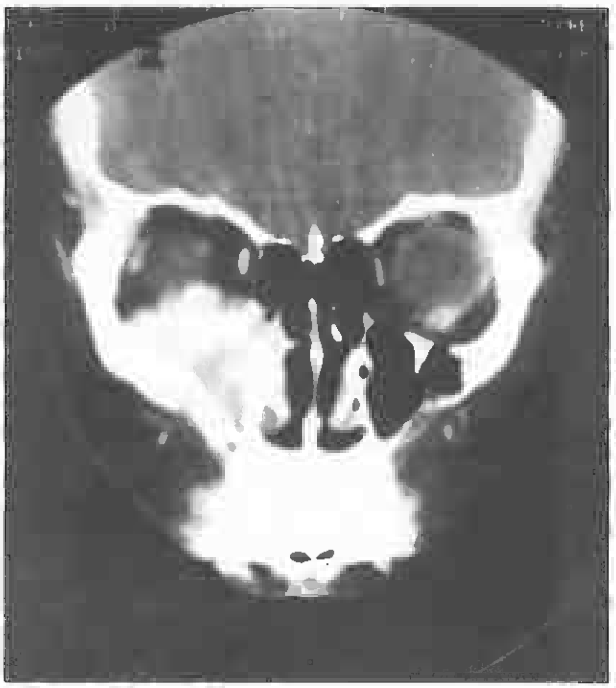


Fig 3a – Fibroinflammatory lesion with spindle-shaped fibroblasts and infiltration of plasma cells and lymphocytes. (H & E x 180)

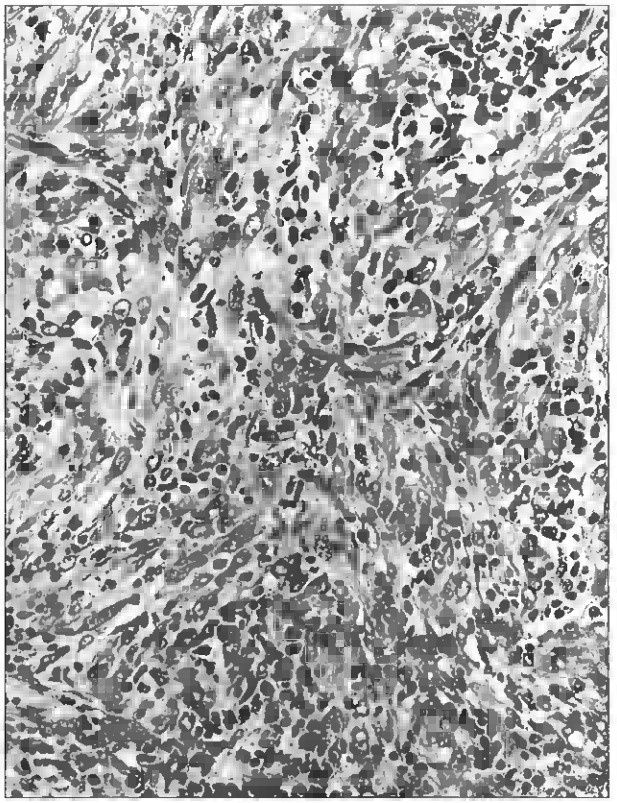
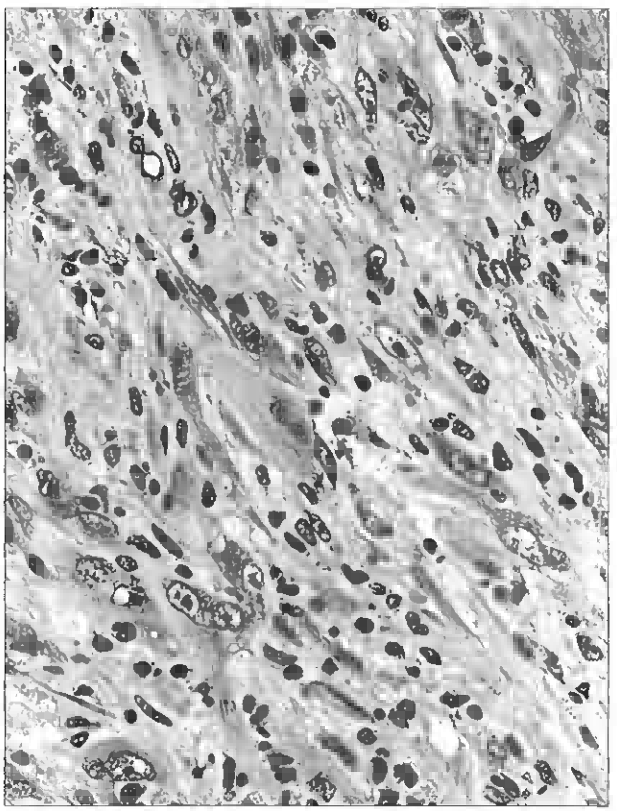


Fig 3a – Atypical fibroblasts with enlarged nuclei and prominent nucleoli (H & E x 280)



Treatment and follow-up

Routine haemotologic and biochemical investigations revealed no abnormality. The erythrocyte sedimentation rate was 9 mm/1 hr. Screening for autoimmune markers showed absence of LE (lupus erythematosus) cells and rheumatoid factor. There was also absence of antibodies against nuclear antigens, smooth muscle and mitochondria. The chest radiograph was normal.

She was started on prednisolone 20 mg tds on 25th September 1987. Towards the end of 3 months of steroid, she developed cushingnoid features. Prednisolone was gradually tailed to 5 mg e.o.d. on 22nd December 1987. Follow up CT scans in September and December 1987 showed no further enlargement of the soft tissue shadow in the right maxillary sinus.

The patient defaulted treatment after a review in June 1988 and was next seen on 6th March 1991. She had remained well for those 33 months without treatment. CT scans done in March 1991 showed a normal right orbit and maxillary antrum (Figs 4a and 4b).

DISCUSSION

Proliferative fibroblastic lesions presenting as tumour-like conditions at different sites of the body have caused much concern to pathologists and clinicians with regard to diagnosis and management[5,6]. The facultative fibroblastic activities of mesenchymal cells like histiocytes, lipoblasts, rhabdomyoblasts, synovial cells and endothelial cells have caused difficulties in determining the histogenesis of these lesions[7].

Fibroblastic lesions comprise a wide spectrum of pathological conditions ranging from stubborn, recurring keloids to tumour-like lesions of such bizarre histological appearance that can sometimes be misdiagnosed as sarcoma. Fibro-inflammatory pseudotumour is a specific entity described in the 1991 WHO classification[8] on histological typing of tumours of the upper respiratory tract. In 1983 Wold and Weiland[9] had given a term "Tumefactive Fibroinflammatory Lesion" when they described 7 patients with fibrosclerosing lesions in the head and neck. The common feature in these tumours is that they appear clinically malignant but are histologically benign; they are locally destructive, often presenting as tumour-like lesions in the

Fig 4a – CT scan (axial) showing a normal right maxillary antrum after Caldwel-Luc operation and steroid (March 1991)

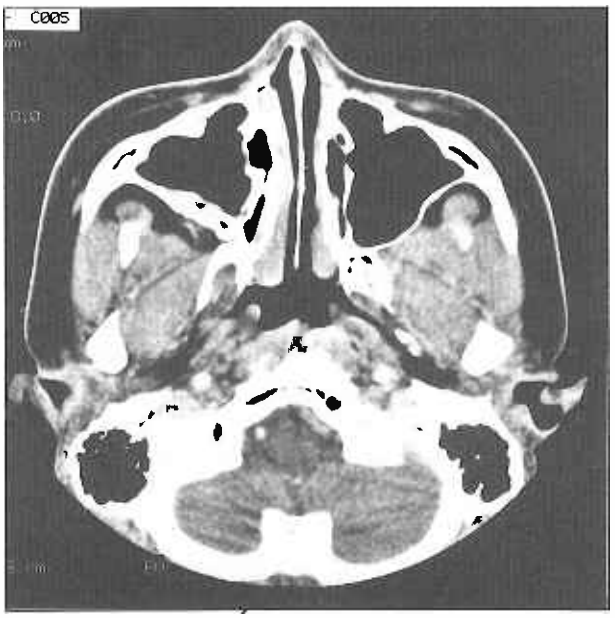
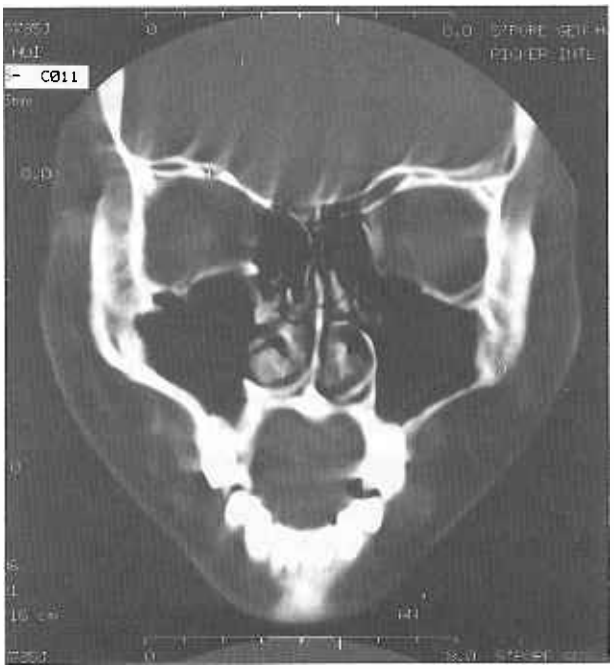


Fig 4b – CT scan (coronal) showing no evidence of tumour in the right orbit and maxillary antrum after treatment (March 1991)



lateral neck compartment, face, oral cavity, tongue, nasal cavity, paranasal sinuses, nasopharynx and orbit.

A search of the literature showed that fibro-inflammatory pseudotumour in the head and neck is a rare condition. The series of twelve patients described by Olsen et al[4] indicated neither sex nor racial preponderance. The age group affected ranged between 33 to 71 years. This case we reported is one of the youngest so far.

The aetiology of fibro-inflammatory pseudotumour in the head and neck is unknown. However the lesion has been considered to be part of the spectrum of 'idiopathic inflammatory fibrosclerosing' syndrome which includes retroperitoneal fibrosis, sclerosing cholangitis, mediastinal fibrosis, Reidel's thyroiditis and orbital pseudotumour[4,10,11]. Retroperitoneal fibrosis is known to be associated with long term usage of methysergide for migraine prophylaxis[12]. There may also be a genetic predisposition. Comings et al[11] reported multifocal involvement of fibrosclerosing lesions in the mediastinum, retroperitoneum, orbit and thyroid in two brothers from a consanguineous marriage.

The presenting signs and symptoms vary according to the sites of involvement. They are the results of the pressure and compression effects of the lesion on adjacent vital structures. The resultant head and neck complaints include pain, stridor from trachea compression, dysphagia and hoarseness from involvement of the vagus nerve etc. Radiological findings of this lesion often show obliteration and destruction of the normal anatomy strongly suggestive of a malignant process. This is exemplified by the CT scans of our patient.

The pathologic findings described by Wold and Weiland[9] consist of proliferating fibrous tissue, lymphocytes, and scattered polymorphonuclear leucocytes. The fibrous proliferation in some areas show a collagenised or hyalinised stroma. Our case differed slightly from their description in that the inflammatory infiltrate consisted of lymphocytes and plasma cells with very few neutrophils. There was neither collagenised nor hyalinised stroma. In addition, the fibroblasts in our case showed nuclear

atypia, a feature that has not been highlighted before. Despite these differences, the lesion we described can be distinguished histologically from fibromatosis which is more cellular and lacks the inflammatory component, and from pseudosarcomatous proliferative lesion such as nodular fasciitis which also generally lacks an inflammatory infiltrate and has prominent mitotic activity[13].

The treatment and prognosis of this lesion in the head and neck depends on the site of involvement and extent. As this lesion tends to recur, adequate surgery in the head and neck can be restricted by the constraints of vital structures and access (eg skull base and root of the neck). As it is a benign tumour histologically. disproportionate radical surgery can result in disfigurement and is therefore unwarranted. There is a case reported of such a lesion in the lateral compartment of the neck who survived with minimal disability for 24 years without any treatment[14]. However, extensive disease in the face, orbit and neck has at least caused death in one of the 12 patients reported by Oslen et al[4].

Corticosteroid has been used successfully in the treatment of retroperitoneal fibrosis[10] and orbital pseudotumour, and it has also been reported to be effective in the treatment of fibroinflammatory tumours of the head and neck. This supports the belief that it may be part of the "idiopathic inflammatory fibrosclerosing" syndrome[11].

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