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Pathology in Focus

Inflammatory myofibroblastic tumour of the tonsil

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Abstract
Inflammatory myofibroblastic tumours are aetiologically enigmatic, nosologically confusing and biologically unpredictable lesions. The lungs are the organs of apparent predilection. These tumours have also been documented in a number of extrapulmonary sites including the head and neck. So far only two cases of inflammatory myofibroblastic tumour of the tonsil have been reported in the English literature. We document another case, occurring in a 41-year-old man with history of cadaveric renal transplant nine years ago. A comprehensive review of the literature is also presented.

Key words: Tonsil; Granuloma, plasma cell

Introduction

Inflammatory myofibroblastic tumour (IMT) is a distinctive pseudosarcomatous inflammatory lesion that occurs in the soft tissue and the viscera of children and young adults. It has a distinctive histological appearance, a benign course with multifocal lesions or recurrence in a proportion of cases, and a disputed nosology (Coffin *et al.*, 1995). Its unknown cause and morphological features have led to several synonyms in the literature such as inflammatory pseudotumour, plasma cell granuloma, plasma cell pseudotumour, xanthomatous pseudotumour, pseudosarcomatous myofibroblastic proliferation and inflammatory myofibroblastic proliferation. IMT occurs most commonly as a solitary lesion in the lung. Extrapulmonary forms have a preference for the abdomen, head and neck, and central nervous system (Treissman *et al.*, 1994). IMT of the head and neck have been reported to occur mostly in the orbit and upper aerodigestive tracts (larynx, oral cavity, oropharynx, oesophagus, and paranasal sinus), but also in the major salivary glands and the parapharyngeal and pterygomaxillary spaces (Batsakis *et al.*, 1995). Only two cases of IMT of the tonsil have been reported in the English literature (Weilbaecher and Sarma, 1984; Newman and Shinn, 1995). We report the third case of tonsillar IMT.

Case report

A 41-year-old man was referred by a nephrologist to our clinic with a chief symptom of 'lump in the throat' with localization to the left side and slight odynophagia for the previous two months. There was no complaint of fever or earache. He had a past medical history of chronic renal failure for which he had received a cadaveric renal transplant nine years ago. He was also known to have had hepatitis C virus (HCV) hepatitis for one year, based on serology and histopathology. He had been on prednisone 5 mg o.d., Azathioprine 100 mg o.d., cyclosporin 75 mg b.i.d. and enalapril 10 mg o.d. since the renal transplant. Clinical examination of his ear, nose and throat revealed a $2 \times 2 \times 2$ cm firm mass with a broad base arising from the left tonsil without any evidence of cervical lymphadenopathy. The rest of the ear, nose, and throat examination was normal. A complete blood count, prothrombin time, partial thromboplastin time, and renal profile were normal. Hepatic profile showed mildly elevated alanine aminotransferase (ALT) and aspartate aminotransferase (AST). The possibility of a neoplastic process and the need for a definitive biopsy was discussed with the patient. He had a tonsillectomy performed without any complication. He recovered with resolution of his symptoms by two weeks after his surgery. It is currently 10 months after surgery without evidence of recurrence.

Pathology

The left tonsil measured $2.5 \times 1.2 \times 0.7$ cm with a roughly spherical, nodular, cream-grey mass 2.2 cm in diameter arising from the medial aspect. On sectioning the cut surface of the mass was cream-grey with yellow streaks. Sections from the mass showed proliferation of spindle cells in a background of inflammatory cells (Figure 1). In some areas the inflammatory cells were sparse and spindle cell proliferation was more compact with a focal storiform pattern (Figure 2). The spindle cells had plump to vesicular nuclei. Immunohistochemistry showed positivity of the spindle cells for vimentin, and smooth muscle actin, and electron microscopy confirmed the myofibroblastic nature of the spindle cells. The right tonsil measured $2.9 \times 1.6 \times 0.8$ cm and histology was normal.

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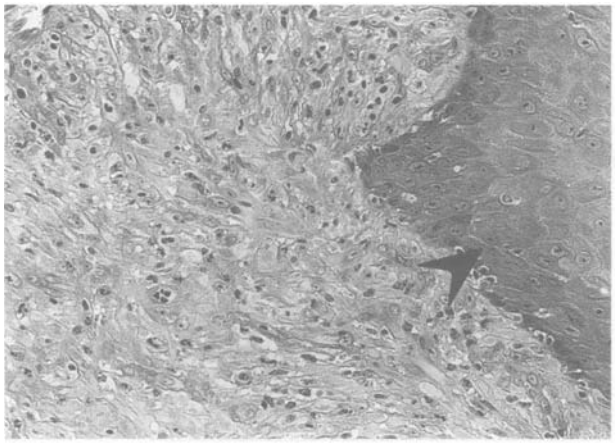

FIG. 1

Inflammatory myofibroblastic tumour – spindle cell proliferation in a background of inflammatory cells is seen. The squamous epithelium of the tonsil is marked with an arrow (H & E stain; × 250).

Discussion

IMT is an uncommon lesion. It is a pathologic diagnosis of exclusion and involves clinicopathological correlation (Hytrioglou *et al.*, 1992). The differential diagnosis of IMT is wide because of the variegated histological picture of the lesion. Pseudosarcomas (nodular fascitis), fibromatosis, fibrous histiocytoma and malignant fibrous histiocytoma as well as other spindle cell sarcomas come in the differential diagnosis.

As is usually the case with IMT, the clinical presentation in our case was suggestive of a neoplastic process.

IMT of the tonsil is extremely rare. An extensive search of the English literature of the last 30 years revealed only two cases. Weilbaecher and Sarma (1984) reported IMT of the tonsil in a 63-year-old man. He had a tonsillectomy but no follow-up was mentioned. The authors chose the term plasma cell granuloma. Newman and Shinn (1995) reported IMT of the tonsil in a 62-year-old woman. She had tonsillectomy without any evidence of recurrence 16 months after surgery. The authors preferred to use the term inflammatory pseudotumour.

In a series of 84 cases of extrapulmonary IMT, two cases arising from the oropharynx were mentioned without any further qualification of exact anatomical location (Coffin *et al.*, 1995).

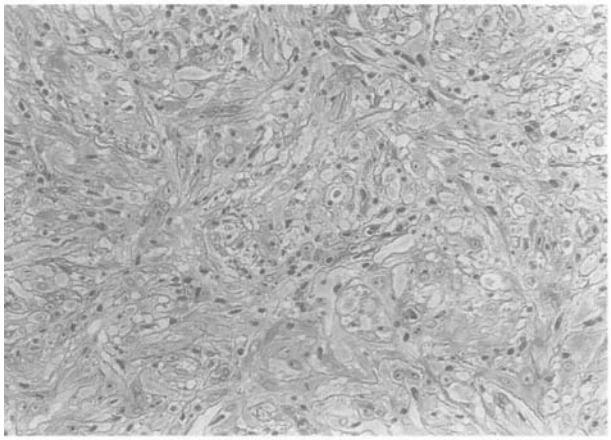

FIG. 2

Inflammatory myofibroblastic tumour – the spindle cells are forming a storiform pattern focally (H & E stain; × 250).

Although the cause of IMT remains unknown, extensive morphological and immunophenotypic studies support the classification of IMT as an inflammatory process rather than a neoplastic one (Facchetti *et al.*, 1990; Davis *et al.*, 1991).

The role of cytokines, particularly interleukin-6 (IL-6) in pathogenesis and the possibility for a specific therapeutic approach has been described (Coffin *et al.*, 1995). It is of interest that an IMT developed in our patient while he was taking long-term prednisone and other immunosuppressants following cadaveric renal transplant nine years ago. The patient with IMT of the tonsil reported by Newman and Shinn (1995) had been on long-term prednisone for asthma and retroperitoneal fibrosis.

A histologically similar lesion, inflammatory fibrosarcoma, has also been described, with a reported metastatic rate of 11 per cent (Meis and Enzinger, 1991). Inflammatory fibrosarcoma and IMT are designated synonymously in the World Health Organization (WHO) classification of soft tissue tumours, with the caveat that 'it is uncertain whether lesions involving multiple sites represent multifocal disease or distant metastasis' (Spencer, 1984). According to Batsakis *et al.* (1995) IMT is of probable neoplastic origin and should be named either IMT or inflammatory fibrosarcoma.

None of the 84 cases of extrapulmonary IMT in the series by Coffin *et al.* (1995) had metastasis documented clinically or pathologically; of the 12 with recurrences, six were successfully treated with excision, one had regression over a six-month period, four were alive with persistent localized disease, and one died of bowel obstruction following recurrence. It has been mentioned that regardless of the site of origin, IMT has a predilection for younger patients, and extrapulmonary IMT shares some clinical and many morphological similarities with the prototypical pulmonary plasma cell granuloma or inflammation (Coffin *et al.*, 1995). Extrapulmonary IMT affects a younger population of patients, with a predilection for the first and the second decades in contrast to a peak incidence in mid-adulthood for the pulmonary form (Spencer, 1984; Pettinato *et al.*, 1990). On the other hand our patient was in his fifth decade, the other two patients of IMT of the tonsil described previously in the literature were in their seventh decade.

The clinical presentation and gross pathologic features of both (pulmonary and extrapulmonary) forms of IMT may mimic a malignancy, yet most tumours behave in a non-aggressive fashion. Although the most common clinical presentation of IMT is an incidentally discovered mass in the chest or abdomen, a minority of cases (15–30 per cent) may be associated with unexplained fever, weight loss, microcytic hypochromic anaemia, thrombocytosis, polyclonal hyperglobulinaemia and elevated erythrocyte sedimentation rate (Coffin *et al.*, 1995). With resection of the pulmonary or extrapulmonary mass, there is resolution of constitutional symptoms and laboratory abnormalities in a few days or weeks, and reappearances of the clinical syndrome heralds the presence of another mass (Tang *et al.*, 1990; Souid *et al.*, 1993).

Treatment of IMT may be divided into surgical and medical on the basis of the site of the lesion and potential for functional impairment (Newman and Shinn, 1995). For this condition, excisional biopsies have been performed primarily for diagnosis, and they have turned out to be curative (Newman and Shinn, 1995). In cases in which significant morbidity could result from surgical treatment, trials of steroids with potential addition of radiotherapy have been described (Sclafani *et al.*, 1993).

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IMT is a well-documented entity that can occur at many sites within the head and neck. As is usually the case with IMT, the clinical presentation in our patient was suggestive of a neoplastic process.

Surgery is curative in cases in which excision can be performed, with acceptable morbidity to the patient.

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