Case Report

**Benign inflammatory myofibroblastic tumour of the nose: a rare case report**


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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) of the head and neck region are rare and benign and of unknown etiology. They mimic as malignant lesions both clinically and radiologically. It encompasses a spectrum of myofibroblastic proliferation along with varying amount of inflammatory infiltrate. Recently, the concept of this lesion being reactive has been challenged based on the clinical demonstration of recurrences and metastasis and cytogenetic evidence of acquired clonal chromosomal abnormalities. Here we report a case of a 16 year old male who presented with a swelling on the nasal dorsum for 6 months. MRI and FNAC revealed a spindle cell neoplasm. The swelling was resected completely under general anaesthesia. Histopathology with immunohistochemistry proved it to be a benign inflammatory myofibroblastic tumour. Complete surgical resection is the best treatment modality for IMT.

Keywords: Benign, Rare, Spindle cells, Surgical excision

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) was first described by Bunn in 1939 when he observed it in the lungs. It was named as IMT by Umiker et al as it was similar to a malignant neoplasm clinically, radiologically and histopathologically. Various pathogenesis have been proposed as initiating factors such as reactive, infections, autoimmune and neoplastic, but the etiology of most of them remains unknown. Recently IMTs being described as reactive has been challenged based on the clinical demonstration of recurrences and metastasis and cytogenetic evidence of acquired clonal chromosomal abnormalities. The aero-digestive tract, major salivary glands and the soft tissues of the neck are the most common sites for IMT. In the head and neck, the epiglottis, endolarynx, parapharyngeal spaces, maxillary sinus, submandibular region and oral cavity are the common sites. IMT in the faciomaxillary region are rare and represent 14–18% of all extra-pulmonary IMTs. The diagnosis is difficult but confirmed by the histologic examination of the lesions. In the oral cavity, IMTs have been reported in varied locations like gingiva, tongue, hard palate, mandible, buccal mucosa and submandibular salivary gland. Clinically, they present as firm, painless, mass or swelling of short duration and are generally asymptomatic or following specific symptoms related to the site of origin. They usually affect children and young adults. Computed tomography (CT) scan and magnetic resonance imaging (MRI) of IMTs in the head and neck region are nonspecific but may suggest infiltration, aggressive growth or granulomatous disease. IMTs of head and neck are generally benign lesions and usually cured by radical excision, steroids, irradiation and/or chemotherapy. CO₂ laser is the latest in the treatment modality.
CASE REPORT

A 16 year old male presented to our department with a swelling over the dorsum of nose for about 6 months. He was apparently normal 6 months ago when he noticed a small pea sized swelling which was spontaneous in onset and has gradually progressed to the present size (Figure 1).

Figure 1: Pre-op photograph (A) front and (B) lateral views.

He complained of occasional pain for the past 1 month, but no history of trauma, bleeding or nasal block. On examination, a 2 cm × 1 cm firm non-tender swelling was present over the dorsum of the nose just above the supratip break region. The overlying skin was free and the swelling had limited mobility. There was no regional lymphadenopathy. A clinical diagnosis of a soft tissue tumour was made. MRI of the face was done which showed a well-defined homogenous T2/STIR hyperintense lesion in the subcutis plane of the supratip region of the nose measuring 15 × 6 × 9 mm occupying the midline and bilateral paramedian planes, with mild scalloping of the underlying cartilages. There were no internal T1 hyperintense components. The MRI suggested a benign lesion and to correlate with histopathology for confirmation (Figure 2).

Figure 2 (A and B): MRI showing a well-defined homogenous T2/STIR hyperintense lesion in the subcutis plane of the supratip region of the nose with no internal T1 hyperintense components.

FNAC smears showed a predominantly scattered with a few clusters of spindle cells with occasional inflammatory cells in a bluish chondroid background with no atypia or necrosis suggestive of a benign spindle cell neoplasm. We proceeded for surgical excision. Under general anaesthesia, we used an open rhinoplasty approach to excise the lesion in toto and sent for histopathology (Figure 3).

Figure 3: Lesion after excision.

It showed a lesion composed of spindle cells arranged in sheets, fascicles and storiform pattern. The cells were spindled with scant to moderate cytoplasm and oval nuclei and seen to infiltrate the adjacent skeletal muscle fibres. No increased mitotic activity or necrosis noted. There are areas of hyalinisation and chondromyxoid stromal tissue (Figure 4 and 5).

Figure 4: Section shows a neoplasm composed of spindle cells arranged in sheets, fascicles and storiform patterns (H&E x100).

This report suggested a low grade spindle cell neoplasm probably fibroblastic. Immunohistochemistry revealed smooth muscle actin (SMA) positive, but negative for Ki 67, CD34, S100 and EMA and thus a diagnosis of benign inflammatory myofibrolastic tumour was made (Figure 6 and 7).

Figure 5: This micrograph shows a lesion with storiform pattern (H&E x100).
DISCUSSION

IMT was first reported in 1905 by Birch-Hirschfield and later by Bunn in 1939 and coined IMT by Umiker et al. Its etiology remains unknown. In 1994, the World Health Organization defined IMT as an intermediate soft tissue tumor that is composed of myofibroblast-differentiated spindle cells and accompanied by numerous inflammatory cells, plasma cells, and/or lymphocytes. IMT is an extremely rare tumor in the head and neck region with only few cases that have been reported in different oral cavity sites including intra and extraosseous sites. Differential diagnoses of IMT are very many, the most significant being nodular fasciitis, solitary fibrous tumor and fibrosarcoma. Histologically, IMT is composed of inflammatory cells, histiocytes, and fibroblasts. There are three variants of IMT as classified by WHO which are: (a) loosely organized myofibroblasts in an edematous myxoid background with plasma cells, lymphocytes, eosinophils, and blood vessels, resembling nodular fasciitis; (b) dense aggregates of spindle cells arrayed in a variable myxoid and collagenized background and admixed with a distinctive inflammatory infiltrate, diffuse clusters of plasma cells, and lymphoid nodules, resembling fibrous histiocytoma or fibromatosis; and (c) collagen sheets with scattered plasma cells and eosinophils resembling a scar or desmoid tumor. Cytologic atypia with nuclear pleomorphism and increased mitotic activity are uncommon features and may be associated with malignant transformation. Immunohistochemical staining is usually utilized to confirm the myofibroblastic phenotype of the tumor spindle cells which are typically reactive to vimentin (99%), SMA (92%), muscle-specific actin (89%), and desmin (69%). Spindle cells may be focally positive to epithelial markers such as cytokeratin, epithelial membrane antigen (EMA; 36%) and CD68 (25%). IMTs are classically negative for myoglobin and S100 protein. Inflammatory mediators such as cytokines and interleukin-1 (IL-1) are released in response to an insult causing fibroblastic proliferation, leaky endothelium in a procoagulant state and extravasation of polymorphous cellular infiltrate into the extracellular spaces. Associated clinical findings include fever, tenderness, and erythema over the affected region. The imaging findings of IMT are a mildly enhancing soft-tissue mass without any internal calcification or bone destruction revealed by contrast-enhanced CT or MRI. Complete surgical resection of the lesion has been reported as the principal treatment choice for IMT.

CONCLUSION

IMTs of the head and neck region are relatively rare and have to be kept in mind when working up for an undiagnosed lesion. CT, MRI and FNAC will aid in the diagnosis. Curative solution is surgical excision and subjecting the specimen to a thorough histopathological examination with appropriate immunohistochemical staining to confirm the diagnosis.

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REFERENCES


