

MAXILLARY INFLAMMATORY MYOFIBROBLASTIC TUMORDEEKSHA P RAO¹, KAILESH PUJARY¹, MARY MATHEW², AJAY M BHANDARKAR^{1*}¹Department of Otorhinolaryngology-Head and Neck Surgery, Kasturba Medical College, Manipal University, Manipal, Karnataka, India.²Department of Pathology, Kasturba Medical College, Manipal University, Manipal, Karnataka, India. Email: bm.ajay@gmail.com

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a benign disorder of locally aggressive nature. It is an indolent tumor with a slowly progressive course and varied manifestation presenting with a wide range of clinical manifestations depending on the site of origin. We present a case of IMT in the maxillary sinus presenting with hemifacial pain, oculomotor palsy and ptosis which was successfully treated with endoscopic local excision and oral steroid therapy.

Keywords: Plasma cell granuloma, Maxillary pseudotumor, Maxillectomy, Steroid therapy, Radiotherapy.

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INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a benign disorder of locally aggressive nature [1]. This tumor provides a dilemma in diagnosis due to the bone-eroding properties and drastic clinical presentation, prompting the clinician to favor a diagnosis of malignancy rather than an indolent tumor with a chronically progressive course [1,2]. We present a case of a 38-year-old man who presented with hemifacial pain, oculomotor palsy, and ptosis of short duration which was an atypical presentation of this benign pathological entity.

CASE REPORT

A 38-year-old nondiabetic, nonhypertensive man presented to the outpatient department with the chief complaints of left hemifacial pain for one and a half months and drooping of the left eye of 10 days duration. The left hemifacial pain initially began as throbbing, continuous pain in the left upper first molar which then progressed to the left cheek and the left parietal region. It relieved on taking analgesics. However, there were no aggravating factors. Drooping of the left eye started after an episode of severe throbbing hemifacial pain. There was no history of blurring of vision, diplopia, nausea, vomiting, and fever. There was no history of nasal symptoms. He revealed a history of tuberculous lymphadenitis 10 years ago and was declared cured after a complete course of antitubercular therapy.

Clinical examination revealed swelling in the left maxillary region. Diagnostic nasal endoscopy revealed a bulge in the lateral wall of the left nasal cavity with no evidence of any mass. Pus was visualized in the accessory ostium of the left maxillary sinus. Examination of the eye revealed ptosis of left upper eyelid with sluggish pupillary reactions and oculomotor nerve palsy, however, the vision was 6/6, and corneal reflex was intact. Neurological and other cranial nerve examination revealed no abnormalities. Ear, throat, and neck examination was normal. Systemic examination was normal.

Blood investigations revealed an elevated erythrocyte sedimentation rate of 52 mm/hrs. Contrast-enhanced computed tomography (CECT) of the nose and paranasal sinuses showed opacity in the left maxillary sinus extending to orbital apex and infratemporal fossa. Magnetic resonance imaging (MRI) of the brain and paranasal sinuses revealed an ill-defined altered signal intensity lesion involving the left maxillary sinus with thickening and sclerosis of the posterolateral wall of the

left maxillary sinus with an extension of the lesion into masticator space involving temporalis and lateral pterygoid muscles. Superiorly, the lesion was abutting the floor of the left orbit with extension into extraconal and intraorbital space. Posteriorly, the lesion was involving the greater wing of the sphenoid with intracranial extension and causing pachymeningitis. Focal enhancement at left supraorbital fissure was evident with a small degree of proptosis (Fig. 1a and b).

Diagnostic nasal endoscopy and biopsy taken from the left maxillary sinus were reported as plasma cell granuloma with no evidence of malignancy. Partial endoscopic maxillectomy was done to obtain deeper tissue from the mass for immunohistochemistry and definitive diagnosis.

Histopathological examination of the mass showed features consistent with IMT (Fig. 2a and b).

Ziehl-Neelsen's staining of the specimen did not reveal any evidence of mycobacteria. Immunohistochemistry showed that the tumor cells were positive for vimentin and smooth muscle actin (SMA) and negative for CD34, anaplastic lymphoma kinase, Ki67 (Fig. 3a and b).

Medical therapy with a tapering dose of oral corticosteroids was given over a period of 3 months.

On follow-up after 1½ months, the ptosis of his left eye had significantly reduced, and the maxillectomy cavity appeared healthy. On subsequent follow-up after 3 months, the ptosis had completely resolved (Fig. 4a and b). Follow-up CECT showed a contracted maxillary sinus with no enhancement on contrast.

DISCUSSION

Plasma cell granuloma is a benign, space occupying lesion; that is locally aggressive [1]. In 1990, Yuan *et al.* studied this entity on 20 patients and coined the term IMT, stressing on the fact that it is just a congregation of reactive, nonspecifically differentiated fibroblastic and myofibroblastic cells which mimic malignancy clinically and radiologically and is comprised infiltrates of inflammatory polymorphous cells, thick-walled vessels, and fibrous stroma [2]. The common sites of occurrence are the abdomen and pelvis. However, it can occur in the peritoneum, central nervous system, bone, and uterus [2-6]. In the head and neck region, the most common site of occurrence is the orbit. It can also occur in the true

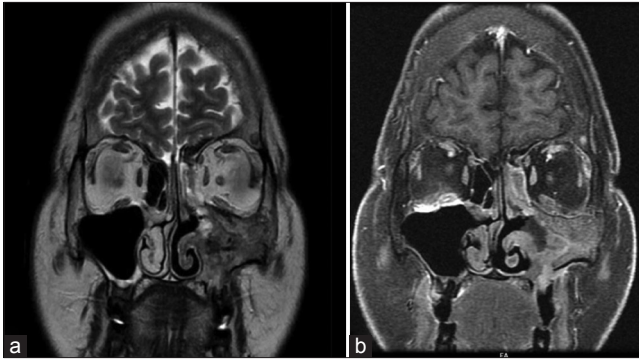


Fig. 1: (a) T1-weighted coronal magnetic resonance image showing altered signal density in the left maxillary sinus extending into the orbit. (b) T2-weighted coronal magnetic resonance image showing altered signal density in the left maxillary sinus extending into the orbit

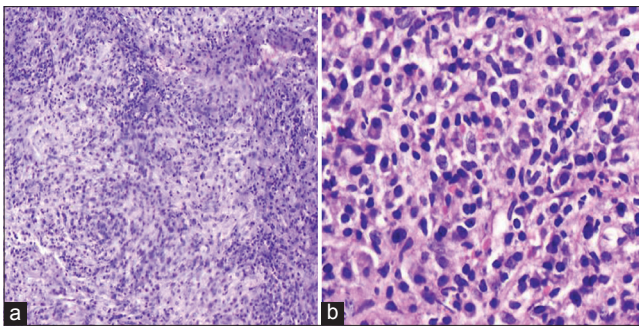


Fig. 2: (a) Proliferating spindle fibroblasts with inflammatory cells (H and E staining, ×100). (b) Plasma cells with intracytoplasmic Russell bodies (H and E staining, ×400)

vocal folds, oral cavity, and oropharynx, nasopharynx, parapharyngeal space, and paranasal sinuses [2,3,5]. It does not occur in a specific age group, although literature reveals a median age group of 47.4 years for the occurrence. The pulmonary variant occurs predominantly in young adults, and the extrapulmonary variant occurs mostly in adolescents and children. It occurs predominantly in females with a ratio of 3:2 [2,3]. Approximately, 30 cases of maxillary sinus plasma cell granuloma have been reported in the literature.

IMT is not a single entity, variations in histological appearance give it some synonyms; namely, plasma cell granuloma, myofibroblastoma, inflammatory pseudotumor, inflammatory myofibrohistiocytic tumor, xanthogranuloma, mast cell granuloma, and xanthoma [2,7,8].

Etiology of IMT is unknown. Multiple theories have been postulated for the development of this tumor. Perrone *et al.* considered adjacent necrosis or infection to be the inciting foci. The role of viral infectious agents such as Epstein-Barr virus and human herpes virus-8 has been discussed in the literature. Autoimmunity has also been considered as an etiological factor. Chronic infection or inflammation leading to differentiation of plasma cells is another theory [1,2,5,7]. In our case, the history of tuberculous lymphadenitis (chronic granulomatous disease), although treated and cured maybe considered as a trigger based on the chronic inflammatory theory.

Clinically, IMT is locally aggressive and causes a varied degree of bone destruction, unlike other benign conditions; therefore, the patient is subjected to multiple biopsies to arrive at this diagnosis. In maxillary sinus involvement, it presents with locoregional features such as nasal obstruction, epistaxis, cheek swelling and facial deformity, numbness due to infraorbital nerve involvement, proptosis and diplopia due to orbital involvement and decreased vision due to optic nerve

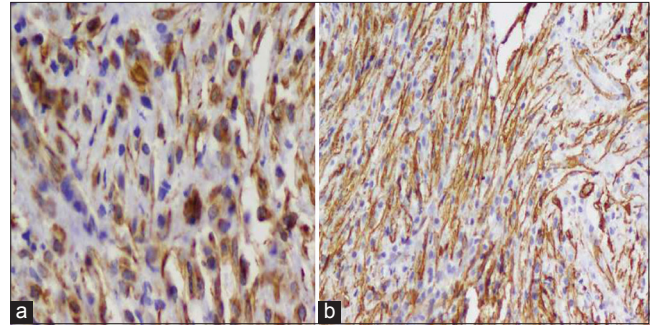


Fig. 3: (a) 400× immunohistochemical staining positive for vimentin. (b) 400× immunohistochemical staining positive for smooth muscle actin



Fig. 4: (a) Pretreatment image showing left maxillary swelling and ptosis. (b) Posttreatment image showing resolution of ptosis after corticosteroid therapy

involvement. Vagus and hypoglossal nerve involvement have been noted with parapharyngeal space involvement. Systemic features such as fever, malaise, and weight loss are reported in patients with thoracic and abdominal involvement and not in head and neck IMT. IMT does not have the property of lymphatic spread although perineural invasion is a known property of IMT. Keen *et al.* suggested perineurofibromatosis and Du Vuysere *et al.* suggested perineural invasion as the causative pathogenesis for neural symptoms of IMT. Our patient presented with hemifacial pain and ptosis which can be attributable to infraorbital nerve and oculomotor nerve involvement by the tumor [1,3,5,6,8,9].

Histopathologically, the characterization of the tumor is based on myofibroblasts and inflammatory cells. Identification of this lesion is by histopathological examination, however, it is a diagnosis of exclusion as this histological picture is very nonspecific and the absence of adequate mitoses and atypia does not characterize it as a neoplastic lesion. Fujii *et al.* have classified the tumor into three subsets based on the varied quantity of myofibroblasts and the inflammatory cells. The lymphoid subset of IMT has lymphocytes predominantly with little fibrosis. The granulomatous subset has an admixture of lymphocytes, histiocytes, plasma cells, eosinophils, fibroblasts, and myofibroblasts. The sclerosing subset predominantly contains fibrotic cells with minimal inflammatory cells. Immunohistochemically, the tumor expresses vimentin and SMA. Antiactin antibody positivity denotes its origin in the muscle. It is immunohistochemically negative to CD68, keratin, desmin, S-100, and caldesmon [1,3,5,6,9].

CT and MRI are not diagnostic of the tumor, only aid in identifying the extent. CECT demonstrates the homogenous or heterogenous appearance of IMT with bony destruction and local invasion with a variably high degree of enhancement on contrast. MRI demonstrates a nonhomogenous

appearance of IMT and an intermediate signal on T2-weighted images due to the paucity of protons in fibrous tissue, high N: C ratio and increased cellularity. The presence of fibrous tissue in IMT is inversely proportional to the intensity on T2-weighted MRI imaging [2,5,6,9,10].

Differential diagnosis of rhinoscleroma, Wegener's granulomatosis, fibrous histiocytoma, fibromatosis, and soft tissue malignancies such as spindle cell sarcoma and myxofibrosarcoma needs to be considered due to the erosive properties of the lesion. Occasionally, the histological appearance of the tumor may resemble plasmacytoma [3,10].

The treatment options depend on the anatomical site and tumor morphology. Lymphoid predominant lesions respond well to radiotherapy, whereas granulomatous lesions to high dose corticosteroids. Sclerosing lesions are more aggressive and show little response to either treatment modality. The initial line of management of IMT is high-dose oral corticosteroids. Endoscopic surgery, in the form of medial maxillectomy, is performed if the response to steroids is poor. This aids in diagnosis by providing biopsy specimens and in clearance of the tumor. Current literature suggests excision of the tumor followed by corticosteroid therapy for 6 months as the best line of management. In our case, we performed endoscopic clearance of the tumor followed by oral corticosteroid therapy which resulted in complete remission of IMT [1,3,5,6,8,9]. Radiotherapy can be employed as therapeutic modality instead of oral corticosteroid therapy and has been known to produce tumor free survival [11].

IMT is a benign, chronically progressive tumor with a good prognosis when treated with combined modality therapy. Literature shows that the following treatment, 80% have remained tumor-free for more than 2 years. Single-modality therapy with surgery/corticosteroids/radiotherapy shows complete remission only in 50% of the cases. However, it has also been noted that radical procedures do not do well when compared to conservative treatment. 25% patients may have a local recurrence following surgical removal and 5% distant spread [1,2,9]. In our case, the patient has been tumor free for 3 months.

CONCLUSION

IMT is a locally erosive benign lesion mimicking malignancy uncommonly occurring in the maxillary sinus. Perineural involvement

is a characteristic feature of IMT. Histology of the tumor contains inflammatory cells and fibroblastic cells. Combined modality therapy with oral corticosteroids and surgical excision is the treatment of choice.

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