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Inflammatory Pseudotumor of Maxilla: Masquerading as Aggressive Malignancy

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Inflammatory pseudotumor is a chronic inflammatory lesion of unknown origin with highly aggressive course. The disease even being benign in nature spreads aggressively, giving a false clinical impression of malignancy. A knowledge of the clinical entity is essential when considering a diagnosis of malignancy.

Keywords: Pseudotumor; tumor; recurrence; chronic inflammation.

1. INTRODUCTION

Inflammatory pseudotumor is a tumor like lesion characterized histologically by fascicles of plump spindle cells in a background of chronic inflammatory cells and plasma cells [1].

Inflammatory pseudotumor was first observed in the lung and described by Brunn in 1939 and was so named by Umiker et al. [2] in 1954 because of its propensity to clinically and radiologically mimic a malignant process. Inflammatory pseudotumor is a quasineoplastic lesion consisting of inflammatory cells and myofibroblastic spindle cells [3,4]. It is a chronic inflammatory lesion of unknown origin. Inflammatory pseudotumor (IPT) are clinicopathologically distinctive but biologically

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controversial entities, which have been described in the lungs, abdomen, retroperitoneum, and extremities, but rarely affect the head and neck region. Clinically it behaves as an aggressive lesion with extensive destruction but considered to be a benign reactive process. Because of its extensive involvement and infiltrative nature, it is confused as a malignancy. Most of them respond very well to corticosteroid and surgical excision. Other synonyms are plasma cell granuloma, histiocytoma, xanthomatous granuloma, inflammatory myofibroblastic tumor, and spindle cell pseudotumor [1].

Though it has a benign clinical course, it is said that at least a subset of inflammatory Pseudotumor (IPTs) represents true neoplasia rather than reactive myofibroblastic proliferation [5].

2. CASE REPORT

A 62 years old female presented to our Outpatient department with left side cheek swelling for 2 months, insidious onset, gradually progressive; and dull aching, mild intensity pain around swelling, that relieved with analgesics. She also had occasional left side nasal obstruction. She has no associated headache, visual impairment, difficulty in swallowing, chewing.

On physical examination examination diffuse, non tender, firm to hard in consistency left side cheek swelling was present, it was also palpable along gingivobuccal sulcus. Eye movements were normal in all directions. No facial weakness, no loose teeth, bulge was detected and mouth opening was normal.

On endoscopic examination pinkish polypoidal mass was seen with smooth surface just coming out of osteo-meatal area, which did not bleed on touch. On contrast enhanced Magnectic resonance imaging of nose and para-nasal sinuses there was soft tissue in left maxillary sinus extending into ostium and erosion of anterolateral wall of left maxillary sinus with mass extending to left cheek.

Patient underwent endoscopic removal of mass from left nasal cavity and maxillary sinus. Mucosa was removed from dehiscent anterolateral wall of maxillary sinus and from soft tissue of cheek. Tissue which was sent for biopsy was reported as chronic inflammatory tissue.

But swelling of left cheek persisted even after surgery. After a brief period, she again developed pain. On repeat endoscopic examination profuse mucopus was seen coming out of antrum, and hypertrophied mucosa of maxillary sinus with small cavity.

She underwent removal of soft tissue by Caldwell- Luc approach after about a month. Tissue was again sent for biopsy. It was reported as inflammatory pseudotumor. Haematoxylin & Eosin stain showed fibroadipose tissue with dense lymphomononuclear cell infiltration along with some lymphoid aggregates. It has a diffuse and cord like arrangement of the infiltrate, composed chiefly of plasma cells and lymphocytes along with occasional histiocytes and neutrophils, separated at places by areas fibrosis and fibroblastic proliferation.

Patient started on oral prednisolone at a dose of 1 mg/kg for 2 weeks and anti- inflammatory medications to which she responded well. Pain and swelling subsided. Prednisolone was tapered to 5 mg/day and now she is completely symptom free and is under regular follow up with us for last 2 years and is completely symptom free.

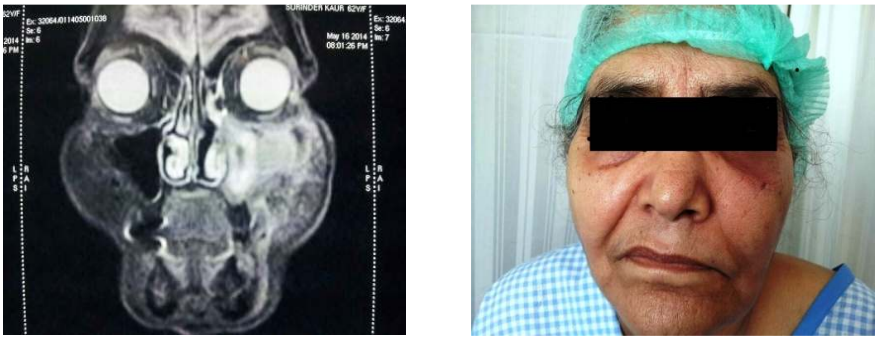


Fig. 1. Coronal section of contrast enhanced MRI of PNS

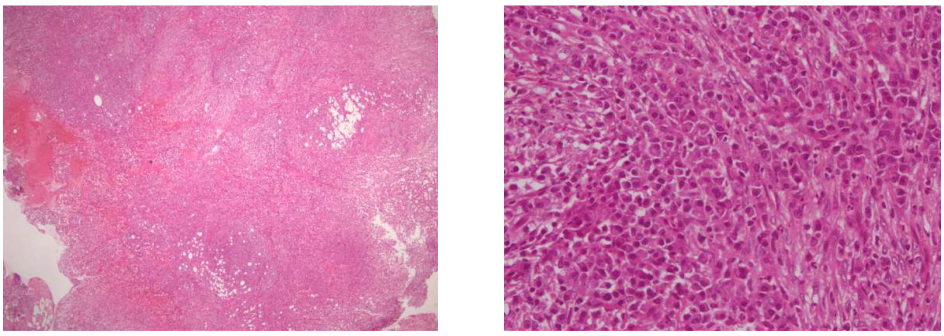


Fig. 2. H & E stain

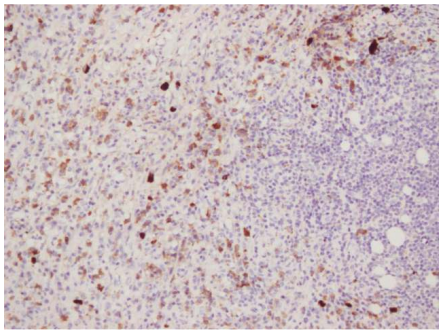
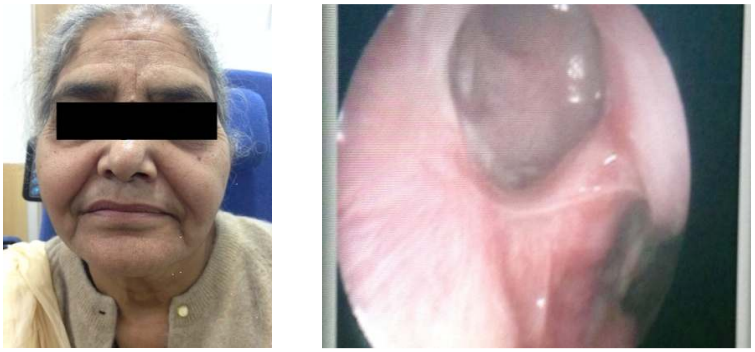


Fig. 3. Staining for kappa showing polyclonal plasma cells



Figs. 4 & 5. Post-operative photograghs showing absence of swelling on the left cheek, absence of any pathology in maxillary sinus on endoscopic examination

3. DISCUSSION

Inflammatory pseudotumor represents a spectrum of myofibroblastic proliferation.

It is reported to occur in nearly every site in the body, most commonly involving the lung and the orbit, and rarely the maxillary sinus [6]. Its diagnosis is usually by exclusion as clinical and histopathology findings are inconsistent. Some are difficult to distinguish from reactive process, whereas others appear sarcomatous on histopathology. Its propensity to be locally aggressive, to frequently be multifocal and to progress to true malignancy supports it being low grade fibrosarcoma. Histology is non- specific, showing two typical cell types: myofibroblasts and inflammatory cell. Myofibroblasts express vimentin in 99% cases, actin in 89-92% cases, desmin in 69% cases, and cytokeratin in 36% cases [7]. Immunohistochemical studies of T- and B-cell subpopulations may be helpful in distinguishing inflammatory pseudotumor from lymphoma.

The exact etiology is not clear. It has been postulated that they might be the result of a post inflammatory repair process, a metabolic disturbance or an antigen-antibody interaction with an agent which is no longer identifiable in aspiration or biopsy material.

Few occurs secondary to infection. Organisms found in association with inflammatory pseudotumor include mycobacteria associated with spindle cell tumor. It has been associated with infections caused by other organisms, including Mycobacterium avium–intracellulare complex, *Corynebacterium equi*, *Escherichia coli*, *Klebsiella*, *Bacillus sphaericus*, Pseudomonas, *Helicobacter pylori* and *Coxiella burnetti* [4,8,9].

Extra orbital inflammatory pseudotumors of head & neck can occur in nasal cavity, nasopharynx, maxillary sinus, larynx and trachea. Perineural spread along maxillary, mandibular, and hypoglossal nerves and a case complicated by internal carotid occlusion have been described [9]. It has no particular age group of presentation. In a review of literature that included 28 cases of mouth & maxillary sinus inflammatory pseudotumors, Vigneshwaran found a found a 1.5:1 male to female ratio & a mean presentation age of 33 years, ranging from 2 to 67 years [10]. It is presented with a variety of symptoms depending on the site of origin. Most frequent clinical picture is a non-specific sinunasal tumor that grows to a stable state over months or years; it may be painful and may be associated with nasal obstruction, nose bleeding, proptosis, increased lymph nodes, dysphagia or cranial nerve dysfunction [11]. Variation in symptoms, edema, erythema and fever may suggest an inflammatory etiology [12]. Systemic symptoms are usually not encountered [11,13].

CT findings usually suggest a more aggressive appearance with associated erosion, remodeling, sclerosis and bone thinning, mimicking malignant tumors [13].

Lab examinations may be altered, such as thrombocytosis, hypochromic microcytic anemia, hypergammaglobulinemia and an increased ESR which resolve following tumor resection [14].

Many a time's multiple biopsies are required to establish a diagnosis of an inflammatory pseudotumor. It is a diagnosis of exclusion, after eliminating the possibility of benign and malignant neoplasias, collagen diseases, vasculitis, infection and other inflammatory diseases [13]. A cytological diagnosis is difficult [15]. Castaneda et al. [16] reported necrotic and inflammatory tissue on histopathology even after taking multiple biopsies. Constantino et al. [14] reported three cases that underwent multiple biopsies in which pathology revealed inflammatory mucosal polyp with non-specific chronic inflammation and ample hemorrhagic, necrotic and hyalinized areas that was suggestive of inflammatory pseudotumor. Lee at al. [17] reported a case in which biopsy specimen showed bundles of fibroblast admixed with inflammatory cells composed of lymphocytes and plasma cells. There was no evidence of mitosis or cellular atypia. This was almost similar to our finding.

Corticosteroids, surgery and radiotherapy have been used in the treatment of inflammatory pseudotumor. Some authors have preferred corticosteroid therapy, while others have chosen surgery as treatment of choice [11,12]. High dose corticosteroid therapy usually yields favorable response; early lesions that include lymphoid follicles tend to be more responsive to corticosteroids, while mature lesions with a more fibrous content respond less [12].

JF Castaneda et al. [16] reported a case of IPT of maxillary sinus in which they attempted surgical removal, but on recurrence they had given intravenous antibiotics and high dose corticosteroids. This resulted in complete disappearance [16]. GTL Constantino et al. [14] reported 3 cases which were cured following complete excision. DT Ginat et al. [18] presented a case series of IPT, which showed complete regression after steroid therapy. JH Lee et al. [17] advocated administration of high-dose corticosteroid as the primary treatment of choice for inflammatory pseudotumor.

Radiotherapy indicated only in patients for whom surgery or corticosteroids is unsuccessful or contraindicated [11].

4. CONCLUSION

Inflammatory pseudotumor should be treated as benign pathology, unless it is showing cellular atypical and/ or frequent recurrence. Because of the unusual clinical presentation, high level suspicion and a scrupulously done histopathology examination is essential for definitive diagnosis. A careful follow up of these cases is mandatory to watch for recurrence.

CONSENT

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this paper and accompanying images.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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