Journal of Advanced Medical and Dental Sciences Research

@Society of Scientific Research and Studies ***NLM ID:*** *101716117*

Journal home page: www.jamdsr.comdoi: 10.21276/jamdsr Indian Citation Index (ICI) Index Copernicus value = 100

(e) ISSN Online: 2321-9599; (p) ISSN Print: 2348-6805

Case Report

Inflammatory myofibroblastoma of maxilla- A rare pseudotumor?

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ABSTRACT:
An inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal tumor of intermediate malignant potential that predominantly affects children, adolescents, and young adults. The lesion has been reported to occur at various sites of the body but rarely affects the head and neck region. Its occurrence in the maxillofacial region is extremely rare and reported sites include buccal mucosa, tongue, gingiva, floor of the mouth, and retromolar area. The clinical presentation usuallymimics malignancy and poses a diagnostic challenge. Hence IMT should be included in the differential diagnosis of locally aggressive lesions of jaw bones. Clinicopathological correlation and immunohistochemical examinations are essential for the confirmation of the diagnosis. Here, we report a case ofinflammatory myofibroblastic tumor of the maxilla in a 27-year-old male.

Keywords: IMT, Inflammatory myofibroblastic tumor, inflammatory pseudotumor, spindle cell tumor

Received: 21 May, 2024 Accepted: 24 June, 2024

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This article may be cited as: Lakshmi B, Sreela LSN, Sivaprasad T, Mathew P, Nair AK. Inflammatory myofibroblastoma of maxilla- A rare pseudotumor? J Adv Med Dent Scie Res 2024;12(7):32-35.

INTRODUCTION

An inflammatory myofibroblastic tumor(IMT) is a rare mesenchymal tumor of unknown etiologywith an estimated prevalence of 0.04%–0.7%. The lung, liver, and orbit are the most affected sites, although it has been documented to affect almost every other part of the body, including the oral cavity and the major salivary glands.[1]IMTs are not common in the head and neck region and contribute to 14–18% of all extra-pulmonary IMTs.[2]The clinical presentation usually mimics malignancy and poses a diagnostic challenge.

In scientific literature, terms like inflammatory pseudotumor (IPT), inflammatory fibrosarcoma, and inflammatory myofibroblastic tumor are used interchangeably for describing this entity.These names reflect the original disagreements about the lesion's etiopathogenesis, histology, and nomenclature. Eventually, it was determined by electron microscopy and immunohistochemistry analyses that the growing spindle cells in these tumors were myofibroblasts; as a result, the condition is now commonly referred as IMT.The name IMT was consolidated by the World Health Organization (WHO) in 1994 for the diagnostic categorization of intermediate soft-tissue myofibroblastic neoplasms with well-reproducible histological appearance [3]Recent research suggests that IMTs may differ from IPT in their origin and clinicopathologic characteristics in the central nervous system, spleen, and lymph nodes.[1]

The destructive character IMTmakes them easily mistaken as malignant neoplastic processes. To prevent mutilating and disfiguring surgical treatments, a proper diagnosis through appropriate biopsies is necessary. Here, we report a case of a maxillary IMT with an atypical clinical presentation.

CASE REPORT

A 27-year-old male patient presented with pain on right upper posterior jaw and difficulty in mouth opening for one month. He also reported pus discharge from that region. He had a history of similar episodes of pain 1 year back for which he underwent extraction of a decayed right upper third molar, but symptoms persisted for almost six months. For the present complaint, the patient consulted a dentist one week prior and he underwent extraction of an upper

right second molar. He was on antibiotics for one week without any relief of symptoms. His medical history family and social history were non-contributory. His vital signs were within the normal range.

Extra orally, he had no gross asymmetry of the face. Hismouth opening was 13mm. On intraoral examination, diffuse erythema was noted over the buccal gingiva of the maxillary right first molar, which was tender on percussion and grade 1 mobility. The extraction socket of the second molar appeared to be inflamed. No active evidence of pus discharge was noted. A complete examination of the site was not possible due to the restricted mouth opening. The initial clinical differential diagnoses considered were post-extraction residual infection, deep fungal infection, and odontogenic/maxillary sinus malignancy.

On radiographic examination, a panoramic radiograph (OPG) revealed an ill-defined radiolucency in relation tothe right maxillary second and third molar region extending anteroposteriorly from the distal aspect ofthe firstmolar to the posterior limit of maxillary tuberosity. The floor of the right maxillary sinus was not evident and appeared discontinuous. Cone beam computed tomography scan displayed the extension of the lesion to the right maxillary sinus completely obliterating the cavity. The buccal and palatal cortical plate discontinuity was also evident.Considering the above findings, radiographic differential diagnoses were more in favour of aggressive lesions like deep fungal infection, odontogenic malignancy and maxillary sinus malignancy.

An incisional biopsy was performed from the right gingiva-buccal sulcus and maxillary sinus and the microscopic examination revealed a spindle cell-rich stroma arranged in a storiform pattern with scattered inflammatory cells, suggestive of inflammatory myofibroblastic tumor. The immunohistochemical analysis showed the tumor cells were positive for smooth muscle actin, and negative for anaplastic lymphoma kinase (ALK). The sample was also subjected to Fluorescence in-situ Hybridization for ALK-1 gene break apart Rearrangement Assay which turned out to be negative confirming the histopathological diagnosis.

The patient was referred to a higher center for further management. He was advised to take oral Prednisolone 20mg and calcium and vitamin D3 supplements initially for 1 month. Later he was prescribed Tab Mycophenolate sodium 360 mg and Tab Prednisolone in tapered doses along with calcium and vitamin supplements. On the review after three months, he showed improvement in mouth opening, and a reduction in the size of the lesion.

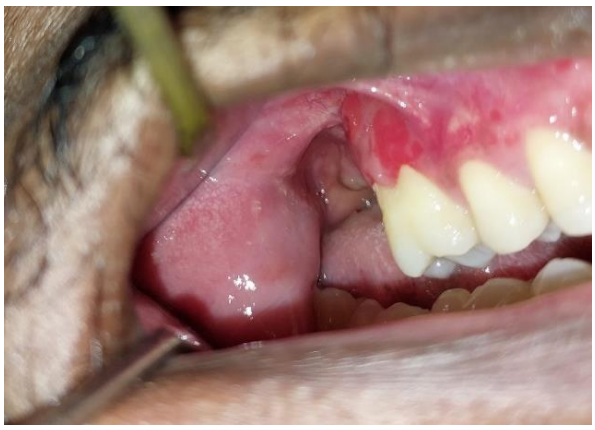

Fig 1. Intraoral image showing eruthema over buccal gingiva of 16

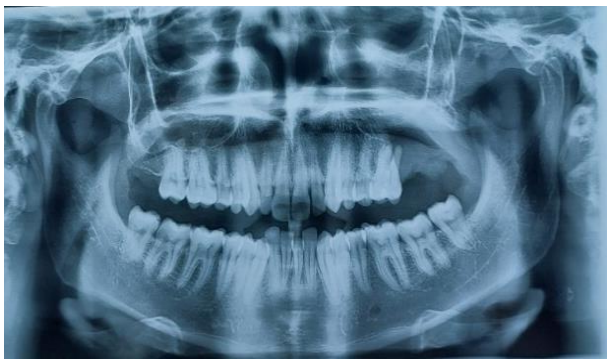

Fig 2. Panoramic radiograph

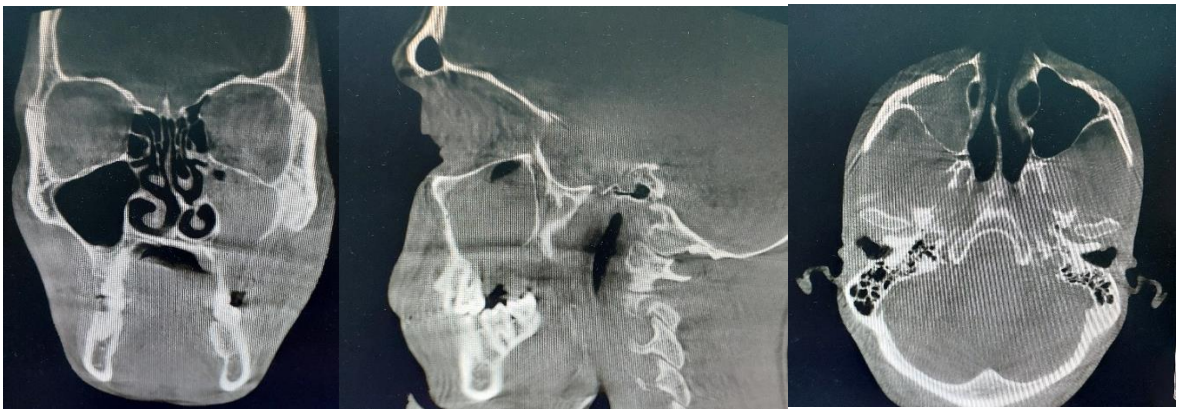

Fig 3. CBCT images

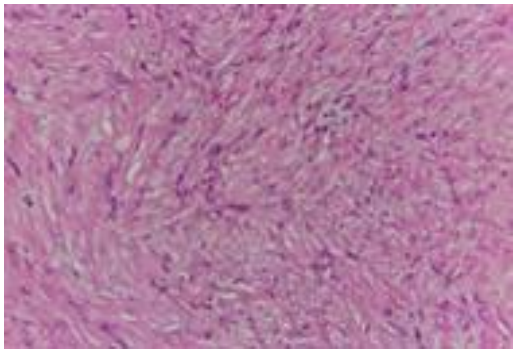

Fig 4. Histopathology showing stroma of spindle cells

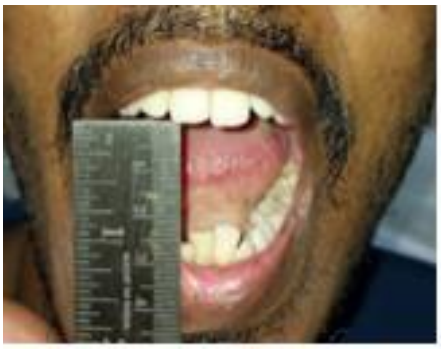

Fig 5. Three months Follow-up

DISCUSSION

IMT is a benign tumor that consists of varying numbers of myofibroblastic spindle cells and inflammatory cells. Traditionally, these tumors have been combined under the general heading of inflammatory pseudotumor.[6]IMT was first identified in the lung and described by Brunn in 1939which was subsequently named by Umiker et al. in 1954.[4]The first report of IMTs in the oral cavity in three children was reported by Liston et al. in 1981.[7]Owing to the clinical and radiological behavior that resembles a malignant disease,IMTs are categorized as tumors of intermediate biological potential by the World Health Organization.[8]

The etiopathogenesis of IMT is obscure and controversial, but neoplastic origin seems to be most acceptable.[11] The pathophysiology of IMT has been linked to the ALK (chromosome band 2p23) gene, which supports the neoplastic etiology of tumors. Lawrence et al. discovered two distinct balanced chromosomal translocations involving the ALK kinase gene. One at chromosome 2p23 and the other involving the tropomyosin 4 gene (TPM4) at chromosomal locus 19p13.1, or the tropomyosin 3 gene (TPM3) at chromosomal locus lq21, leading to translocations t(1;2) (q21;p23) and t(2;19) (p23;p13.1), respectively.[12]ALK-positive has been linked to about half of all IMTs. These translocations cause ALK kinase to be expressed in IMT, as determined by immunohistochemistry analyses. According to reports, the majority of earlier oral IMT cases were ALK-negative. ALK-positive IMT have in fact sparked debate on their potential as a distant clinicopathologic entity with neoplastic origin, more aggressive behaviour, and even the ability to become malignant.[13] However,in the present case, ALK expression was found to be negative.

The clinical presentation of IMT can vary.The buccal mucosa is the most commonly affected intraoral site. Even though, IMT predominantly affects children, adolescents, and young adults it can affect any age group ranging from 2 to 82 years, with a mean of 32 years, and slight female predilection.[9,10]As observed in the current case, radiological pictures or clinical features of IMT had low specificity for diagnosing the case.The classification of soft tissue tumors by the World Health Organization (WHO) is used to make the diagnosis of IMT based on histopathological and immunohistochemical results.[14]Histologically, the presence of myofibroblastic spindle cell fascicles intermingled with both acute and chronic inflammatory cells in varying amounts are the key features seen in IMT cases occurring in the maxillofacial region.[9] Atypical mitotic features are rare and stroma frequently is myxoid or collagenous with significant vascularity.[1]

IMTs are generally managed by complete surgical excision with a lower recurrence rate. Conservative management with systemic immunosuppressants and corticosteroids has been found effective in the management of IMTs mostly indicated in cases of inoperable sites.[5]A total of 29 cases of oral IMT have been documented till 2019.[4] However, intrabony IMT

is extremely rare with only three recorded cases till date.[1]

In summary, IMT is an uncommon tumor in the maxillofacial region, and it may be challenging to diagnose the tumor or distinguish it from other mucosal lesions.Oral IMT should be considered in the differential diagnosis of localized gingival expansion mimicking oral hyperplastic/reactive lesions, notwithstanding its exceptional rarity. Furthermore,IMT in the oral cavity might resemble other malignant tumors in terms of histology, radiography, and clinical presentation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given hisconsent for his images and other clinical information to be reported in the journal. The patient understand that his name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

None

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