https://doi.org/ 10.33472/AFJBS.6.5.2024. 6427-6437

African Journal of Biological Sciences

Inflammatory myofibroblastic tumour: A rare finding in the jaws

TYPE OF ARTICLE: Case Report

AUTHOR: Shivakumar L[1]

[1]Senior Lecturer, Department of Oral and Maxillofacial Surgery, Sree Balaji Dental College and Hospital, Bharath Institute of Higher Education and Research, Chennai.

Article History
Volume 6, Issue 5, 2024
Received: 15 May 2024
Accepted: 22 May 2024
doi: *10.33472/AFJBS.6.5.2024. 6427-6437*

ABSTRACT:

Inflammatory Myofibroblastic Tumor (IMT) is an uncommon benign lesion, identified first in the lungs. However, it exhibits aggressive growth, leading to clinical suspicions of malignancy. It was also known as inflammatory pseudotumor because of this presentation. It is composed of myofibroblastic spindle cell proliferation with a prominent chronic inflammatory cell infiltrate. Immunohistochemistry to demonstrate Anaplastic Lymphoma Kinase (ALK) should also be used to aid in the diagnosis. IMT in the head & neck region is exceedingly rare, but not unheard of. In this paper, we present a case report of an IMT that occurred in the mandible of a young girl, which was managed by surgical resection, which is the treatment of choice, as there have been no recorded cases of recurrence post resection.

KEYWORDS:

Inflammatory myofibroblastic tumour, Mandibular tumour, Immunohistochemistry, Anaplastic Lymphoma Kinase.

INTRODUCTION

Inflammatory Myofibroblastic Tumour (IMT) is a benign tumour of unknown etiology. It was first observed in the lungs by Brunn in 1939 and named by Umiker in 1954.[1] Extrapulmonary sites such as the liver and gastrointestinal tract are the other common sites.[2] It is a rare lesion in the head and neck region, where it most commonly occurs in the orbit and upper airway. It is rarer still in the oral cavity. However, cases of IMT in the retromolar and

pterygopalatine area, tongue, maxilla, hard palate, mandible, floor of the mouth, buccal mucosa, and major salivary glands have been reported.[3]

In 2013, the WHO classification of soft tissue and bone tumours defined IMT as a distinctive neoplasm composed of myofibroblastic and fibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and/or eosinophils.[4]

Although various pathogeneses such as trauma, Epstein-Barr virus, radiotherapy have been suggested, the etiology remains unclear.[4] Current evidence suggests that IMTs are neoplastic processes resulting from chromosomal translocations that cause an overexpression of ALK (Anaplastic Lymphoma Kinase) tyrosine kinase.[5]

Till recently, IMT was more commonly known as Inflammatory pseudotumor because it mimics a malignant lesion both clinically and radiographically. It was also known by several other names such as plasma cell granuloma, pseudosarcoma, myxoid hamartoma, inflammatory myofibrohistiocytic proliferation, and benign myofibroblastoma.[1] Its current nomenclature was decided by the WHO in 1994.[5] These multiple descriptive terminologies only further reflect the uncertainty regarding the true nature of this lesion.[1]

In this case report, we describe a rare case of IMT in the mandibular body region in a young girl, which was managed by surgical resection.

CASE REPORT

A 17-year-old female reported to the department of Oral & Maxillofacial Surgery with a chief complaint of a swelling inside her mouth, on the left side of the lower jaw, for the previous 4-5 months.

The patient gave a history of an initial small swelling that gradually increased in size over the course of 4-5 months. It was painless and there were no other symptoms such as discharge or numbness associated with the swelling. The patient did not have any significant medical history.

On extraoral examination, a diffuse swelling was observed over the left lower 3rd of the face involving the inferior border of mandible (figure 1). The swelling was bony hard and tender on palpation. No abnormal changes were observed on the skin overlying the swelling. Mouth opening was unrestricted. Intraorally, a well-defined sessile growth is seen extending along the lower left alveolus in the premolar-molar region (between tooth numbers 35 & 36), extending over both the buccal and lingual surfaces. But the major portion of the lesion was present lingually. The swelling was irregular in shape, firm on palpation and non-tender. The

mucosa over the swelling was of the same colour as the adjacent normal mucosa with a mix of erythematous and keratotic areas. There was mild ulceration in the areas where the maxillary teeth occluded. The surface of the swelling was otherwise smooth and there were no discharging sinuses present. There was no bleeding on provocation. There was associated mobility of tooth number 35 (figure 2).

Radiographically, Cone Beam CT of mandible revealed the presence of a well-defined, irregular radiolucency in the left mandibular body region, extending from distal to tooth number 35 to just in front of the angle region. There was involvement of the inferior border with slight bowing. The axial and coronal sections showed expansion and thinning of both buccal and lingual cortices (figure 3).

The swelling was provisionally diagnosed as a giant cell lesion and an incisional biopsy was performed, which revealed the diagnosis to be "Inflammatory Myofibroblastic Tumour". Surgical management was planned, and a segmental resection of the involved part of the mandible was done under general anesthesia, followed by fixation of a titanium reconstruction plate spanning the defect (figure 4) (figure 5).

DISCUSSION

IMTs may occur anywhere in the body, including the head and neck, but its occurrence in this region is extremely rare. Very few cases of IMT in the oral cavity have been reported. Although this can be attributed to its rarity intraorally, it can also be explained by the fact that its classification was unified only recently.[5]

Demographically, IMT is a mixed bag as it occurs equally both in males and females, spread across a wide age range of 19-77 years. However, it has a predilection to occur in children and young adults, which is also the case in this report. Intraoral lesions are known to exhibit a rapid growth rate in a few instances.[6] In this case, although the lesion was not huge by any standard, it did reach a considerable size in only a few months' time.

Clinically, IMTs are painless swellings or indurated masses of short duration. The patient in this report also had a painless swelling growing for about 5 months. Radiological findings are non-specific as it was in this case, where the irregular radiolucent lesion could have been mistaken for any osteolytic lesion such as a cyst or an odontogenic tumour.[7]

Histopathologically, our case demonstrated "***multiple fasciculi of cells with abundant cytoplasm and prominent nuclei against a background of fibromyxoid stroma with abundant inflammatory cell infiltrate"*** (figure 6). These cells resembled myofibroblasts,

which along with the other features led to the diagnosis of IMT being made. However, these features might be present in any tumour or inflammatory process that possesses fibroblasts and myofibroblasts.[5] Diagnosis is based on exclusion. It is common for patients to undergo multiple biopsies before a diagnosis can be established.[7,8]

Definitive diagnosis depends on immunohistochemistry (IHC) by demonstration of positive staining for ALK-1. Although this is highly specific, it is not 100% sensitive as ALK negative IMTs are also known to exist. They are morphologically indistinguishable from ALK positive lesions.[1] At the time of writing this paper, IHC was not done for this case and our diagnosis was based only on histopathological features.

Non-surgical methods of management such as radiotherapy has been tried in a few instances such as in unresectable or non-responsive cases and as an adjuvant to surgery. But it carries with it, a risk of malignant transformation. Intralesional injection of corticosteroids has also been tried in a few cases with not much efficacy, the possible mechanism of action being the reduction of inflammatory cell infiltration.[4,9]

Complete excision of lesion/resection with negative margins has been suggested as the best method of management of this lesion as it provides the best prognosis.[10] There has been no reports of recurrence of the lesion or metastasis occurring with IMTs till date.[2] Segmental resection was done in this case, with the patient doing well post-surgery (figure 7) (figure 8). In the follow-up period so far, no signs of recurrence have been observed.

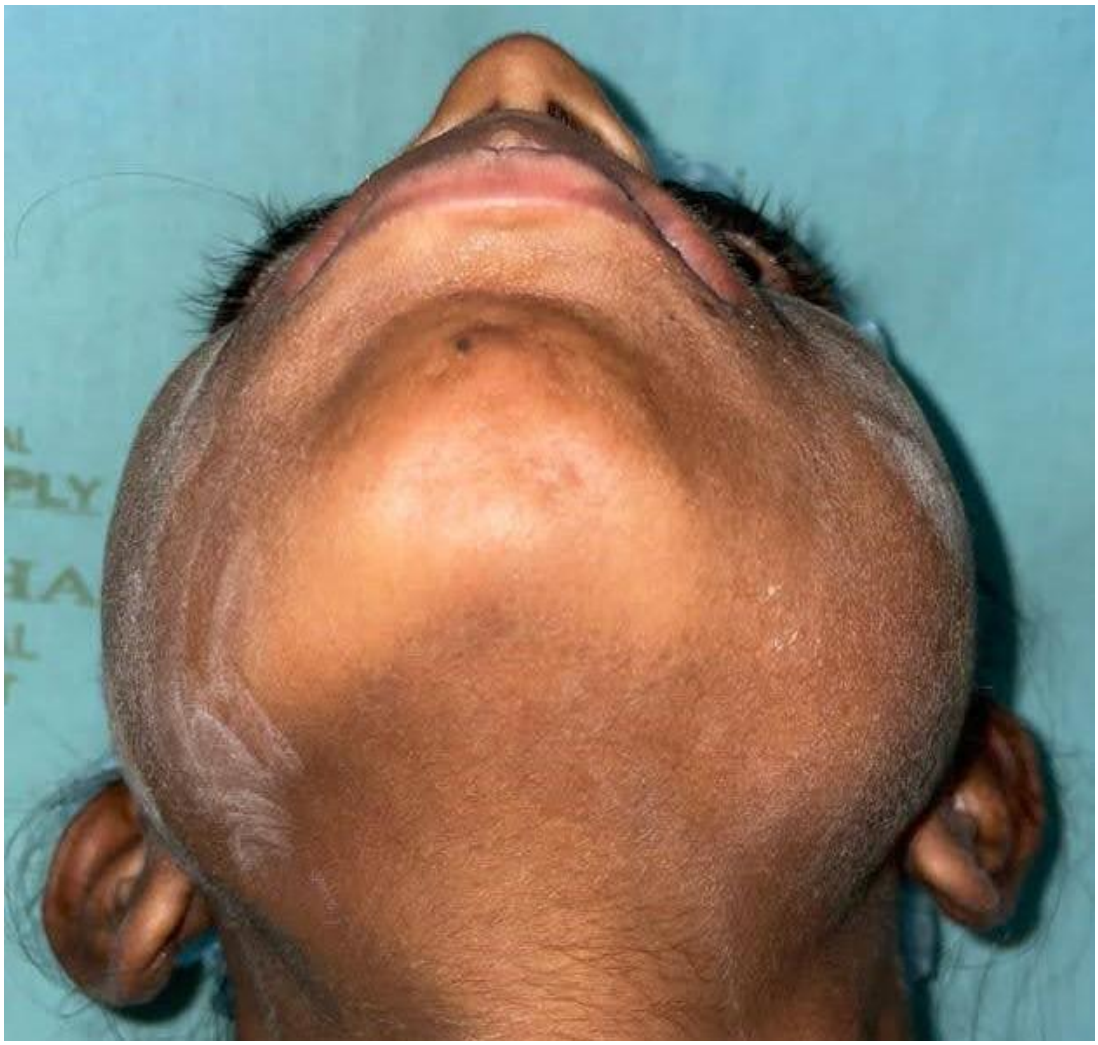

Figure 1: Extraoral diffuse swelling involving the inferior border of mandible

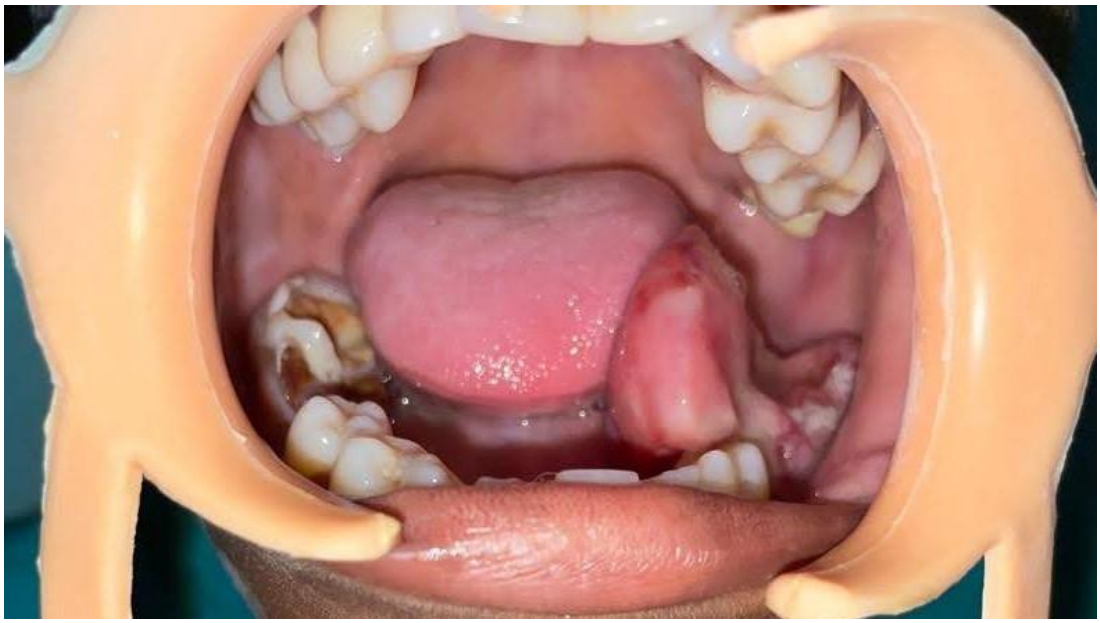

Figure 2: Intraoral swelling involving the mandibular alveolus

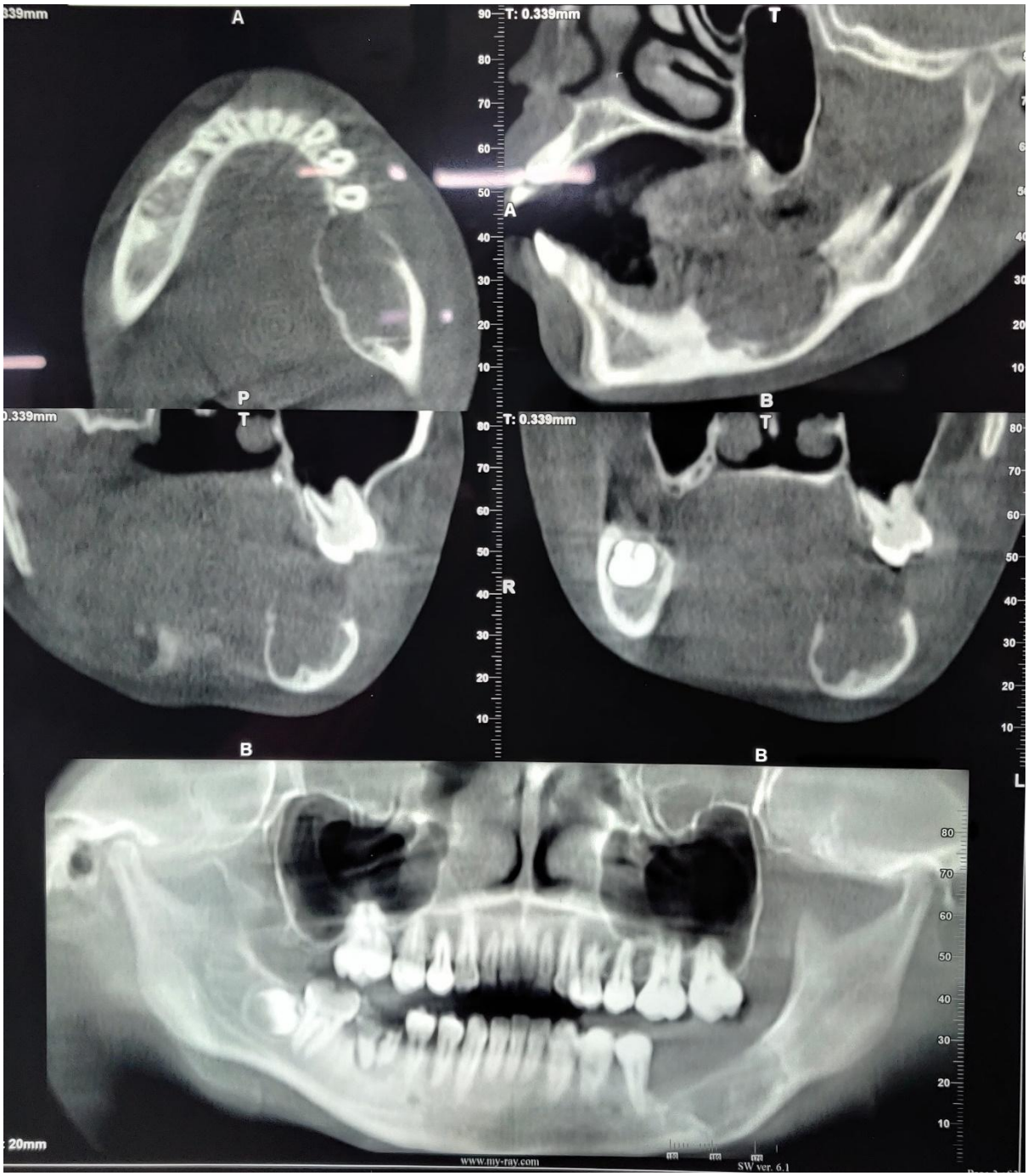

Figure 3: CBCT showing a radiolucent lesion involving the left body of the mandible with expansion of both buccal and lingual cortices seen on axial and coronal sections

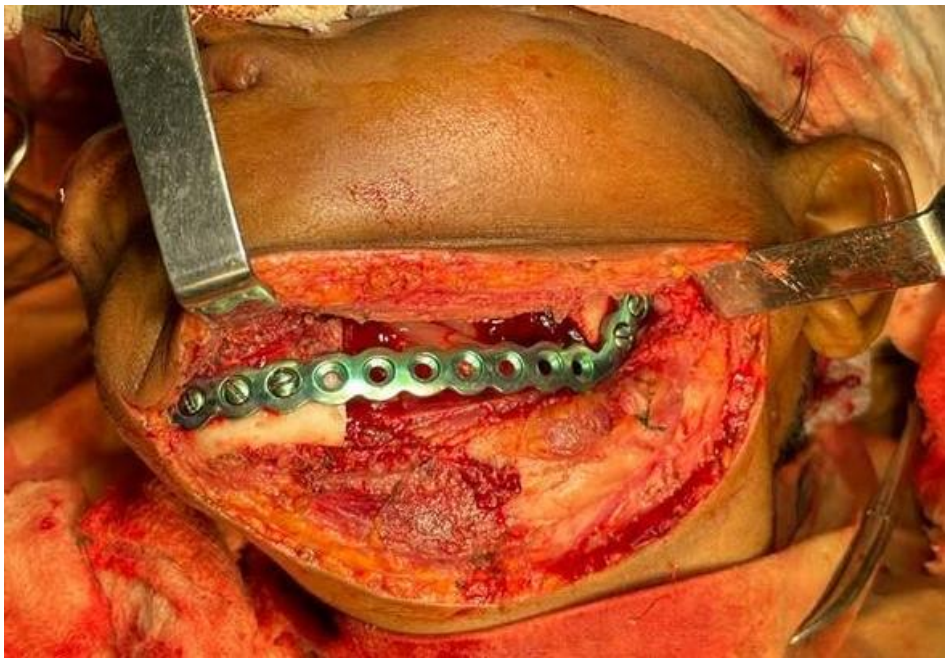

Figure 4: Intra-op image showing Titanium Reconstruction plate fixed after segmental resection of the involved part of the mandible

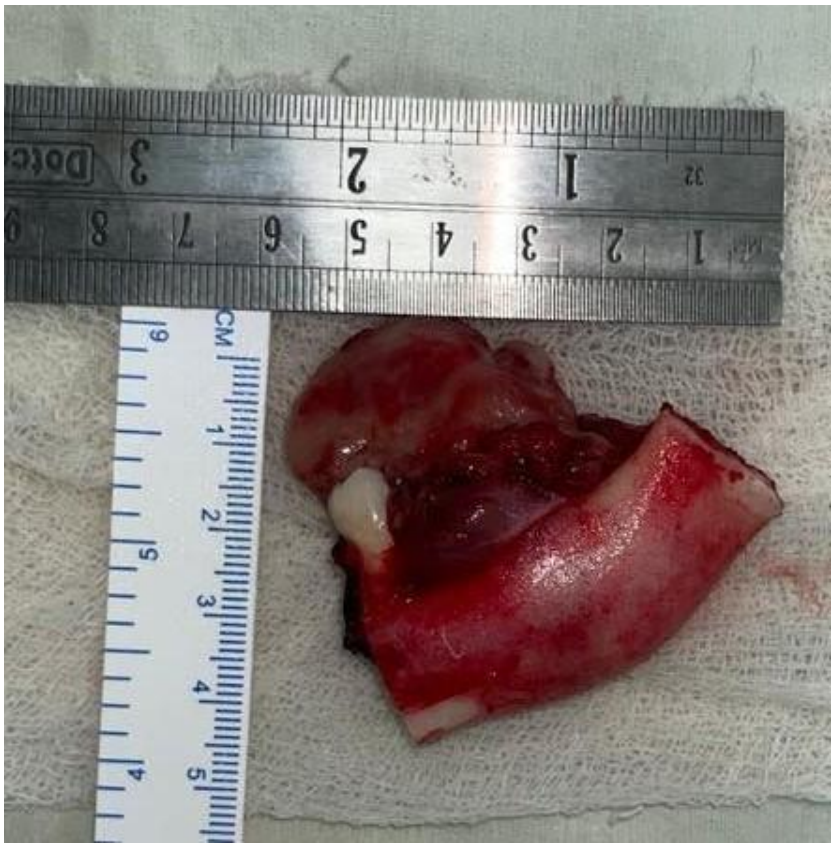

Figure 5: Image of the resected specimen

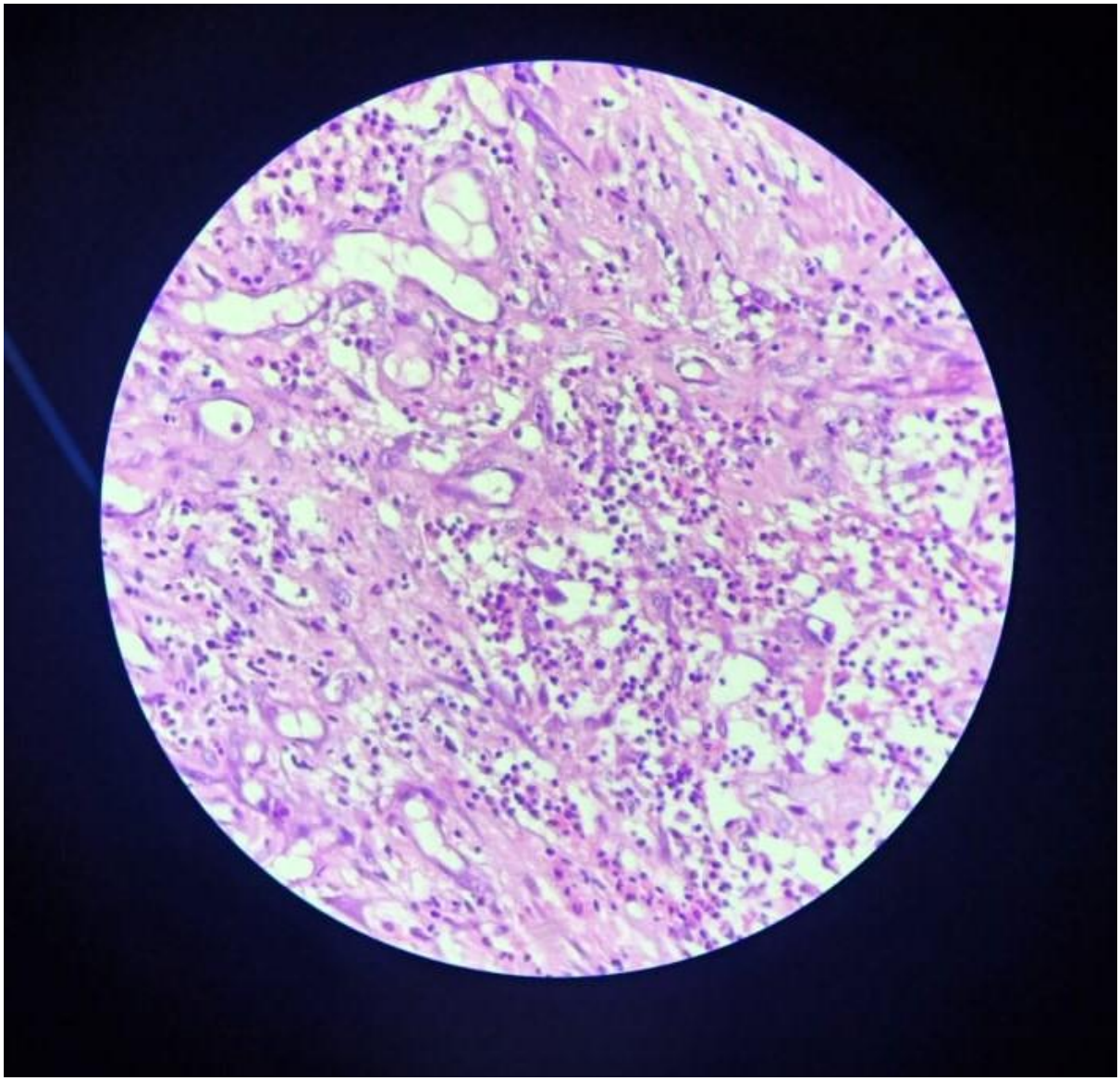

Figure 6: Photomicrograph of the slide showing characteristic myofibroblasts and inflammatory cells on a fibro-myxoid background

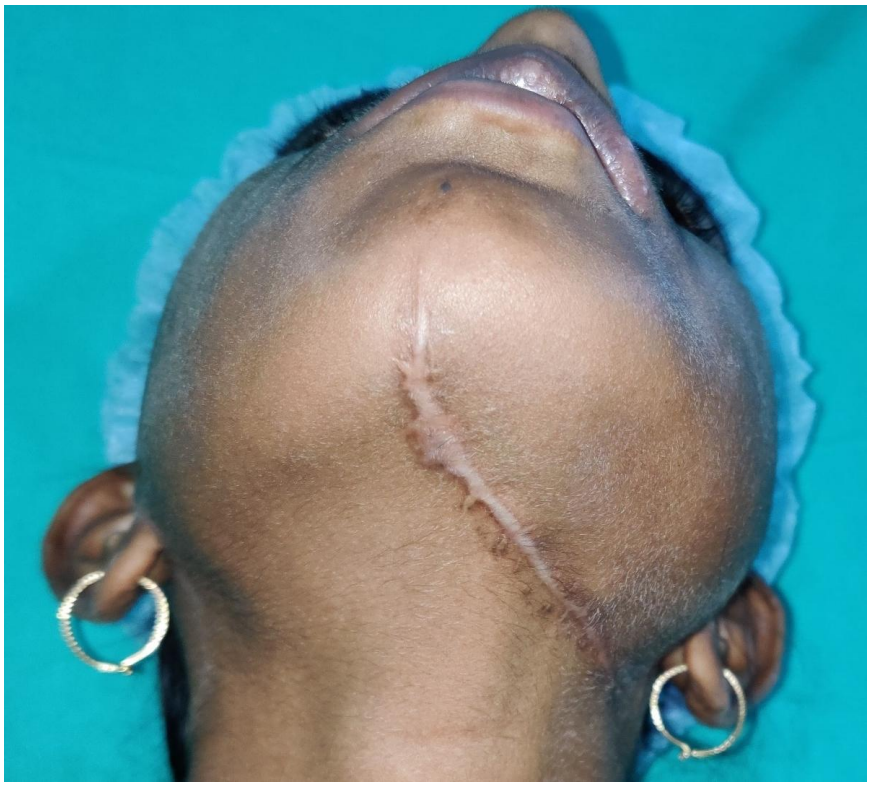

Figure 7: Follow-up image of the patient showing uneventful healing but with prominent scarring along the incision line

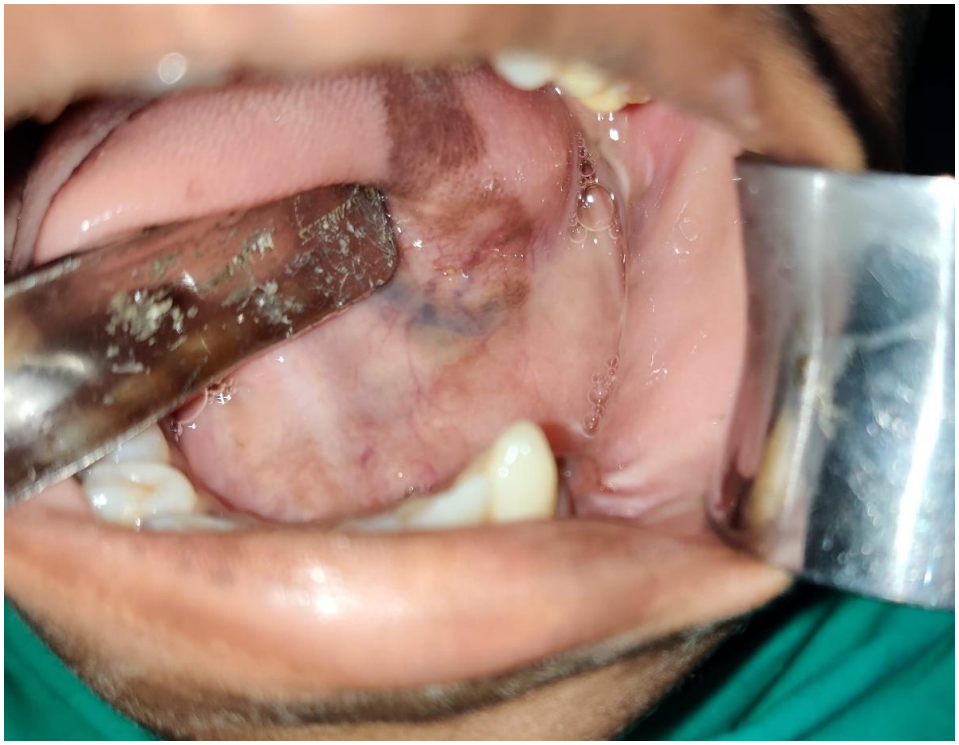

Figure 8: Intraoral follow-up image showing complete healing with healthy mucosa

CONCLUSION

IMTs are extremely rare lesions, especially in the head and neck region, whose non-specific features make it difficult to establish a diagnosis and come up with an appropriate treatment plan. The surgeon should have in his arsenal, knowledge of various inflammatory and neoplastic lesions of the head and neck to arrive at a diagnosis by exclusion. Since the body of knowledge on this lesion is insufficient at present, all patients undergoing treatment for this lesion must be kept on regular follow up to identify any clinical/radiographic signs of recurrence. As it is an uncommon lesion, the most important thing is to not be hasty in establishing the diagnosis as even histopathology can be misleading in this lesion and multiple biopsies & IHC must be used whenever required.

REFERENCES:

1. Palaskar S, Koshti S, Maralingannavar M and Bartake A. Inflammatory myofibroblastic tumor. *Contemp Clin Dent* 2011;2(4):274-7.

2. Korlepara R, Guttikonda VR, Madala J andTaneeru S. Inflammatory myofibroblastic tumor of mandible: A rare case report and review of literature. *J Oral Maxillofac Pathol* 2017;21(1):136-9.

3. AlKindi MG. A rare case of inflammatory myofibroblastic tumor of the mandible mimicking a malignant tumor. *Saudi Dent J* 2017;29(1):36-40.

4. Monteiro L, Alhassani M, Resende T and Albuquerque R. Inflammatory myofibroblastic tumour of the jaw: A rare presentation. *J Stomatol Oral Maxillofac Surg* 2020;121(1):90-4.

5. Stringer DE, Allen CN, Nguyen K and Tandon R. Intraosseous inflammatory myofibroblastic tumor in the mandible: a rare pathologic case report. *Case Rep Surg* 2014;2014:565478. .

6. Rahman MA. Inflammatory myofibroblastic tumour of the mandible – A case report. *UpDCJ* 2020;10(1):21-4.

7. Al-Sindi K, Al-Shehabi MH and Al-Khalifa SA. Inflammatory myofibroblastic tumor of paranasal sinuses. *Saudi Med J* 2007;28(4):623-7.

8. Deshingkar SA, Tupkari JV and Barpande SR. Inflammatory myofibroblastic tumour of maxilla. *J Oral Maxillofac Pathol* 2007;11:76-9.

9. Narla LD, Newman B, Spottswood SS, Narla S and Koll IR. Inflammatory pseudotumor. *Radiographics* 2003;23:719-29.

10. Ong HS, Ji T, Zhang CP, Li J, Wang LZ, Li RR *et al*. Head and neck inflammatory myofibroblastic tumor (IMT): evaluation of clinicopathologic and prognostic features. *Oral Oncol* 2012;48(2):141-8.