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A RARE CASE OF LOW GRADE MYOFIBROBLASTIC SARCOMA OF THE MANDIBLE

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ABSTRACT
A 19 year young girl was referred to our institute with a swelling over right alveolus that had progressed over last two months. She was having history of frequent oral bleed without any other associated systemic or local symptoms. No significant personal or family history was present. Her systemic examination was unremarkable and on local examination, there was a 3x4 cm fleshy, bosselated mass involving right lower alveolus at the level of last premolar to 2nd molar and crossing laterally to gingivo-buccal sulcus through the inter-molar space without involving any other adjacent structures (tongue/ floor of mouth) and neck nodes. Routine haematological and biochemical investigations were within normal range except haemoglobin of 10.3 gm%. Contrast enhanced computed tomography (CECT) scan of face and neck revealed soft tissue mass attached to right lower alveolus with feeders in arterial and venous phase; angiography suggested all these feeders from facial artery (figure 1). Possibility of arterio-venous malformation was kept in mind and planned for excision. Histopathology was suggestive of low grade myofibroblastic sarcoma with negative margins, which was further confirmed by immunohistochemistry of tumor cells (positive for SMA and negative for Desmin, CD34 and S-100).

INTRODUCTION

Myofibroblasts are mesenchymal spindle cells sharing immunohistochemical and ultra-structural features of both fibroblasts as well as smooth muscle cells [1-2]. They have been shown to participate in wound healing and various benign and malignant soft tissue tumours [3]. Low-grade myofibroblastic sarcoma (LGMS) has been classified as a distinct entity in the newly published World Health Organisation classification of soft tissue tumors [4]. LGMS mainly affects the soft tissue of the oral cavity, limbs, trunk or abdominal/pelvic cavities and rarely bone [5-11]. In the world literature, there have been 51 published cases of LGMS [5-11]. Among these cases, the most common location has been the soft tissue of the head and neck followed by extremities, trunk, retro-peritoneum, bone, chest wall and breast [5-11].

Figure 1. CECT scan of face and neck

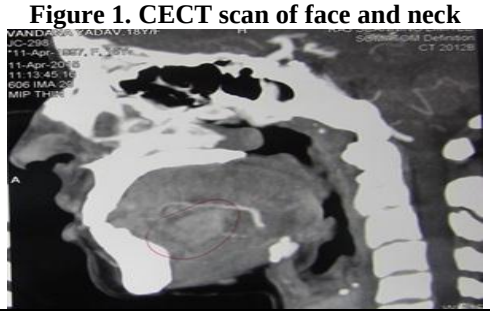

Table 1. **Summary of reported cases of Low-grade myofibroblastic sarcoma affecting mandible [3, 14-16]**

Case	Age (year)/Sex	Size/cm	Treatment	Recurrence
1	9/F	NA	NA	Y
2	9/F	NA	NA	N
3	19/M	3.5	Local excision, RT, CT	N
4	51/M	3.0	Wide excision	N
5	54/M	6.0	Local excision	N
6 (our case)	19/F	4.0	Local excision	N

DISCUSSION AND CONCLUSION

The diagnosis of LGMS is usually made on clinical and pathological grounds including morphological, immunohistochemical and ultra-structural features [5,12]. As LGMS is a rare disease, there are few treated cases. Surgery is the main therapeutic treatment of modality [5,13]. There have been only 6 cases of LGMS affecting the mandible till now [3,14-16] (table 1). The average age was 26.8 years (range: 9-54 years; median: 19 years) and the male to female ratio was 1:1. The recurrence rate was 16.7%.

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CONFLICT OF INTEREST: NIL

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