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CLINICAL REPORT

Low Grade Fibromyxoid Sarcoma in Oral Cavity: A Rare Case Report

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Abstract Low-grade fibromyxoid sarcoma (LGFMS) is a rare, low-grade malignant soft tissue neoplasm. It is a distinctive variant of fibrosarcoma with a high metastasizing potential and sometimes long interval between tumor presentation and metastasis. It may present in any part of body. However in the region of the head and neck, they are seldom seen and hence easily misdiagnosed. We report the case of an 18-year-old female who developed a large mass in the left oral cavity. The tumor was excised. On pathology evaluation was diagnosed as LGFMS. Due to the relative rarity of LGFMS, there is no dedicated protocol regarding follow-up recommendations.

Keywords Oral cavity · Low-grade fibromyxoid sarcoma · Excision and follow-up

Introduction

Low grade fibromyxoid sarcoma is a rare soft tissue tumor first describe by Evans [1]. It is characterized by its relatively benign histological appearance with spindle cells in a whorling pattern, as well as collagenized and myxoid areas. In spite of the low-grade and benign histological appearance it has paradoxically aggressive behavior, with a high rate of recurrence and metastasis [2]. LGFMS commonly arise from the deep soft tissues of the lower extremities, but occasionally reported to arise also, from the chest wall, axilla, inguinal region, buttock, neck, the mediastinum and retroperitoneum [3]. It may present in any part of the body however in the oral they are seldom seen and hence easily misdiagnosed.

Herein, we present a case of an 18-year-old female who developed a mass in the left oral cavity.

Case report

An 18 years old female presented in department of Otorhinolaryngology RIMS Imphal with a two month history of an enlarged mass in her left buccal mucosa. The mass was painless but it was constantly enlarging. On examination a reddish fleshy mass of around $5 \times 3 \times 2$ cm well circumscribed firm in consistency non tender with smooth surface (Fig. 1). Routine investigations were within normal limit, FNAC- suggests pleomorphic adenoma of minor salivary gland and histopathology of biopsy shows Low Grade fibromyxoid Sarcoma. Wide excision of the swelling was done and the specimen sends for Histopathological examination. Histology shows low-grade fibromyxoid sarcoma consisting of bland spindle cells in a collagen and myxoid background (Fig. 2). In 18 month of follow up there was no local recurrence.

Discussion

Low grade fibromyxoid sarcoma as describe first by Evans in 1987 is a slow growing tumor with apparently benign histological characteristic with a high metastatic potential [1]. The usual presentation of the tumor is painless soft tissue mass mostly in the lower extremity other region can be affected chest wall, abdomen inguinal region, buttock

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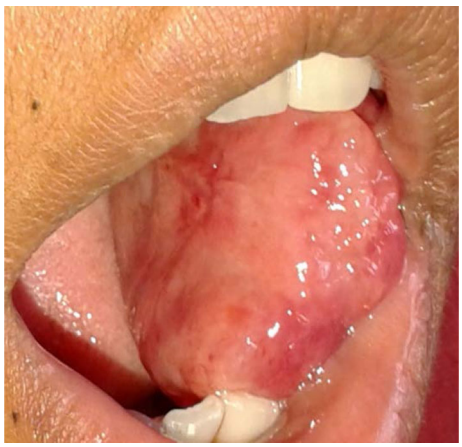

Fig. 1 Mass in left oral cavity

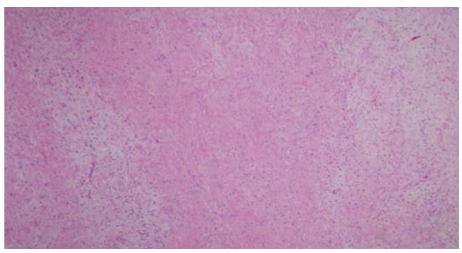

Fig. 2 Histopathology of low grade fibromyxoid sarcoma

and neck [3, 4]. Rare cases have also been reported in in unusual sites including the retro peritoneum, small bowel mesentery and paravertebral region [5, 6].

The diagnosis done by histopathology characterized by presence of bland spindle cells, showing mainly of whorled or focally linear arrangement, set in alternating areas with fibrous or myxoid stroma. Immunohistochemicaly the tumor cells are strongly positive for vimentin [3]. Fibromyxoid sarcoma is difficult to diagnose by fine needle aspiration cytology because of morphological overlap with other spindle cell and myxoid lesion [7]. In this case wide excision with normal margin was done to avoid recurrence [2]. Evans [2] and Goodlad et al. [3] suggested that LGFMS were paradoxically aggressive tumors and local recurrence was noted in 68 %, metastasis in 41 %, and death from disease in 18 % [2].

Conclusion

There is very rare known occurrence of Low Grade fibromyxoid sarcoma in oral cavity. However the current case report present further information regarding the diagnosis and management of LGFMS. There is no definite protocol for the early diagnosis, treatment or follow up. Since, it is associated with a high incidence of recurrence and metastasis after a long duration. All LGFMS must be treated as a malignant tumor and thus, undergo a wide excision, followed by full oncological assessment and follow up.

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Compliance with Ethical Standards

Conflict of interest The article "fibromyxoid sarcoma in oral cavity a rare case report" is for academic interest and there is no funding and no conflict regarding this article.

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