Oral Health Case Reports
ISSN: 2471-8726
Patil et al., Oral health case Rep 2018, 4:2
DOI: 10.4172/2471-8726.1000147
Case Report Open Access

Unhurried Growth of Low-Grade Myofibroblastic Sarcoma of the Mandible: A Rare Report

Bharati Patil, Abhisheak R Naik* and Dhanalakshmi T*

Department of Oral Medicine and Radiology, The Oxford Dental College, Bommanhalli, Bangalore, India

Abstract

Low-grade myofibroblastic sarcoma (LGMS) is a rare, mesenchymal malignant tumor with myofibroblastic differentiation. It has been reclassified as a distinct entity in the newly published World Health Organization classification. Only a few cases have been reported in the oral and maxillofacial region. Here, a LGMS developed in the right retromolar region of a 12-year-old boy who presented with a mass of 2.5 cm × 1 cm × 1 cm size. Based on the histological and immunohistochemical findings, a diagnosis of LGMS was established. The tumor was resected, and no recurrence was observed over one year. Follow-up is essential for early recognition and intervention of late recurrence and metastases.

Keywords: Low-grade myofibroblastic sarcoma; Oral; Mandible

Introduction

As the name signifies, Myofibroblastic Sarcoma (MS) is a malignant tumor mainly composed of myofibroblasts. Myofibroblasts are spindle cells of mesenchymal origin, mainly discovered in healing wounds within granulation tissue and play a role in the production of contractile force. Myofibroblasts subsist in various benign and malignant soft tissue tumors [1]. Infrequently, myofibroblasts undergoes carcinogenic transformation and become malignant [2]. Many researchers failed to recognize the reason and phenomenological interpretation of the myofibroblast's differentiation into tumor cell, also the treatment method and its prognosis [3]. Low, intermediate and high-grade myofibroblastic sarcomas are rarely reported in the literature. In 2002, World Health Organization (WHO) included a new disease entity low-grade myofibroblasts sarcoma and thus proclaims the existence of such tumors under the classification of bone and soft tissue tumors [4].

Low-grade myofibroblastic sarcoma (LGMS) represents an atypical tumor composed of myofibroblastic differentiation of cell. Very few cases been reported till today [5]. The correct diagnosis of myofibroblastic sarcomas is challenging because patients complain of painless swelling or an enlarging firm mass with pale and fibrous cut surfaces [6]. We report a case of LGMS seen in the right retromolar region in a young patient with clinical, histological and immunohistochemical features.

Case Report

A 12-year-old patient was referred to our department by his dentist for painful gingival lesion in the right retromolar region, persisting for more than 3 months and not responding to antibiotic and periodontal therapy. His personal and familial medical histories were unremarkable. Oral examination revealed a well-defined exophytic growth in the right retro-molar region measuring 2.5 cm × 1 cm × 1.5 cm in size. Lesion extended antero-posteriorly from mesial surface of 47 to right retromolar region, medio-laterally from buccal sulcus of 47 to 1 cm away from the arch and supero-inferiorly 2 cm above the occlusion to 2 cm into the lingual sulcus. The overlying mucosa appeared reddish and irregular with indentation of the teeth. On palpation, it was firm in consistency, tender, and bleeding was noted (Figure 1). A solitary right submandibular lymph node measuring 1 × 2 cm, slightly tender, firm to hard in consistency, non-fixed to underlying tissue was observed. Based on the above findings a provisional diagnosis of Ameloblastic fibroma of the right retro-molar region was given with differential diagnosis of Eosinophilic granuloma.

OPG revealed presence of 3rd molar tooth buds in all quadrants. Discontinuity of the radiopaque line in the coronal portion of 48 was noted suggestive of an outgrowth of the follicle. Further there was widening of PDL space in the apical region of 47 (Figure 2).

The patient underwent complete surgical excision of the overlying gingival growth. Histological examination of the mass revealed the diagnosis of myofibroblastic sarcoma and tumor-free resection margins. Histologically, the lesion was composed of hypercellular areas and revealed infiltration of spindle tumor cells surrounded by a fibrous capsule. The neoplastic cells were dense in some parts but

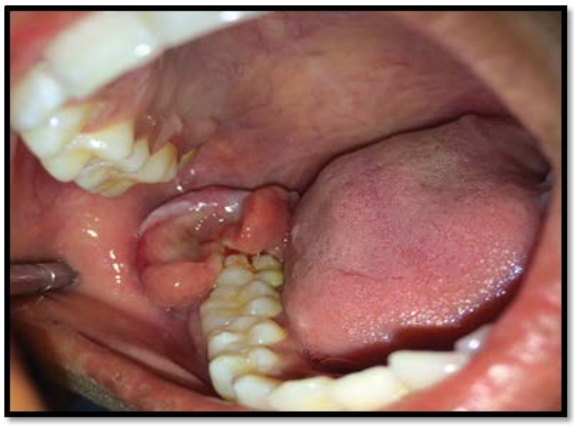

Figure 1: Intraoral view of soft tissue mass in the retromolar region of the mandible showing well defined exophytic growth.

***Corresponding author:** Abhisheak R Naik, Student in Department of Oral Medicine and Radiology, The Oxford Dental College, Bommanhalli, Bangalore, India, Tel: +8095384902; E-mail: abhisheakrnaik@gmail.com

Dhanalakshmi T, Student in Department of Oral Medicine and Radiology, The Oxford Dental College, Bommanhalli, Bangalore, India, Tel: +8150820555; E-mail: drdhanalakshmi1994@gmail.com

Received: September 18, 2018; **Accepted:** November 20, 2018; **Published:** November 22, 2018

Citation: Patil B, Naik AR, Dhanalakshmi T (2018) Unhurried Growth of Low-Grade Myofibroblastic Sarcoma of the Mandible: A Rare Report. Oral Health Case Rep 4: 147. doi:10.4172/2471-8726.1000147

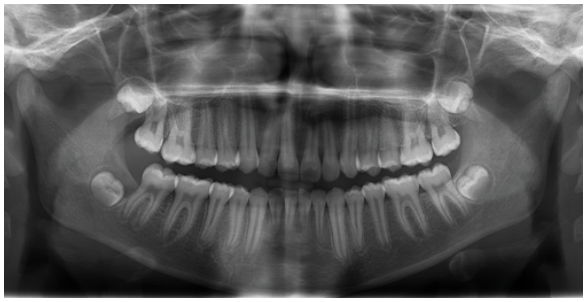

Figure 2: Initial panoramic view showing an outgrowth of the follicle further there was widening of PDL space in the apical region of 47 was present.

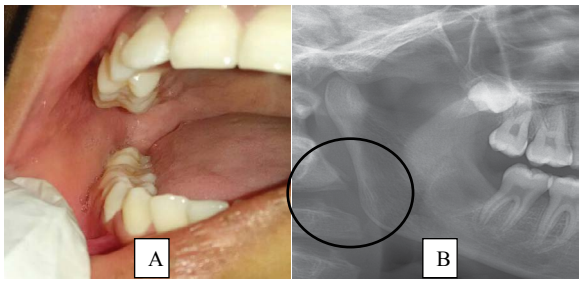

Figure 3: (A) Post-operative clinical photo (**B**) Panoramic X-ray after 12 months.

sparse in other areas which were round-shaped or spindle nuclei, and the cytoplasm was eosinophilic. There were few mitotic cells, however, atypical cells with large nuclei were observed. Immunohistochemically, the neoplastic cells were diffusely vimentin positive, focally smooth muscle actin and calponin positive. All the features were consistent with an LGMS. No adjuvant chemotherapy and radiotherapy was performed to allow for normal growth of the patient. One year post surgery, clinical and radiological follow-up did not show signs of recurrence or metastatic disease. (Figures 3A and 3B). Patient was further advised regular follow-up once in every 6 months [4].

Discussion

Low-grade myofibroblastic sarcoma is a rare, mesenchymal malignant tumor showing myofibroblastic differentiation and has been reclassified as a distinct entity in the newly published World Health Organization classification of soft tissue tumors [4]. In 1971, Gabbiani et al. first described mesenchymal spindle-shaped cells, present in connective tissue that share ultrastructural features with both fibroblasts and smooth muscle cells and contribute to reparative and reactive conditions, such as granulation tissue, hypertrophic scars, and various benign and malignant soft tissue tumors. Clinically, LGMS usually behaves as a slow-growing low-grade malignant sarcoma and exhibits a pattern of aggressive local spread with common local recurrences. Only after a prolonged time metastatic dissemination is seen [5].

LGMS is mainly seen in adults with a mean age of 40.7 years and a preponderance of men is reported.3 Children are rarely affected. LGMS has been reported at a variety of sites, including the extremities, trunk, abdominal and pelvic cavities [6]. About 55 cases have been reported in the oral and maxillofacial region [7]. Tongue is the most common site, followed by maxilla, palate, mandible, nasal/paranasal cavity and deep tissue spaces including the parapharyngeal spaces. Gingiva as the primary site is seen in only one case.

The diagnosis of myofibroblastic sarcomas can be challenging. Size of tumor varies from 1.5 cm to 17 cm [6]. The tumors generally present as a painless or a slow-growing mass with a lethargic course. Absence of a detectable epithelial discontinuity increases the risk of delay in diagnosis and subsequent worsening of prognosis [8]. The conclusive diagnosis and histological grading of myofibroblastic sarcoma are determined through histological and immunohistochemical examination. Study of this tumor tissue by electron microscopy provides the highlight of the myofibroblastic differentiation. Although the diagnosis is difficult which is mainly based on the basis of morphological architecture and the expression of myofibroblastic profile antigens, like smooth muscle actin, desmin, caldesmon and myogenin [9].

In our present case though the tumor was closer to bone, no bone involvement was demonstrated. On histopathological study spindle cells were seen and immunohistochemical analyses showed strong positive immunoreactivity for myogenic markers like α-smooth muscle actin, Vimentin, calponin and rarely desmin which provided a definitive diagnosis of low grade Myofibroblastic sarcoma. According to the study conducted by Binmadi et al. [10] there are about 21 cases of myofibroblastic sarcoma which are seen in the oral cavity, among them 5 cases showed mandible as prominent site between the age group of 20-82 years. In 2012, Park et al. encountered a myofibroblastic sarcoma of the mandible in a 9-year-old male followed by our present case reported as LGMS in the posterior mandible in pediatric patient. Although the treatment has not been clearly defined, an aggressive surgical resection with wide tumor-free margins is the preferred therapeutic option. Occasionally, adjuvant radiotherapy or chemotherapy is considered [11].

LGMS tends to recur locally rather than to metastasize with the rate of 33%. Nasal cavity/paranasal sinus has the highest recrurrence rate (RR) followed by jawbone and deep tissue space. As LGMS is associated with fewer symptoms, early detection may be difficult especially when tumors are in deep tissues, such as a nasal cavity, paranasal sinus, jawbone and masticatory spaces. RR is 21.4% when tumor size less than 3 cm, whereas that of over 3 cm tumor is 46.2%. Recurrence rate was seen in 75% of patients who were treated with local excision alone, while 7% was seen in patients who underwent wide surgical excision with or without radiotherapy.5 As the early diagnosis of LGMS is difficult mainly when it involves nasal cavity, jaw bone, sinus and masticatory space leading to high chances of RR. In our case, the patient underwent a wide surgical excision showed no recurrence after one and half year follow-up postoperatively.

Conclusion

Myofibroblasts are involved in the contractile function and also in the malignant transformation. Diagnosis of myofibroblastic sarcomas can be challenging and it is made on the basis of ultrastructural findings as determined by light microscopy and immunocytochemistry. It's biological behaviour and exact treatment options are left unclear. However, LGMS can be best managed with wide surgical resection and regular follow-up.

References

1. Rao S (2017) Myofibroblast: Overview of a controversial cell. J Interdiscipl Histopathol 5: 12-17.
2. Jayaraj G, Sherlin HJ, Ramani P, Premkumar P, Natesan A (2015) Stromal myofibroblasts in oral squamous cell carcinoma and potentially malignant disorders. Indian J Cancer 52: 87-92.
3. Cai ZG, Pan CC, Yu DH, Feng ZZ, Ma L, et al. (2016) Myofibroblastic sarcomas: Clinicopathologic analysis of 15 cases and review of literature. Int J Clin Exp Pathol 9: 1568-1577.

4. Yamada T, Yoshimura T, Kitamura N, Sasabe E, Ohno S, et al. (2012) Myofibroblastic sarcoma of the palate. International Journal of Oral Science 4: 170-173.

5. Oliveira GG, García IA, Buylla ASA, Burgos RS, Pérez FM, et al. (2015) Low-grade myofibroblastic sarcoma. Two rare tumors in two rare locations. Rev esp cir oral maxilofac 37: 108-112.

6. Qiu JY, Liu P, Shi C, Han B (2015) Low-grade myofibroblastic sarcomas of the maxilla. Oncology letters 9: 619-625.

7. Maruyama T, Nakasone T, Nimura F, Matayoshi A, Kawano T, et al. (2017) Indolent growth of low-grade myofibroblastic sarcoma of the cheek mimics benign lesions: A case report and literature review. Oncology letters 13: 4307-4314.

8. Montebugnoli L, Venturi M, Gissi DB, Flamminio F, Foschini MP (2010) Low-grade myofibroblastic sarcoma of the gingival. BMJ Case Reports 10: 1-4.

9. Diembi S, Moukassa D, Odzili FAI, Ngouoni CG, Dzon BO, et al. (2018) Low grade myofibroblastic sarcoma of the tongue: A case report in an infant. Open Journal of Pathology 8: 24-30.

10. Park KR, Jang HW, Won JH, Kim HS, Cha IH, et al. (2012) Myofibroblastic sarcoma of the mandible: A case report. J Korean Assoc Oral Maxillofac Surg 38: 240-244.

11. Demarosi F, Bay A, Moneghini L, Carrassi A (2009) Low-grade myofibroblastic sarcoma of the oral cavity. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 108: 248-254.