International Journal of Oral & Maxillofacial Pathology. 2013;4(1):85-89
Available online at http://www.journalgateway.com or www.ijomp.org
ISSN 2231 – 2250

Case Report

Odontogenic Myxoma of Oral Cavity: A Case Report

Nidhi Sharma, Dolly Prashant, Saurabh Sharma, Viplav Prashant

Abstract

Odontogenic myxoma is a relatively rare neoplasm which is almost exclusively seen in tooth bearing areas. They constitute around 3-6% of total odontogenic tumors. Odontogenic myxoma occurs commonly in mandible and their presentation in maxilla is rare. This article presents a rare case of odontogenic myxoma occurring in the maxilla of an 18 years old male patient with a brief review of its pathogenesis, clinical, histopathological and immunohistochemical characteristics.

Keywords: Odontogenic Myxoma; Fibromyxoma; Odontogenic Tumors; Connective Tissue; Maxilla.

Nidhi Sharma, Dolly Prashant, Saurabh Sharma, Viplav Prashant. Odontogenic Myxoma of Oral Cavity: A Case Report. International Journal of Oral & Maxillofacial Pathology; 2013:4(1):85-89.

Received on: 01/02/2013 Accepted on: 03/06/2013

Introduction

Odontogenic Myxoma (OM) are benign tumors derived from embryonic mesenchymal elements of dental anlage.[1,2] OM appears to originate from dental papillae, follicle or periodontal ligament. The evidence for its odontogenic origin arises from its almost exclusive location in the tooth bearing areas of the jaws, its occasional association with missing or unerupted teeth, and the presence of odontogenic epithelium.[1,3] According to the World Health Organization, OM is classified as benign tumor of ectomesenchymal origin with or without odontogenic epithelium.[1]

It was probably first described by Rudolf Virchow as myxofibroma in 1863.[4] In 1992, WHO defined OM as a locally invasive neoplasm consisting of rounded and angular cells lying in an abundant mucoid stroma.[5] OM represents an uncommon benign neoplasm comprising of 3-6% of all odontogenic tumors. It is a sporadic neoplasm with an annual incidence of 0.07 per million.[6] The occurrence of OM in the maxilla is rare when compared to the mandible(1:2) and slight predilection to females can be noted.[2,7] Commonly, OM occur in molar and premolar region in the mandible (65.1% of total mandibular cases), while in the maxilla the occurrence in these areas is around 73.8%.[2] OMs usually are unilateral lesions and they rarely cross the midline. Farman et al, have put the mean age at the time of diagnosis of maxillary OMs in men is around 29.2 years.[8]

OM is a locally invasive benign neoplasm. The invasiveness is attributed to the biological nature of tumour. The OM exhibits abundant extracellular production of ground substance and thin fibrils by the delicate spindle shaped cells. These undifferentiated mesenchymal cells are capable of fibroblastic differentiation also.[9-11] Depending upon the pattern of differentiation; the histological nature of the tumor varies. It may have complete myxomatous tissue or varying proportions of myxomatous and fibrous tissue. In the latter case it can be designated either as odontogenic fibromyxoma, in which the myxomatous element predominates; or odontogenic myxofibroma with predominance of fibrous tissue. Some regard OM as a modified form of fibroma in which the myxoid intercellular substance separates the connective tissue.[4,11]

Clinically, OMs are slow-growing, painless and site-aggressive tumors. Since pain and hypoesthesia are not common, the lesions may reach a considerable size before patient perceives its existence and seeks treatment. Larger lesions may cause tooth displacement and cortical bone expansion.[12,13] Radiologically, the appearance may vary from a unilocular radiolucency to a multicystic lesion with well-defined or diffused margins with fine, bony trabeculae within its interior structure expressing a “honey combed”, ”soap-bubble” or “tennis racket” appearance.[1,14] A unilocular appearance may be seen more commonly in children and in anterior parts of the jaws. Root resorption is rarely seen, and

the tumor is often scalloped between the roots.[13] The treatment of choice for OM is surgical excision by enucleation, curettage, or block resection. OM carries a high recurrence rate. Due to poor follow up and lack of reports a precise and accurate recurrence rate is still missing. The high recurrence rate of 25% is reported when more conservative treatments are used.[15]

Case Report

An 18 years old male patient reported with swelling in his gums since 5 months. Extraorally, swelling extended from nasal septum till left ala of nose. On palpation, swelling was firm in consistency and tender having irregular and non-indurated margins. Intraorally, the swelling was reddish in color present at left maxillary anterior region extending from maxillary right central incisor to maxillary left canine. Benign odontogenic tumors were considered and swelling was surgically excised.

Gross examination showed lesion of 6 x 4cm size. Further palpitation of the lesion showed that it was soft in consistency and the cut surface showed greyish-white appearance. The hematoxylin and eosin (H&E) stained section of the lesion revealed loose stellate cells (Fig 1a) with long branching cytoplasmic processes with bland and uniform nuclei were seen, the connective tissue stroma was myxoid and hyper cellular (Fig 1b). No other special stains were used. On immunohistochemistry, tumor cells showed strong immunoreactivity for Vimentin (Fig 1c) and SMA (Fig 1d & e) and focal immunoreactivity for S-100 protein (Fig 1f). They were immunonegative for Bcl-2, P-53, CK-19 and Ki-67. On basis of clinical, histopathological and immunohistochemical analysis diagnosis of odontogenic myxoma was made.

Discussion

Odontogenic myxoma is a rare aggressive intraosseous lesion derived from embryonic mesenchymal tissue associated with odontogenesis and primarily consisting of a myxomatous ground substance with widely scattered undifferentiated spindled mesenchymal cells.[16] Though it is a benign neoplasm, it may be infiltrative, aggressive and may recur.[17-20]

The prevalence of OM is principally quoted between 0.04% and 3.7%.[18] In Asia, Europe and America, relative frequencies between 0.5% and 17.7% have been reported.[21] There was lack of uniformity in the most common age group studies of OM but most of the studies showed 22.7 to 36.9 years and it is rarely seen in patients younger than 10 years of age or older than 50 years.[22,23] The mandible appears to be more frequently affected than maxilla.[7,22,24] Farman et al differentiated between maxillary and mandibular odontogenic myxoma and suggested that the mean age at the time of diagnosis of maxillary OM in men was 29.2 years and in women was 35.3 years, while the mandibular OM in men occur at a mean age of 25.8 years and in women they occur at 29.3 years.[8] Gunhan et al[25] and Regezi et al[29] reported a higher incidence of these tumors in women (64-95%) than in men. In present case, the tumor was present in 18 years old male patient involving the maxilla, extending from upper right central incisor to upper left canine.

There are no clinical or radiological signs that would allow a physician to distinguish myxoma from odontogenic and non-odontogenic lesions; however, histological analysis shows several lesions that could be misinterpreted as myxoma.[26] Myxoma of the head and neck are rare tumors. Two forms have been identified: (1) facial bone derived myxoma, which had been subdivided in the past into true osteogenic myxoma and odontogenic myxoma and (2) ”soft tissue”-derived myxoma, derived from the perioral soft tissue, parotid gland, ear and larynx.[27]

There has been a great deal of controversy regarding the origin of myxomatous tumors. Virchow, in 1863, coined the term myxoma for a group of tumors that had histologic resemblance to the mucinous substance of the umbilical cord.[13] Traditionally, the myxoma of the maxilla and mandible has been considered to be a neoplasm of odontogenic origin. Although the evidence is mainly circumstantial, support of an odontogenic origin has been perpetuated by its almost exclusive occurrence in the tooth bearing areas of the jaws, its common association with an unerupted tooth or a developmentally absent tooth, its frequent occurrence in young individuals, its histologic resemblance to dental mesenchyme, especially the dental papilla and occasional presence of sparse amounts of odontogenic epithelium.[28]

Slootweg and Wittkampf, on the other hand, showed that the matrix of myxomas of the jaw is entirely different from the matrix seen

in the dental pulp and periodontal ligament. According to them, even the presence of odontogenic epithelium is not necessary to make the diagnosis of myxoma of bone.[18] Contrary to the findings of Slootweg and Wittkampf, McClure and Dahlin reviewed more than 600 bone tumors of patients of Mayo clinic and concluded that there were no true myxomas of the bone except for those found in the mandible and maxilla.[29]

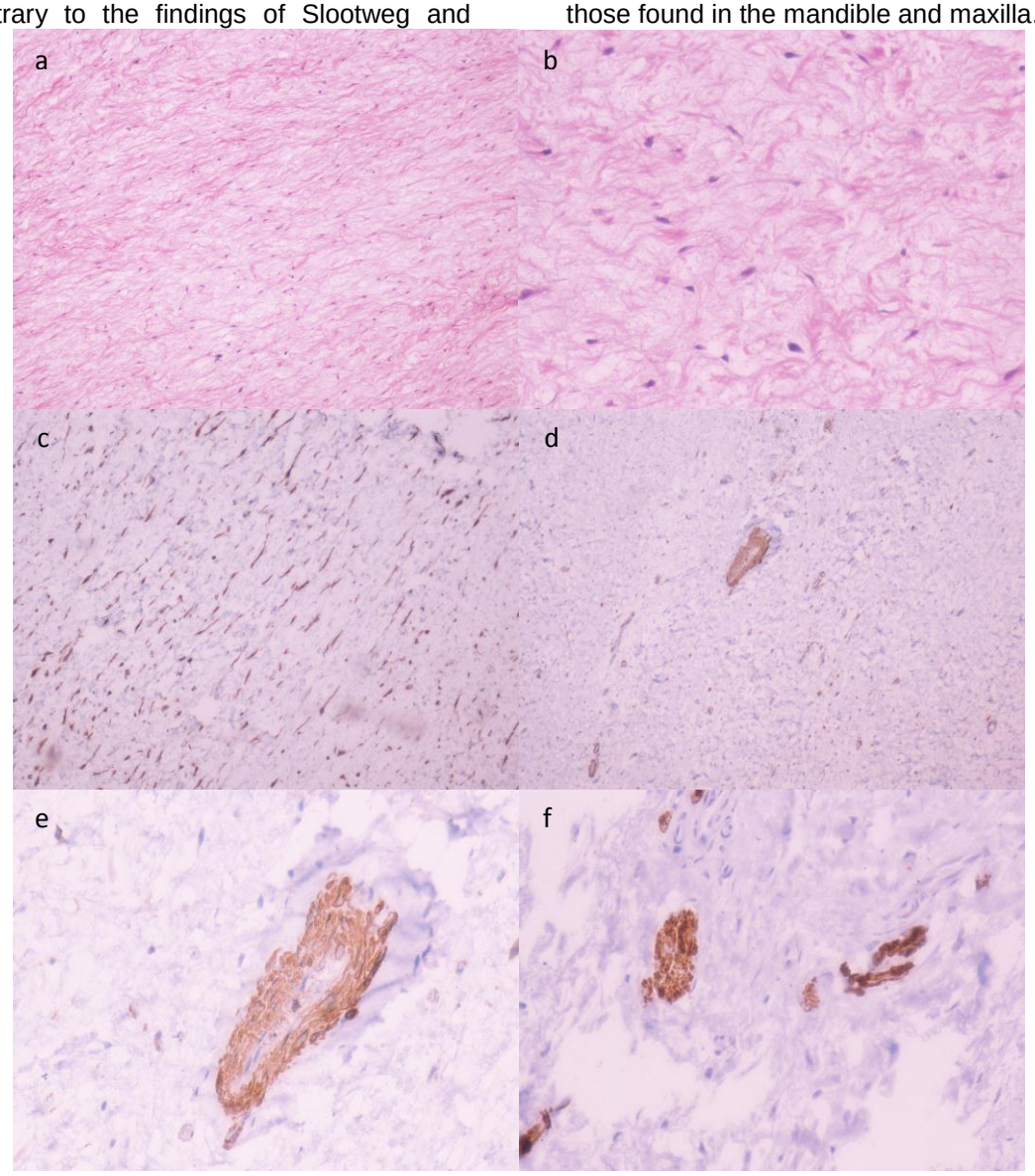

Figure 1: The Photomicrograph of hematoxylin and eosin stained sections shows loose stellate cells with long branching cytoplasmic processes at low power (a) and hypercellular and myxoid connective tissue stroma at high power (b). The immunohistochemical staining shows strong immunopositivity for vimentin (c), for SMA at low (d) & high power (e) and focal immunoreactivity for S-100 (f).

Most OMs are first noticed as a result of a slowly increasing swelling or asymmetry of the affected jaw. Lesions are generally painless and ulceration of the overlying oral mucosa only occurs when the tumour interferes with dental occlusion. Growth may be rapid and infiltration of neighboring soft tissue structure may occur. Both the buccal and the lingual cortical plates of the mandible may expand occasionally.[21] Kaffe et al, found expansion of the jaws in 74% of the cases.[30] When the maxillary sinus is involved, the OMs often fill the entire antrum. Displacement of teeth has been registered in 9.5% of the cases. Both the features were appreciated in the present case. In the present case, there was firm and tender swelling present in gums and extraorally the swelling extended from nasal septum till left ala of nose. OM is a benign neoplasm without encapsulation. A spectrum of fibrous connective tissue stroma is present from myxoid to densely hyalinized and from relatively acellular to cellular.[31] Calcification may or may not be present. It is

distinguished by the presence of sparse cords and islands of inactive odontogenic epithelium.[20] The histopathological examination in the present case showed presence of loose stellate cells with long branching cytoplasmic processes with bland and uniform nuclei with myxoid and hyper cellular connective tissue stroma.

An extensive study on the ultrastructure of odontogenic myxoma was published by Goldblat in 1976. Two basic types of tumour cells were described: secretory and non secretory. The secretory cell type was considered the principal tumor cell and resembles fibroblasts[4] whereas the non-secretory cells had morphological and functional criteria of a so called myxoblast. While generally considered a slow growing neoplasm, OMs may be infiltrative and aggressive, with high recurrence rates. Because of poor follow-up and lack of reports, a precise analysis of recurrence rates is still missing. Treatment of OM varies from local excision, curettage or enucleation to radical resection. Recurrence is considered to be directly related to the type of therapy, with conservative surgery resulting in a higher number of reccurences.[4]

In a recent immunohistochemical and ultrastructural study, Moshiri et al, supported the notion of odontogenic origin of myxoma by suggesting that fibroblasts that compose the tooth germ undergo modification to give rise to OM.[15] An immunohistochemical panel of polyclonal and monoclonal antibodies were used to characterize and distinguish the nature of cells as fibroblastic, histiocytic, myoblastic and neural origin. Three types of odontogenic myxoma cells were discriminated: spindle cells, stellate cells and hyaline cells. Neoplastic cells of myxoma were positively stained for transferrin, ferritin, alpha-1-antichymotrypsin (alpha-1-ACT), alpha-1-antitrypsin (alpha-1-AT), S-100 protein, vimentin (pan-mesenchymal marker) and actin; however, neuron specific enolase (NSE), S-100 alpha subunit, S-100 beta subunit, factor VII related antigen (FVII-AG) and cytokeratin (CKI) were negative.[1,20] Antibodies directed against vimentin are used to identify mesenchymal cells, and leatin antibodies detect epithelial differentiation. Spindle cells were positive for transferrin, ferritin, alpha-1-ACT, alpha 1-AT, S-100 protein and vimentin. Stellate cells were strongly positive for transferrin, alpha-1-AT, S-100 protein and vimentin. Hyaline cells reacted with alpha-1-ACT and alpha-1-AT. Myxomatous matrix showed negative reaction for all the antibodies used. These results have confirmed that OM is a tumor of a dual fibroblastic-histiocytic origin and also suggest that the cells comprising OM are of myofibroblastic origin.[7,19,20] The immunohistochemical panel in the present case showed strong immunoreactivity for vimentin and SMA and focal immunoreactivity for S-100 protein and immunonegative for Bcl-2, p-53, CK-19 and Ki-67 confirming the diagnosis of OM.

Acknowledgement

We would like to acknowledge all the staff members form the department of oral and maxillofacial pathology for their support and guidance.

Author Affiliations

1.Dr.Nidhi Sharma, Assistant Professor, Department of Oral Pathology, 2.Dr.Dolly Prashant, Assistant Professor, Department of Pathology, Government Dental College, Raipur, 3.Dr.Saurabh Sharma, Assistant Professor, Department of Orthodontics & Dentofacial Orthopaedics, Chhattisgarh Ayush & Health Science University, Maitri College of Dentistry & Research Centre, Anjora, Durg, Chhattisgarh, 4.Dr.Viplav Prashant, ex. Senior registrar, Department of Pathology, pt.ravishankar shukla university, Pt.JNM Medical college, Raipur, Chhattisgarh, India.

Reference

1. Lo Muzio L, Nocini P, Favia G, Procaccini M, Mignogna MD. Odontogenic myxoma of jaws: A clinical, radiologic, immunohistochemical and ultrastructural study. Oral Surg Oral Med Oral Pathol Radiol Endod 1996;82:426-33.
2. Peltola J, Magnusson B, Happonen RP, Borrman H. Odontogenic myxoma - a radiographic study of 21 tumors. Br J Oral Maxillofac Surg 1994;32:298-302.
3. Asaumi J, Konouchi H, Hisatomi M, Kishi K. Odontogenic myxoma of maxillary sinus: CT and MRI-pathologic correlation. Eur J Radiol 2001;37:1-4.
4. Reichart PA, Philipsen HP. Odontogenic tumors and allied lesions. London: Quintessence Publishing; 2004. 189-96p.
5. Kramer IRH, Pindborg JJ, Shear M. Histologic Typing of Odontogenic tumors. 2nd ed. Berlin: Springer-Verlag; 1992.
6. Simon EN, Merkx MA, Vuhahula E, Ngassapa D, Stoelinga PJ. Odontogenic myxoma: A clinic pathological study of

33 cases. Int J Oral Maxillofac Surg 2004;33:333-7.
7. Abiose BO, Ajagbe HA, Thomas O. Fibromyxoma of the jaw bones: A study of ten cases. Br J Oral Maxillofac Surg1987;25:415-21.
8. Farman AG, Nortje CJ, Grotepass FW, Farman FJ, vanZyl JA. Myxofibroma of the jaws. Br J Oral Surg 1977;15:3-18.
9. Regizi JA, Scuiba J. Oral pathology clinical-Pathologic correlations. WB Saunders Company Ltd: 2003. p278-9.
10. Zimmermann DC, Dahlin DC. Myxomatus tumors of the jaws. Oral Surg Oral Med Oral Pathol 1958;11:1069-80.
11. Adekeye EO, Avery BS, Edwards MB, Williams HK. Advanced central myxoma of the jaws in Nigeria: Clinical features, treatment and pathogenesis. Int J Oral Surg 1984;13:177-86.
12. Bast BT, Pogrel MA, Regezi JA. The expression of apoptotic proteins and matrix metalloproteinases in odontogenic myxoma. J Oral Maxillofac Surg 2003;61:1463-6.
13. Landa LE, Hedrick MH, Nepomuceno-Perez MC. Recurrent Myxoma of the Zygoma: A case report. J Oral Maxillofac Surg 2002;60:764-8.
14. Regezi JA, Kerr DA, Courtney RM. Odontogenic tumors: Analysis of 706 cases. J Oral Surg 1978;36:771-8.
15. Moshiri S, Oda D, Worthington P, Myal lR. Odontogenic myxoma: Histochemical and Ultrastructural study. J Oral Pathol Med 1992;2:401-3.
16. Kim J, Ellis GL. Dental follicular tissue: Misinterpretation as odontogenic tumors. J Oral Maxillofac Surg 1993;51:762-8.
17. Chuchurru JA, Luberti R, Cornicelli JC, Dominguez FV. Myxoma of the mandible with unusual radiographic appearance. J Oral Maxillofac Surg 1985;43:987-90.
18. Slootwegv PJ, Wittkampf RM. Myxoma of the jaws: An analysis of 15 cases. J Maxillofac Surg 1986;46-52.
19. Green TL, Leighty SM, Walters R. Immunohistochemical evaluation of oral myxoid lesions. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1992;73:407-71.
20. Zhao M, Lu Y, Takata T, Ogawa I, Miyauchim, Mock D, et al. Immunohistochemical and histochemical characterization of the mucosubstances of odontogenic myxoma: Histogenesis and differential diagnosis. Pathol Res Pract 1999;195:391-7.
21. Lu Y, Xuan M, Takata T, et al. Odontogenic tumors: a demographic study of 759 cases in a Chinese population. Oral Surg Oral Med Oral Pathol Oral Radiol and Endod 1998;86(6):707-14.
22. Sivakumar G, Kavitha B, Saraswathi T, Sivapathasundharam B. Odontogenic myxoma of maxilla. Indian J Dent Res 2008;19(1):62-5.
23. Ajayi OF, Ladeinde AL, Adeyemo WL, Ogunlewe MO. Odontogenic tumors in Nigerian children and adolescents - a retrospective study of 92 cases. World J Surg Oncol 2004; 2, article 39.
24. Lin YL, Basile JR. A case of odontogenic myxoma with unusual histological features mimicking a fibro-osseous process. Head Neck Pathol 2010;4(3):253-6.
25. Gunhan O, Erseven G, Ruacan S, Celasun B, Aydintug Y, Ergun E, et al. Odontogenic tumors: A series of 409 cases. Aust Dent J 1990;35:518-22.
26. Rocha, Gaujac C, Ceccheti MM, Amato-Filho G, Machado GG. Treatment of recurrent mandibular myxoma by curettage and cryotherapy after thirty years. Clinics 2009;64(2):149-52.
27. Gnepp D. Diagnostic Surgical pathology of Head and Neck. Philadelphia; W.B. Saunders Co; 1997.
28. Halfpenny W, Verey A, Bardsley V. Myxoma of the mandibular condyle: A case report and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2000;90:348-53.
29. Mc Clure DK, Dahlin DC. Myxoma of bone: Report of three cases. Mayo Clin Proc 1977;52:249-53.
30. Kaffel, Naor H, Buchner A. Clinical and radiological features of odontogenic myxoma of the jaws. Dento Maxillofac Radiol 1997;26:299-303.
31. Gormley MB, Mallin RE, Solomon M, Jarrett W, Bromberg B. Odontogenic myxofibroma: Report of two cases. J Oral Surg 1975;33:356-9.

Corresponding Author
Dr.Dolly Prashant,
A-1, Rajiv Nagar, Raipur,
Chhatisgarh 492007, India.
Ph: +91 9630008003
E-mail: dollydolly03@rediffmail.com

Source of Support: Nil, Conflict of Interest: None Declared.