

Odontogenic Fibromyxoma: A Diagnostic Dilemma

Shivangi Singh¹, Sonika Singh⁴, Ankit Sachdeva³, Komal Gupta⁴

1-Sr. Lecturer, Department of Oral Medicine and Radiology, ITS Dental College and Research Centre, Ghaziabad, Uttar Pradesh. 2,3,4-Post graduate student, Department of Oral Medicine and Radiology, Shree Bankey Bihari Dental College and Research Centre, Ghaziabad, Uttar Pradesh.

Correspondence to:
Dr. Shivangi Singh, Sr. Lecturer, Department of Oral Medicine and Radiology, ITS Dental College and Research Centre, Ghaziabad, Uttar Pradesh.
Contact Us: www.ijohmr.com

ABSTRACT

Odontogenic fibromyxoma is a slow growing, locally invasive, non metastasizing neoplasm found exclusively in the bones of the facial skeleton. Although benign, it is locally very aggressive making it difficult to eradicate. It commonly occurs in the second and third decade, and the mandible is involved more commonly than the maxilla. The lesion often grows without symptoms and presents as a painless swelling. The radiographic features are variable, and the diagnosis is therefore not easy in many cases. This article presents a rare case of Odontogenic fibromyxoma occurring in the posterior mandible of a twenty year old male patient with the review of literature.

KEYWORDS: Fibromyxoma, Odontogenic Tumor, Multilocular Radiolucency, Lichen Planus of jaw

INTRODUCTION

Odontogenic myxoma (OM) is a rare tumor that occurs in the mesenchymal tissue of the the mandible and maxilla and represents 3 – 6% of all odontogenic tumors.¹ Fibromyxomas are benign but aggressive mesenchymal tumors that are slow growing, expansile and locally destructive². It usually occurs in the 2nd and 3rd decade of life, rarely in children or adults over 50 years of age.³ They occur commonly in women with a mean age of 31 years.² The tumor occurs almost equally in the maxilla and mandible with a slight predilection for the posterior mandible.⁴ Clinically it presents as a slow growing asymptomatic swelling, however, pain and paresthesia can occur in advanced stages. Facial asymmetry may occur due to large lesions. Displacement and mobility of teeth are relatively common. It may be associated with unerupted teeth. Cortical expansion can occur and large lesions can cause perforation.⁵ Radiographically, OM most commonly presents a unilocular or multilocular, well-defined radiolucency.⁶ In this article, we present a case of Odontogenic fibromyxoma alongwith focal reparative giant cell granuloma with its review of literature and the contribution of the radiological examination to the differential diagnosis and the importance of a meticulous enucleation in order to prevent recurrence.

CASE REPORT

A 21 year old male patient (Fig.1) reported with a chief complaint of swelling in left lower back teeth region since 1 month which appeared following atraumatic extraction of a mobile tooth in the left mandibular posterior region. The swelling was peanut in size to begin with and had gradually attained the present size. There was pus discharge from the swelling which subsided on

medication. There was no history of anesthesia or paresthesia. The swelling was followed by pain which was intermittent, piercing and localized and got relieved on medication. No significant medical anamnesis was reported.

On clinical examination, a diffuse oval shaped swelling measuring approximately 5cm × 4cm in its greatest dimensions, was present extraorally on the lower third of the face extending from 1cm to the left corner of mouth to left angle of the mandible anteroposteriorly. Superoinferiorly, it was extending from line joining the corner of mouth and ear lobe to 1cm below inferior border of the mandible. Overlying skin was normal (Fig.1). On palpation, it was soft and nontender with local



Figure 1: - Extra oral view of patient showing ill defined swelling on left side of face.

How to cite this article:

Singh S, Singh S, Sachdeva A, Gupta K. Odontogenic Fibromyxoma: A Diagnostic Dilemma. *Int J Oral Health Med Res* 2016;3(1): 111-115.

rise in temperature. A solitary left submandibular lymph node was palpable roughly oval in shape, firm in consistency, tender, and mobile. Intraorally there was a solitary diffuse ill – defined exophytic growth having irregular shape measuring 4cm x 3cm in its greatest dimensions extending mesiodistally from distal aspect of 36 to retromolar area and mediolaterally from lingual vestibule to buccal vestibule (Fig.2). There was interspersed ulcerated area. On palpation it was pedunculated with its base arising lingually, firm in consistency and nontender. There was mild bleeding on probing. The buccal and lingual cortices were expanded with grade II mobility in relation to 36. Based on the history and clinical features a provisional diagnosis of epulis granulomatousum was given.

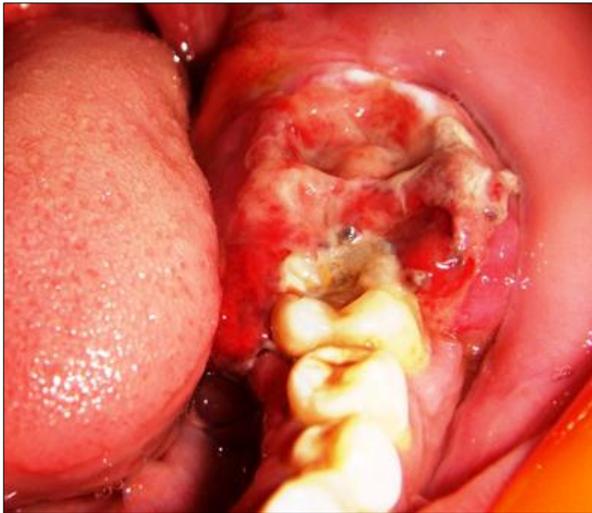


Figure 2:- Exophytic growth present distal to 36.

Panoramic radiograph was taken as the size of the lesion was large, which revealed a solitary well defined multilocular radiolucent lesion on the left mandible roughly oval in shape measuring about 4cm x 2cm extending anteroposteriorly from the mesial root of 36 to distal aspect of 38 and superoinferiorly from 0.5cm below the distal root of 36 to 1cm above the inferior border of mandible. The borders were well defined with loss of cortication seen in the anterior aspect of the lesion. Internal structure revealed multiple areas of increased radiolucency separated by few thin curved septas giving it a soap bubble appearance. Mandibular canal could not be traced in anterior part of the lesion, but it was slightly displaced inferiorly in the posterior aspect. 37 was missing, and root formation had not completed in 38, there was resorption of distal root of 36 (Fig.3). Intraoral periapical radiograph revealed a well-defined oval shaped radiolucency of size 3cm x 2cm apical to 36. The radiolucency had sclerotic borders with few thin curved and straight septas within the radiolucency giving it a multilocular appearance. Root resorption was seen in relation to distal root of 36 (Fig.4). Odontogenic myxoma was considered as radiographic diagnosis and ameloblastoma and central giant cell granuloma as radiographic differential diagnosis for the case.



Figure 3:- Panoramic radiograph showing multilocular radiolucency

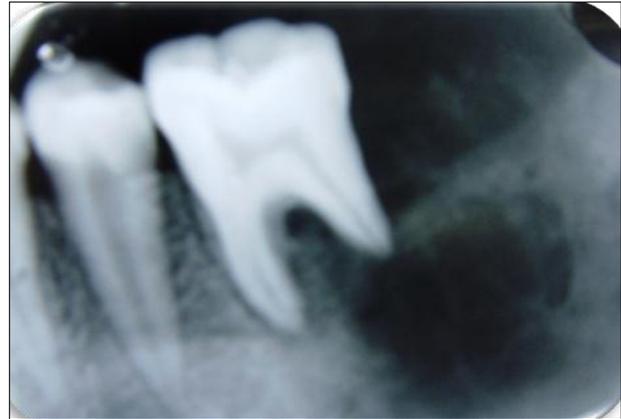


Figure 4:- Intra oral periapical radiograph showing radiolucency apical to 36

Sanguineous material was obtained on aspiration which showed chiefly blood and fibrin with only a few leucocytes, a small number of benign squamous and polygonal epithelial cells. Enucleation of the lesion was carried out, and the specimen was sent for histopathological examination which revealed clusters of multinucleated osteoclast like giant cells mixed with mononuclear oval to spindle cells. In deeper areas of tissues, the tumor is seen composed of scattered plump oval to spindle and stellate cells lying within the myxoid stroma. The myxoid areas were divided into lobules of fibrous bands. New lamellar bone formation with along with bone resorption was seen in some pieces suggestive of Odontogenic fibromyxoma alongwith focal reparative giant cell granuloma (Fig.5). The patient is on follow up (Fig.6).

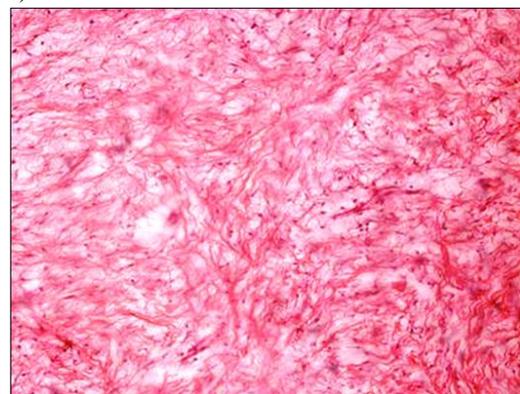


Figure 5:- Photomicrograph showing histological features (Hematoxylin-Eosin stain X 10x)



Figure 6:- Post operative panoramic radiograph

DISCUSSION

Myxomas are benign soft tissue or bone neoplasms that may appear anywhere in the body. In the head and neck region they have been specifically found in the: (1) tongue; (2) nose; (3) cheek; (4) neck muscles; (5) larynx; (6) pharynx; and (7) parotid gland.⁷ Most central myxomas occur in the jaws, where they are called odontogenic myxomas (OM) because of their presumably odontogenic origin—although their histologic origin is still controversial.⁸ Odontogenic myxoma (OM) is classified as a benign tumor of ectomesenchymal origin with or without odontogenic epithelium. It is defined as — “a locally invasive neoplasm consisting of rounded and angular cells lying in an abundant mucoid stroma” according to the World Health Organization.⁹ Fibromyxoma is classified as a specific type of myxoma with a higher fibrous/myxoid tissue ratio than myxoma. There is a discrepancy regarding the reports of fibromyxoma, as many of them are classified under the general term “myxoma”, making the review of the literature difficult.¹⁰

According to Dutz and Stout, the term myxoma was first used by Virchow in 1863, but the term fibromyxoma was described by Marcove et al. in 1964 who reported extragnathic locations of fibromyxoma.^{11,12}

The histologic similarity to stellate reticulum of a developing tooth, its association with a missing or unerupted tooth, odontogenic epithelium may be present in a minority of cases and the fact that it rarely appears in other parts of skeleton often supports its odontogenic origin. In a recent immunohistochemical and ultrastructural study, Moshiri et al. supports the odontogenic origin of myxomas by suggesting that fibroblasts that compose the tooth germ undergo modification to give rise to odontogenic myxoma.¹³

Slootweg and Wittkamp 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. In addition, they also argued that myxomas may also develop in the sinonasal tract and other facial bones that originate from the nonodontogenic mesenchyme. According to them, even the presence of odontogenic epithelium is not necessary to make the diagnosis of myxoma of bone.¹⁴

Aibose et al. reported that fibromyxomas constituted 3.73% of all benign and malignant oral tumors and 20% of tumors of dental origin, second in incidence to ameloblastoma. Most arise from second and third decade of life with an age range varying from 22.6 – 36.9 years.¹⁵ However, James et al. reported the case of a maxillary myxoma in a child of 11 months.¹⁶ Interestingly, Keszler observed a higher frequency of this neoplasm than other aggressive tumors in children. He concluded that OM should be considered in the differential diagnosis of radiolucent lesions in children and adolescents.¹⁷

Fibromyxomas are usually located intraorally most often in the posterior regions of the mandible, its angle and ramus and rarely extraorally with ratios of 3:1. The female to male ratio is 1.5:1 although some reports state no sex predilection and equal frequencies in maxilla and mandible.⁹ Nevertheless, most reports indicate that the posterior region of jaw is the most commonly affected site.¹⁸ Besides the alveolar process, maxillary involvement may include the zygomatic processes. Mandibular involvement, on the other hand, may include the posterior body of the mandible, angle, and ramus. Moreover, the OM is localized on one side of the jaw and rarely crosses the midline.¹⁹ According to Kaffe, in only 5% of the cases is OM associated with an unerupted tooth.²⁰ In the present case also the age of the patient was 21 years, and the lesion was located in the posterior mandible which was in accordance with that reported in literature.

Clinically it presents as a slow growing asymptomatic swelling, however, pain and paresthesia can occur in advanced stages. Facial asymmetry may occur due to large lesions. Displacement and mobility of teeth are relatively common although root resorption may be rare.⁹ It may be associated with unerupted teeth. Cortical expansion can occur and large lesions can cause perforation.⁵ All these features were in accordance with the present case.

WHO classified it as multiple radiolucent areas of varying size, separated by straight or curved bony septa with poorly defined borders.⁹ On conventional radiographs, myxomas of the jaws often show multilocular radiolucencies which helps in distinguishing this entity from malignant tumors arising centrally within the jaw bones, because the latter usually cause massive bone destruction without compartments by bony trabeculations or bony septa.²¹ The internal trabecular pattern has been described as “honey- comb,” “soap-bubble,” or “tennis racquet.”⁶ The latter appearance is characterized by angular or straight trabeculations forming square or triangular compartments²² and has been considered almost pathognomonic of OM. Eversole (1980) said that the internal configuration of the bony speta resembled “lichen planus of the jaw bone”.²³ In the present also, panoramic radiograph revealed a large well defined multilocular radiolucency having soap bubble appearance with bony septa.

Computed tomographic images of odontogenic myxomas may show any of the following features:

- Osteolytic expansile lesions with mild enhancement of the solid portion of the mass in the myxoma of the mandible.
- Bony expansion and thinning of cortical plates with strong enhancement of the mass lesion in the anterior maxilla.
- A soft tissue mass with bone destruction and thinning and strands of fine lace-like density representing ossifications in the maxillary sinus.²⁴

Magnetic resonance imaging (MRI) revealed a well defined, well-enhanced lesion with homogeneous signal intensity on every pulse sequence. The lesion showed intermediate signal intensity on the T1-T2- weighted images. Unfortunately, CT and MRI were not performed in this case.²⁵

OM should be included in the differential diagnosis of both radiolucent and mixed lesions, in both the jaws, for individuals of all age groups. When unilocular and without trabeculae, the tumor closely resembles periapical, lateral, periodontal, and traumatic bone cysts. When multilocular, it must be distinguished from ameloblastoma, central hemangioma and Odontogenic keratocyst.²⁶

Upon gross examination, OM appears as a white or yellow, gelatinous, lobulated mass.¹⁸ Histologically, it is rarely encapsulated and is composed of spindle-shaped and stellate cells interspersed in the loose mucoid background. Collagen fibers may also be seen scattered in the mucopolysaccharide ground substance, and their amounts determine the tumor's texture and whether it is called myxoma or myxofibroma. Odontogenic epithelium may occasionally be found, but its role as a tumor-inducing agent is controversial and, thus, its presence is not a requirement for the diagnosis of OM.²⁷

OM tumor cells are mesenchymal in origin and express vimentin and muscle-specific actin. Conflicting description of S-100 and GFAP positivity has been reported. The matrix exhibits different proteins, mostly type-I and type IV collagen, fibronectin, and proteoglycans.²⁸

An extensive study on the ultrastructure of OM was published by Goldblat in 1976. Two basic types of tumor cells were described, secretory and nonsecretory. The secretory cell type was considered the principal tumor cell and resembled fibroblasts.²⁹

The tumor is not radiosensitive, and surgery is the treatment of choice.³⁰ The generally accepted treatment for OM includes resection of the tumor with a greater than 1.5 cm margin of surrounding tissue.¹⁸ The lack of a capsule and infiltrative growth pattern is responsible for high recurrence rate (upto 25%) when conservative enucleation and curettage are performed. Prognosis of myxomas of the jaw is generally good. Recurrence typically occurs during the first 2 years after removal

although recurrence has been described over 30 years after original surgery.³¹

CONCLUSION

This case highlights the wide variety in clinical and radiologic appearance of odontogenic myxomas, being the most common form of presentation as an asymptomatic expansion in the jaw and a multilocular radiolucent image and its importance of including myxoma in the differential diagnosis of radiolucent multilocular lesions in the maxilla or mandible. Since, odontogenic myxomas poses a diagnostic and therapeutic challenge hence, correlating clinical, radiological and histopathological features is essential. For the successful management of the myxomatous tumours, a complete surgical excision along with proper long term follow up is essential keeping in mind the high recurrence rate.

REFERENCES

1. Dalbo Contrera Toro M, Siqueira Barreto I, Amstalden EM, Takahiro Chone C, Nizam Pfeilsticker L. Odontogenic Myxoma in Children: A Case report and Literature Review. *Case Rep Oncol Med*. 2016;9017421.
2. Shivakumar GC, Sahana S. Fibromyxoma of the Maxilla – A case report. *Int J Oral Med Sci*. 2010; 9(1):36 – 41.
3. Deitrich EM, Papaemmanouil S, Koloutsos G, Antoniadis H, Antoniadis K. Odontogenic Fibromyxoma of the Maxilla: A Case Report and Review of the Literature. *Case Report Med*. 2011. 238712
4. Craig GT. The Paradental Cyst. A specific inflammatory odontogenic cyst. *Br Dent J*. 1976; 141: 9 – 14.
5. Brannon RB. Odontogenic Fibroma, myxoma (odontogenic myxoma, fibromyxoma), and central odontogenic granular cell tumor. *Oral Maxillofac Surg Clin North Am*. 2004; 16(3): 359 – 374.
6. MacDonald-Jankowski DS, Yeung RW, Li T, Lee KM. Computed tomography of odontogenic myxoma. *Clin Radiol*. 2004; 59:281-7.
7. Robledo J, Alderson GL, Jones AC, McGuff HS, Peterson T, Potter M. Oral and maxillofacial pathology case of the month. Odontogenic myxoma. *Tex Dent J*. 2003; 120(7): 616-21.
8. Fujita S, Hideshima K, Ikeda T. Nestin expression in odontoblasts and odontogenic ectomesenchymal tissue of odontogenic tumors. *J Clin Pathol*. 2006; 59(3): 240-5.
9. Gupta I, Keluskar V, Shetti A. Odontogenic fibromyxoma of left maxilla - A case report. *J Int Oral Health*. 201; 2(4): 79 – 86.
10. Dietrich EM, Papaemmanouil S, Koloutsos G, Antoniadis H, Antoniadis K. Odontogenic fibromyxoma of the maxilla: a case report and review of literature. *Case Report Med*. 2011. 238712.
11. Dutz W, Stout AP. The myxoma in childhood. *Cancer*. 1961; 14:629–635
12. Marcove RC, Kambolis C, Bullough PG, and Jaffe HL. Fibromyxoma of bone. A report of 3 cases. *Cancer* 1964; 17:1209–1213
13. Moshiri S, Oda D, Worthington P, Myall R. Odontogenic myxoma: Histochemical and ultrastructural study. *J Oral Pathol Med*. 1992; 21 (3): 401–3.
14. Slootweg PJ, Wittkampf RM. Myxoma of the jaws: An analysis of 15 cases. *J Maxillofac Surg*. 1986; 14(1):46-52.

15. Abiose BO, Ajagbe HA, Thomas O. Fibromyxomas of jaw bone- a study of 10 cases. *Br J Oral Maxillofac Surg.* 1987; 25(5):415-421.
16. James DR, Lucas VS. Maxillary myxoma in a child of 11 months. A case report. *J Craniomaxillofac Surg.* 1987; 15(1):42 – 44.
17. Keszler A, Dominguez FV, Giannunzio G. Myxoma in childhood: An analysis of 10 cases. *J Oral Maxillofac Surg.* 1995; 53(5):518-21.
18. Chiodo AA, Strumas N, Gilbert RW, Birt BD. Management of odontogenic myxoma of the maxilla. *Otolaryngol Head Neck Surg.* 1997; 117:S73-6.
19. Simon EN, Merckx MA, Vuhahula E, Ngassapa D, Stoelinga PJ. Odontogenic myxoma: A clinicopathological study of 33 cases. *Int J Oral Maxillofac Surg* 2004; 33(4):333-7.
20. Kaffe I, Naor H, Buchner A. Clinical and radiological features of odontogenic myxoma of the jaws. *Dentomaxillofac Radiol* 1997; 26(5):299-303.
21. Kawai T, Murakami S, Nishiyama H, Kishino M, Sakuda M, Fuhihata H. Diagnostic imaging for a case of maxillary myxoma with a review of the magnetic resonance images of myxoid lesions. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 1997;84(4):449-54.
22. Koseki T, Kobayashi K, Hashimoto K, Arijji Y, Tsuchimochi M, Toyama M, et al. Computed tomography of odontogenic myxoma. *Dentomaxillofac Radiol.* 2003; 32(3):160-5.
23. Langlais RP, Langland OE, Nortje CJ. Diagnostic Imaging of the Jaws. 1st edition, USA, Williams & Wilkins; 1995. pp 348 – 349.
24. Asaumi J, Konouchi H, Hisatomi M, Kishi K. Odontogenic myxoma of maxillary sinus: CT and MR -pathologic correlation. *Eur J Radiol* 2001;37:1-4.
25. Reichart PA, Philipsen HP. Odontogenic tumors and allied lesions. Illinois: Quintessence Publishing Co Ltd; 2004.
26. Chuchurru JA, Luberti R, Cornicelli JC, Dominguez FV. Myxoma of the mandible with unusual radiographic appearance. *J Oral Maxillofac Surg* 1985; 43(12):987-90.
27. Chiodo AA, Strumas N, Gilbert RW, Birt BD. Management of odontogenic myxoma of the maxilla. *Otolaryngol Head Neck Surg.* 1997; 117(6):S73-6.
28. Bucci E, Lo Muzio L, Mignogna MD, De Rosa G. Odontogenic myxoma: Report of a case with peculiar features. *J Oral Maxillofac Surg.* 1991; 49(1):91-4.
29. Gnepp DR. *Diagnostic Surgical Pathology of head and neck.* London: WB Saunders Company Ltd; 2001. p. 6
30. Farman AG, Nortje CJ, Grotepass FW, Farman FJ, Van Zyl JA. Myxofibroma of the jaws. *Br J Oral Surg* 1977; 15:3-18.
31. Lo Muzio L, Nocini P, Favia G, Procaccini M, Mignogna MD. Odontogenic myxoma of the jaws: A clinical, radiologic, immunohistochemical, and ultrastructural study. *Oral Surg Oral Med Oral Pathol Radiol Endod* 1996;82:426-33.

Source of Support: Nil
Conflict of Interest: Nil