Journal of Advanced Clinical & Research Insights (2018), 5, 88–91

CASE REPORT

A pediatric odontogenic myxoma of the maxilla: A case report and review

Preeti Singh[1], K. Shwetha Nambiar[1], Roopa S. Rao[1], Prathibha Shridhar[2], Kavitha Prasad[2], S. V. Sowmya[1], Dominic Augustine[1], Vanishri C. Haragannavar[1]

[1]Department of Oral Pathology and Microbiology, Faculty of Dental Sciences, Ramaiah University of Applied Sciences, Bengaluru, Karnataka, India, [2]Department of Oral and Maxillofacial Surgery, Faculty of Dental Sciences, Ramaiah University of Applied Sciences, Bengaluru, Karnataka, India

Keywords:
Maxilla, odontogenic myxoma, pediatric

Correspondence: Dr. Preeti Singh, Department of Oral Pathology and Microbiology, Faculty of Dental Sciences, Ramaiah University of Applied Sciences, Bengaluru – 560 054, Karnataka, India. E-mail: singhpreeti7389@gmail.com

Received: 09 April 2018
Accepted: 26 May 2018

doi: 10.15713/ins.jcri.219

Abstract

Odontogenic tumors represent a broad spectrum of lesions ranging from benign to malignant lesions. Benign odontogenic lesions are rare entities that are important due to their local aggressive nature and equally challenging to handle. Odontogenic myxoma (OM) in children is unusual and extremely rare. Till date, only 40 such cases have been reported in the literature with high recurrence rate between 10 and 33% with an average of 25%. We report a rare case of aggressive OM in the maxilla of a female child of 1 year and 8 months. Timely and apt histopathological diagnosis of OM enabled the surgeons to arrive at a conservative treatment plan considering esthetic and age of the child. No recurrence was noted on follow-up of the case for 6 months. The patient is kept under observation.

Introduction

Odontogenic myxoma (OM) of the jaws was first described by Goldman and Thoma in 1947. It represents 3%–6% of all odontogenic tumors.[1] The first to describe the histologic features of myxofibroma was German pathologist Virchow (1863), although the lesions of jaws were not specifically mentioned..[2] The World Health Organization (WHO) in 2003 classified "OM as a benign neoplasm arising from odontogenic ectomesenchyme with or without odontogenic epithelium." The odontogenic nature of the myxomas has been questioned by some authors because of the appearances that represent a more primitive fibroblastic or undifferentiated tissue.[3] OM can occur both in bone and soft tissue.[2,3] Although intraosseous myxoma has been reported in various anatomical sites, in head and neck region, the majority of these occur in mandible followed by maxilla.[3-5] Melo *et al.* (2008) showed that there is higher prevalence of mandibular tumors (56.56%) than that of maxillary tumors (42.16%) with the ratio of mandible: maxilla 1.34:1. Similarly, Nonaka *et al.* (2010) reported that involvement of mandible is greater than maxilla (57.14% and 35.71%), respectively. The mandibular site most often affected is posterior region with incidence of 35.5% followed by maxillary posteriors (21%).[6] Majority of OM occurs in the second and third decade of life.[7] According to Harder (1978), the occurrence of OM is rarely seen before the age of 10 years.[1,8] OMs can cause marked asymmetry of face because of its site-specific aggressiveness of lesion although the pain is uncommon clinical findings. Its existence is unnoticed by patient as the pain and hypoesthesia are not common, so the lesions may reach a considerable size at the time of diagnosis. Cortical expansion and tooth displacement are encountered in relation to the larger lesions.[2,9,10] In infants, OM seems to display mostly the same clinical, radiological, and pathological characteristics.[11] Usually, OMs affect anterior parts of jaw in children with a unilocular appearance,[1,3] while multilocular lesions occur mainly in the posterior region.[7,12,13]

Case Report

A 1 year 8 months female child was referred to the Department of Oral and Maxillofacial Surgery, Faculty of Dental Sciences, Ramaiah University of Applied Sciences, for the evaluation and management of a swelling on the right side of the nose. History was elicited from the parent with respect to the child. It was a painless swelling which rapidly grew and was sudden in onset. The child had sustained blunt injury to right side of the face about 20 days back. Two days after the injury , she developed the swelling which was initially small but has increased to the

current size. On general physical examination, swelling was not associated with any systemic illness. History of present illness revealed a gradual increase in the swelling on the right side of nose in a span of approximately 20 days to the present size of 2 cm × 2.5 cm diameter from a small pea-sized growth. However, they noticed rapid increment in its size in the past 2 days. On extraoral examination, a non-tender, bony hard swelling at the right lateral nasal region was present, extending from inner canthus of eye superiorly to the middle third of the right lateral aspect of ala of nose inferiorly and anterioposteriorly from right infraorbital margin to the right lateral aspect of nose [Figure 1a]. The entire skin overlying the area appeared smooth and erythematous. On palpation, the swelling was firm in consistency and non-tender. No clinically, significant findings were present in intraoral examination. Computed tomography revealed a well-defined radiolucency in relation to right lateral aspect of the nose with the erosion of the nasal bone and obliteration of the nasal cavity [Figure 1b]. Under general anaesthesia, an incision was made in the inferior turbinate of the right nasal cavity. Greyish-white slimy tissue surfaced from the incision site which was completely evacuated by compressing the left lateral aspect of nose. Through irrigation was done and a nasal pack was given.. On gross examination, the excisional biopsy specimen was in multiple bits, largest bit measuring about 1 cm × 0.7 cm × 2.5 cm (l × b × h) in size, and they were gelatinous in nature color ranging from brownish-white to amber having soft consistency [Figure 1c]. The unique presentation of gross specimen directed toward the provisional diagnosis of OM.

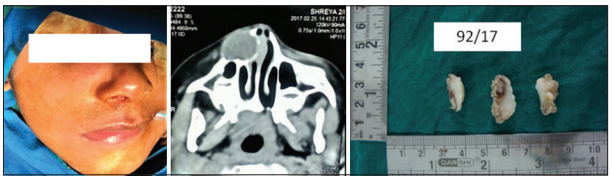

Figure 1: (a) Extraoral swelling extending superiorly from inner canthus of eye to the middle third of the right lateral aspect of ala of nose inferiorly and anterioposteriorly from right infraorbital margin to right lateral aspect of nose, (b) a well-defined radiolucency with erosion of the nasal bone and obliteration of the nasal cavity, (c) smooth, gelatinous, glistening, lobulated mass on gross specimen

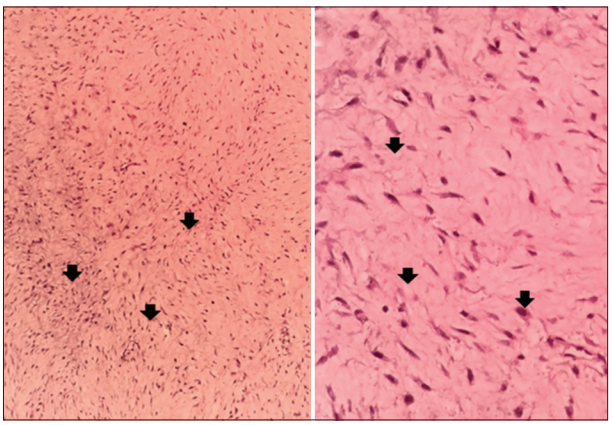

Figure 2: (a and b) H and E stained histopathology with abundance of stellate and spindle cells in a myxoid background (black arrows showing stellate and spindle cells ×A-4, ×B-40)

Histological examination

The H and E stained sections exhibited the abundant extracellular myxomatous stroma of ground substance and thin fibrils characterized by the presence of stellate and spindle cells in a predominantly myxoid connective tissue stroma along with few blood vessels suggesting of OM. [Figure 2]. Since it was a straightforward diagnosis of OM, immunohistochemistry (IHC) was not opted.

Treatment and prognosis

On post-histopathological diagnosis, conservative approach was the choice of treatment for the present case.

Discussion

The term myxoma was coined by Virchow (1863) for a group of tumors that histologically resembled the mucinous substance of the umbilical chord. In 1968, Stout stated that myxomas are true neoplasms that do not metastasize and the absence of cellular components that, especially, include lipoblasts, chondroblasts, and rhabdomyoblasts. Myxoma is the type of benign tumor that can occur at any site of the body including heart, skin, and subcutaneous tissue and centrally in the bone. In head and neck regions, myxomas are considered as rare tumors. Facial bone derived and soft tissue derived are the two forms of myxoma which can be identified. Further, these facial bone derived are subdivided into true osteogenic and OM, whereas soft tissue type myxoma are usually derived from the perioral, parotid gland, ear and larynx. In the International Histological Classification of Odontogenic Tumors (1971), OM is defined as a benign odontogenic tumor of mesenchymal origin that is locally invasive and consists of rounded and angular cells that lie in the abundant mucoid stroma.[14] Our case aligned with the above features suggestive of OM.

Radiologically, the lesion appears as a unilocular to a multilocular radiolucent lesion with well-defined or diffused margins. Bony trabeculation is present in its interior margin giving the lesion a "honeycombed," "soap bubble," or "tennis racket" appearance.[1,3,9] The present case was differentiated from other lesions that included odontogenic keratocyst, central giant cell granuloma, and neoplasm like ameloblastoma.

Odontogenic fibroma (OF) is the other major histological differential diagnosis for the current case which is also a benign odontogenic tumor histologically composed of stellate or spindle fibroblasts with hypercellular and myxomatous stroma. According to the WHO (1971), the presence of odontogenic epithelium is necessary for the diagnosis of OF. The choice between OF and OM for the current case is straightforward since, in OF, stroma will have higher cellular component than

collagenous component as well as the presence of odontogenic epithelium.[1] Chondromyxoid fibroma and chondrosarcoma can mimic OM as these lesions exhibit myxoid lesion with calcified products. Although both lesions can have a myxoid stroma, they also comprise areas of tumor with chondrocytes within lacunae of cartilaginous material.[1,15] For conventional OM, desmoplastic fibroma (DF) and neurofibroma (NF) also need to be considered in the differential diagnosis Histologically, DF is composed of a proliferation of spindle cells which are arranged in long and short fascicles in a fibromyxoid stroma, with hypercellular zone and dense interstitial collagen. NF exhibits a proliferation of spindle cells which are usually slender shaped with wavy nuclei in a myxoid stroma in contrast to plump spindle cells in OM.[15,16]

There are differences of opinion regarding the origin of myxomatous tumors. In past myxomatous, component of myxoma was compared with primitive mesenchyme that is present through the body. In past OM has also been compared with the dental papilla and the dental follicle. Ultrastructural and immunohistochemical studies discussed by Moshiri et al (1992) concluded that origin of myxomatous component of OM either could be fibroblastic or myofibroblastic or both. In their study, tumor cells showed positivity for vimentin and actin, respectively.[17] Hence, specific IHC markers can be employed for the diagnosis of OM in case of diagnostic difficulties.

Rare existence of tumor makes its outcome difficult leading in controversial treatment modality. Due its locally aggressive nature, tissue destruction and disfigurement are inevitable if left untreated. Some literature advocate for conservative approach like excision with narrow margins or curettage, whereas others for surgical therapy. 10 pediatric patients, in Kessler's (1995) series on follow-up recurrence, were evident in two cases; among them, one was treated with simple curettage within 1 year of surgery. In Slootweg *et al.* (2006), series 5 maxillary cases were treated following the conservative approach, and in follow-up of about 18.5 years, no evidence of recurrence was noted. Kleiber *et al.* (2014) presented a case of OM in a 3-year-old patient with an involvement of both mandible and maxilla. The patient underwent radical surgery and previous chemotherapy considering the aggressiveness of lesion. After complete resection, over 4 years of follow-up, the patient did not experience disease recurrence. According to Kawase-Koga *et al.* (2014) literature review of 44 cases, 45% underwent conservative surgery with 15% recurrence of the lesion, whereas no recurrence was observed in other group were radical surgery was performed after considering 46 months.of follow up. Among those cases, three cases were children that were 6, 7, and 12 years old, one underwent radical surgery, but none presented with recurrence. In review of 20 cases of infants by Kadlub *et al.* (2014), all cases presented in maxilla and one case presented with recurrence among 62.5% of cases which were treated conservative excision. In Rootenberg (2004) series of five cases pediatric patient treated with conservative excision, there was no evidence of recurrence over 8.5 years. Hence, according to Rootenberg data and others, in case of pediatric OM conservative approach can be taken considering the esthetics of the patient.[18-23] Hence, in the present case, treatment choice was conservative, concerning the age and esthetic of the patient with follow-up.

OM at pediatric age is a rare pathology. OM is a rare benign intraosseous neoplasm, termed "locally malignant" due to its exceptionally high local aggressiveness, high recurrence rate, and non-metastasizing nature. Unencapsulation and infiltrative growth pattern are responsible for a high rate of recurrence when conservative enucleation and curettage are performed.[1,24-26] Precise analysis of recurrence rates in pediatric cases is still missing due to poor follow-up and lack of reports in literature. Till date, there is no strong evidence in literature that highlights the best treatment approach to OM in pediatric case. Its radiologically varied appearance and histological mucoid stroma that can mimic few malignant lesions impose a diagnostic and operative dilemma; hence, warrants accurate clinical, radiological, and histopathological interpretation to arrive at a definitive diagnosis and effective management of patient considering the age of the patient.

Conclusion

This case report adds one more rare pediatric case presentation of OM to the present literature. Although aggressive in nature, conservative approach was the treatment of choice concerning the age, function, and esthetics of the patient. Appropriate diagnosis along with adequate treatment plan is mandatory for treating a pediatric case of OM.

References

1. Singaraju S, Wanjari SP, Parwani RN. Odontogenic myxoma of the maxilla: A report of a rare case and review of the literature. J Oral Maxillofac Pathol 2010;14:19-23.
2. Reddy SP, Naag A, Kashyap B. Odontogenic myxoma: Report of two cases. Natl J Maxillofac Surg 2010;1:183-6.
3. Manne RK, Sarath PV, Anumula L, Mundlapudi S, Tanikonda R. Odontogenic myxoma of the mandible. Case Rep Dent 2012;3:214704.
4. Limdiwala P, Shah J. Odontogenic myxoma of maxilla: A review discussion with two case reports. Contemp Clin Dent 2015;6:131-6.
5. Leiser Y, Abu-El-Naaj I, Peled M. Odontogenic myxoma-a case series and review of the surgical management. J Craniomaxillofac Surg 2009;37:206-9.
6. Neto VN, Sartori I, Oliveira L, Júnior JB, Souza D, Cardoso F, *et al.* Odontogenic myxoma in children: Case report. J Dent Res 2014;2:447-52.
7. Shah A, Lone P, Latoo S, Ahmed I, Malik A, Hassan S, *et al.* Odontogenic myxoma of the maxilla: A report of a rare case and review on histogenetic and diagnostic concepts. Natl J Maxillofac Surg 2011;2:189-95.
8. Harder F. Myxoma of the jaws. Int J Oral Surg 1978;7:148-55.
9. Li TJ, Sun LS, Luo HY. Odontogenic myxoma: A clinicopathologic study of 25 cases. Arch Pathol Lab Med 2006;130:1799-806.
10. Sivakumar G, Kavitha B, Saraswathi TR, Sivapathasundharam B.

Odontogenic myxoma of maxilla. Indian J Dent Res 2008;19:62-6.
11. Kadlub N, Mbou VB, Leboulanger N, Lepointe HD, Ansari E, L'hermine AC, *et al*. Infant odontogenic myxoma: A specific entity. J Craniomaxillofac Surg 2014;42:2082-6.
12. Spencer KR, Smith A. Odontogenic myxoma: Case report with reconstructive considerations. Aust Dent J 1998;43:209-12.
13. Chuchurru A, Uerti R, Cornicelli C, Dominguez FV. Myoma of the mandile ith unusual radiographic appearance. Oral Maillofac Surg 1985;43:987-90.
14. Farman AG, Nortje CJ, Grotepass FW, Farman FJ, Van Zyl JA. Myxofibroma of the jaws. Br J Oral Surg 1977;15:3-18.
15. Kramer IR, Pindborg JJ, Shear M. The world health organization histological typing of odontogenic tumours. Eur JCancer Care (Engl) Oral Oncol 1993;29:169-71.
16. de un Mixoma EC, con Desplazamiento O, Altug HA, Gulses A, Sencimen M. Clinico-radiographic examination of odontogenic myxoma with displacement of unerupted upper third molar: Review of the literature. Int J Morphol 2011;29:930-3.
17. Moshiri S, Oda D, Worthington P, Myall R. Odontogenic myxoma: Histochemical and ultrastructural study. J Oral Pathol Med 1992;21:401-3.
18. Takahashi H, Fujita S, Okabe H. Immunohistochemical investigation in odontogenic myxoma. J Oral Pathol Med 1991;20:114-9.
19. Gupta S, Grover N, Kadam A, Gupta S, Sah K, Sunitha JD. Odontogenic myxoma. Natl J Maxillofac Surg 2013;4:81-3.
20. Kiresur MA, Hemavathy S. An aggressive odontogenic myxoma of the maxilla. Indian J Dent Res 2014;5:214.
21. Rotenberg BW, Daniel SJ, Nish IA, Ngan BY, Forte V. Myxomatous lesions of the maxilla in children: A case series and review of management. Int J Pediatr otorhinolaryngol 2004;68:1251-6.
22. Keszler A, Dominguez FV, Giannunzio G. Myxoma in childhood: an analysis of 10 cases. J Oral Surg 1995;53:518-2.
23. Slootweg PJ, Wittkampf RM. Myxoma of the jaws: An analysis of 15 cases. J Maxillofac Surg 1986;14:46-52.
24. Abiose BO, Ajagbe HA, Thomas O. Fibromyxomas of the jawbones-a study of ten cases. Br J Oral Maxillofac Surg 1987;25:415-21.
25. Gnepp DR. Diagnostic Surgical Pathology of Head and Neck. Vol. 21. London: Saunders Company Ltd.; 2000. p. 643.
26. Lombardi T, Lock C, Samson J, Odell EW. S100, α smooth muscle actin and cytokeratin 19 immunohistochemistry in odontogenic and soft tissue myxomas. J Clin Pathol 1995;48:759-62.

How to cite this article: Singh P, Nambiar KS, Rao RS, Shridhar P, Prasad K, Sowmya SV, Augustine D, Haragannavar VC. A pediatric odontogenic myxoma of the maxilla: A case report and review. J Adv Clin Res Insights 2018; 5: 88-91.