Case Report
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Odontogenic Myxoma with Osteocementum-like Calcifications: A Rare Histological Variant

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Abstract

Odontogenic myxoma is a benign, locally aggressive neoplasm of mesenchymal origin. Histologically, the tumor exhibits spindle or stellate cells embedded in loose myxoid extracellular matrix and sometimes sparsely scattered residual bone and dystrophic mineralization in tumor stroma. Although, lesion sometimes presents with residual bony trabeculae but the formation of osteocementum-like calcifications is an extremely rare phenomenon. The present paper highlights two cases of odontogenic myxoma with osteocementum-like calcifications with emphasis on additional differential diagnosis that arose because of them and their effect on behavior of the lesion.

Keywords: Differential diagnosis, Odontogenic myxoma, Osteocementum-like calcifications, Residual bone

1. Introduction

Myxomas are rare benign, slow growing tumors of mesenchymal origin found in various sites of body including heart, skin, subcutaneous tissue, skeletal muscle, aponeuroses, genitourinary tract and bone. They are rare in head and neck region and mainly occur in the jawbones, with very small number of cases occurring in the larynx, pharynx, paranasal sinuses and other soft tissue[1-2]. Literature reported that Thoma and Goldman in 1947 first described the myxoma of odontogenic origin based on age, site of occurrence, association with missing teeth, histopathological resemblance to dental mesenchyme and sporadic presence of odontogenic islands[1]. OM can be divided into two groups that include (1) tumors that arise particularly in jawbones (most common type) and (2) tumors arising in soft tissues of that area[2].

OM is categorized by the World Health Organization as a mesenchymal odontogenic tumor that does not produce calcifications[3]. The aggressive nature and high recurrence rate of OM is well documented in the literature and could be attributed to the local infiltration by the tumor. Histologically, it sometimes reveals scattered residual bony trabeculae, which could be the result of local infiltration by the tumor but the presence of osteocementum-like calcifications is a rare phenomenon[4-5]. On extensive literature search, few cases of OM with osteocementum-like calcifications were found. Keeping in mind the plausible literature on OM with osteocementum-like

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calcifications, two such cases are being highlighted with emphasis on additional differential diagnosis that arose due to their presence and the corresponding effect on the behavior of the tumor.

2. Case Report

2.1 Case 1

A 45-year old male reported to our institute with a complaint of swelling in the left posterior region of maxilla since 8-9 months and inability to breathe from left side of nose for past 3-4 months. The swelling gradually increased in size and attained the present size. Patient gave history of surgery for the myxoma of nasal cavity 1-2 years back. Intraorally, swelling extended anteroposteriorly from maxillary left second premolar to maxillary tuberosity, laterally occluding the left vestibule and medially 1cm away from the midline. The overlying mucosa appeared to be normal. On palpation, swelling was hard, non-compressible and non-tender. Computed Tomography (CT) scan showed a well-defined radiolucency on left side of maxilla causing erosion of the maxillary sinus walls and displacement of the nasal wall towards opposite side. Based on clinico-radiological evaluation a provisional diagnosis of myxoma or salivary gland tumor was made. Incisional biopsy of the lesion was done and on histological examination, a diagnosis of myxofibroma was given. Patient underwent surgical excision and tissue was sent for histopathological examination to our department. Macroscopically, tissue was firm to gritty in consistency and gelatinous in appearance (Figure 1a**).** Microscopically, multiple sections obtained from hard and soft tissue revealed extensive areas of myxomatous tissue admixed with fibrous tissue. Myxomatous areas showed stellate and angular shaped cells (Figure 1b). Few sections revealed presence of residual lamellar bone (Figure 1c) and areas of metaplastic bone formation (osteocementum-like calcifications) (Figure 1d). Hence,

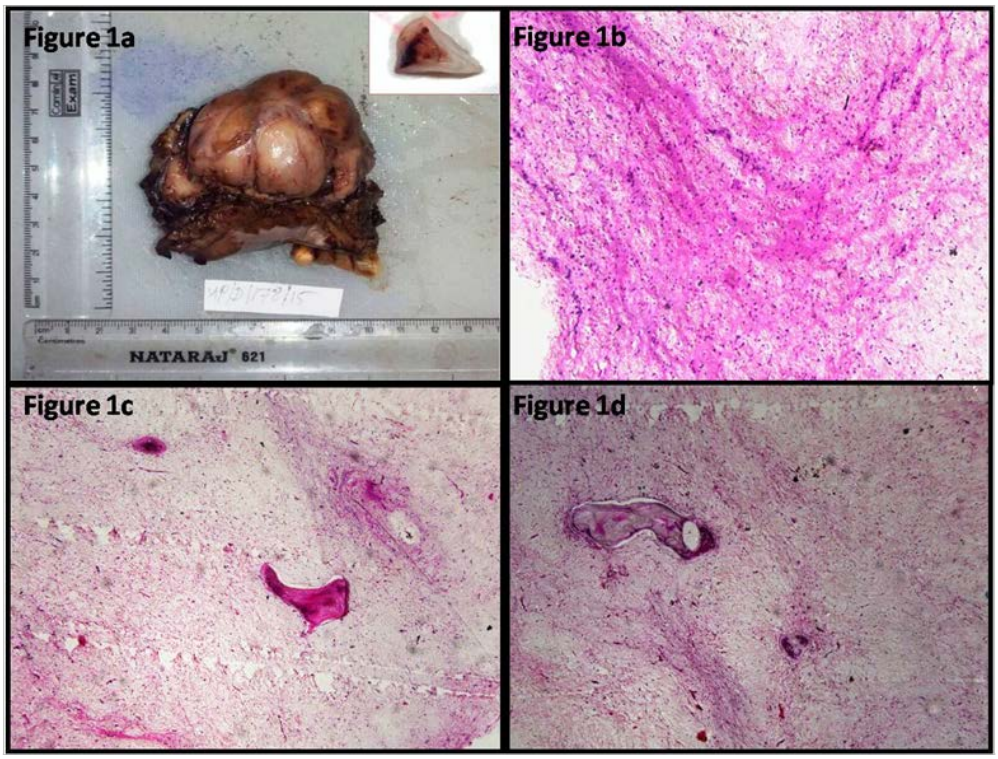


Figure **1a.** Gross Specimen of Case 1: soft tissue was firm to gritty in consistency and gelatinous in appearance; **b.** Photomicrograph reveals myxomatous areas admixed with fibrous tissue. Myxomatous areas show stellate and angular shaped cells (H&E 10X; Case 1); **c.** Photomicrograph reveals myxomatous area with residual bone (H&E 4X; Case 1); **d.** Photomicrograph reveals myxomatous area with areas of metaplastic bone formation (osteocementum–like calcifications) (H&E 4X; Case 1).

a final diagnosis of odontogenic myxofibroma with osteocementum-like calcifications was made.

2.2 Case 2

A 16-year-old female reported to our institute with complaint of swelling on right side of mandible in the tooth-bearing area since past 2 months. Swelling mildly increased in size and was not associated with pain. Intraorally, swelling was present on right mandibular body region extending from 43 to 47. On palpation, swelling was firm in consistency, sessile, diffuse, non-tender and lead to expansion of both buccal and lingual cortical plate. Orthopantomogram (OPG) showed mixed radiolucent and radiopaque lesion with respect to right side of mandible extending from distal of 42 to mesial of 47. Tooth displacement with respect to 43 to 46 was also observed. Based on clinico-radiological evaluation, a provisional diagnosis of ameloblastoma or central giant cell granuloma was given. Incisional biopsy of the lesion was done and tissue was sent for histopathological examination. The tissue sections revealed abundant haphazardly arranged stellate, spindle or round cells in myxoid ground substance. Few areas with loose myxoid stroma also revealed abundant collagen bundles. Some areas showed osteocementum-like calcifications with minimal inflammation. Clinico-radiological correlation confirmed the diagnosis of odontogenic myxoma with osteocementum-like calcifications.

3. Discussion

OM is a rare benign neoplasm that constitutes about 3% of all odontogenic tumors which is believed to arise from the mesenchymal tissue of the tooth germ, either the dental papilla, follicle or the periodontal ligament[4,6]. It occurs most commonly in the second or third decade of life and rarely occurs before the age of 10 or after 50 years of age[6]. Both of the present cases were between 10-50 years of age. No gender predilection is evident for this tumor but it shows slight predilection for mandible. It is a slow growing lesion thus may or may not be associated with pain. As it mainly occurs as the central lesion of jaws, it is associated with bone expansion and might cause destruction of the cortex[6].

Radiographically, it may present with uni- or multi-locular radiolucency but some authors reported that lesion could be mixed radiolucent and radiopaque. A "tennis racket" appearance and diffuse calcifications have also been reported in literature. Displacement of tooth by the tumor is a common radiographic finding although root resorption is rare[3-6].

Microscopically, tumor consists of haphazardly arranged spindle, stellate or round cells with fine, long, eosinophilic cytoplasmic processes extending from centrally placed nucleolus within an abundant loose myxoid or mucoid stroma that contains few fine collagen fibrils[3,5]. The loose myxoid tissue is not highly cellular and the cell does not show pleomorphism, prominent nucleoli or mitotic figures. The tumor exhibits variable number of tiny capillaries and occasionally nests of odontogenic epithelium[6]. Isolated foci of residual bone and dystrophic mineralization were also evident in the tumor[3]. Histologically, both of our cases showed classical features of OM along with presence of calcifications. Some of them exhibited characteristic lamellar pattern but others were hematoxyphilic, hypocellular to acellular, present in the form of small spherules or large masses throughout the lesion, with no distinct cellular rimming around them and were non-polarizable under polarized light (Figure 2a), thus, they were best described as osteocementum-like in nature[3-4,7]. Osteocementum-like calcifications are believed to arise by metaplasia as typical rimming by plump osteoblasts or cementoblasts is absent[8]. In contrast, the residual bone is normal bone which is trabecular in shape, exhibits lamellar pattern, present focally or at the tumor periphery or between the lobules of tumor and shows birefringence under the polarized light[3-4,8-9]. (Figure 2b) In addition, literature states the presence of peripheral radiating collagen fibers from the calcified spherules there by recapitulating Sharpey's fibers, thus, confirming the presence of osteocementum-like calcifications in OM[3]. Till date, only 25 cases of OM with osteocementum-like calcifications have been reported in literature worldwide (Table 1)[3-5,7-13].

Histopathologically, odontogenic mesenchymal tumors and non-odontogenic tumors with myxomatous degeneration should be considered in the differential diagnosis of odontogenic myxoma[7]. The unusual presence of osteocementum-like calcifications gave rise to an additional differential diagnosis of central odontogenic fibroma, fibro-osseous lesions (ossifying fibroma/cemento-ossifying fibroma, fibrous dysplasia), chondromyxoid fibroma and chondrosarcoma[3-4, 7-10]. Central odontogenic fibroma (COF) should be considered in differential diagnosis as WHO type of COF may present with foci of

Table 1. Reported cases of odontogenic myxoma with osteocementum-like calcifications

Author	No. of cases reported	Age (yrs)	Gender	Site	Radiographic Findings	Type of Calcification
Rennie *et al.* 1985	1	5	Male	Right Mandible	Well-defined intra-bony radiolucency with discrete radioopaque mass with several smaller peripheral masses at anterior-superior edge of radiolucency. Tooth germ of the second permanent molar displaced upwards and posteriorly.	Cementum-like calcified material
Ogyur *et al.* 2001	1	19	Female	Left Maxilla	Irregularly outlined radiolucency associated with an unerupted tooth, traversed by fine crossing trabeculae.	Osteocementum-like spherular calcified bodies
Lin and Basile 2010	1	25	Female	Right Mandible	Unilocular radiolucency causing tooth displacement.	Osteocementum calcified products
Effiom *et al.* 2011	16	-	-	-	-	16 out of 63 cases showed cementum-like calcified material
Sriram *et al.* 2012	1	33	Female	Left Mandible	Multilocular radiolucency with fine internal trabeculations.	Acellular, globular calcified products arisen by metaplasia
Lahey *et al.* 2013	1	69	Female	Right Mandible	Mixed radiolucent/radiopaque lesion with poorly defined borders.	Osteocementum-like calcifications
Vecchio *et al.* 2014	1	50	Male	Left Maxilla	Ill-defined radiolucent lesion associated with unerupted tooth.	Osteocementum-like spheroid bodies
Rallis *et al.* 2015	1	32	Male	Left Maxilla	CT scan revealed a left maxillary lesion. Diffuse ossifications were present and an impacted molar was repulsed to the anterior wall of the left sphenoid sinus.	Osteocementum calcified spherules
Hammad *et al.* 2016	1	45	Female	Left Mandible	Well-defined, multilocular radiolucency with root resorption of first and second molars	Cementum-like material
Poudel *et al.* 2016	1	53	Female	Right Mandible	Multilocular radiolucency	Cementum- like calcifications
Our Series	2 cases					
Our Case 1		45	Male	Left Maxilla	Well-defined radiolucency causing erosion of the maxillary sinus walls and displacement of the nasal wall towards opposite side	Osteocementum – like calcifications
Our Case 2		16	Female	Right Mandible	Mixed radiolucent and radiopaque lesion with displacement of tooth	Osteocementum – like calcifications

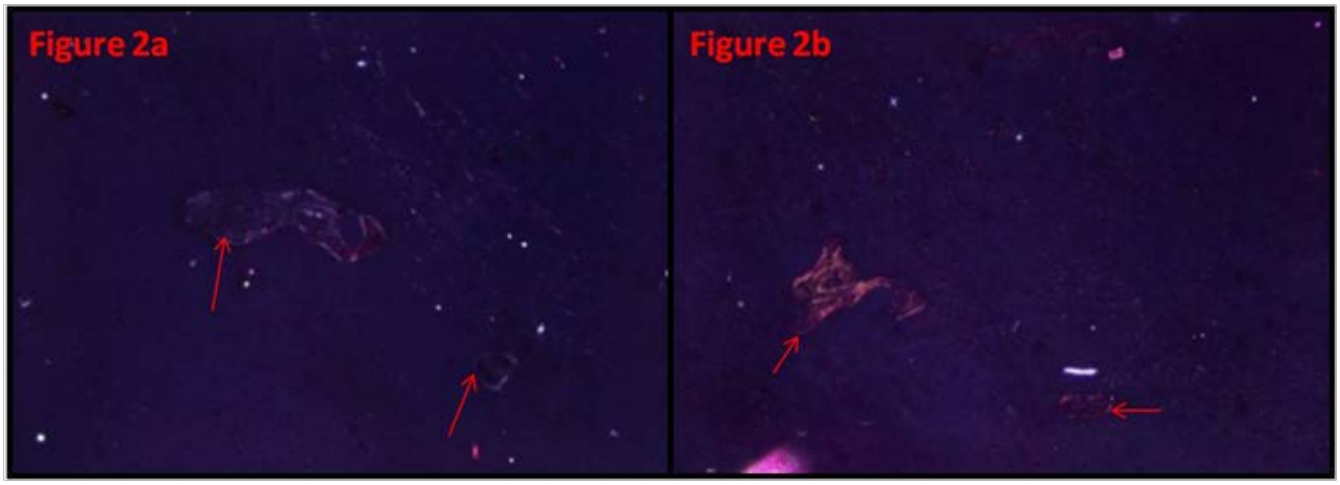


Figure 2a. A photomicrograph reveals presence of osteocementum-like calcifications present in large masses or small spherules, most of them were hypocellular to acellular and are non-polarizable under polarized light (H& E 10X, Polarized microscope); **b.** A photomicrograph reveals presence of residual bone with lamellar pattern and birefringence under polarized light. (H& E 10X, Polarized microscope).

calcified material, which resembles dysplastic cementum, a tubular dysplastic dentin or osteoid. Simple type of COF exhibits a cellular areas with delicate fibers scattered in considerable amount of ground substance yielding fibromyxoid quality. COF can be differentiated from the OM, as COF is more cellular and fibrous than myxoma, does not show clear-cut myxoma-like areas and has a well-demarcated margin in contrast to infiltrative margins of myxoma[7-8]. OM with calcifications could resemble ossifying fibroma/cemento-ossifying fibroma as they exhibit formation of osteoid or cementum in various proportions. OM with calcifications could be demarcated from the ossifying fibroma/cemento-ossifying fibroma as they are more densely collagenized, encapsulated, does not exhibit islands of odontogenic epithelium and are characterized by absence of predominant myxoid area[3,9]. Fibrous dysplasia may show myxoid change therefore, it may be considered in differential diagnosis of OM with calcifications. However, fibrous dysplasia shows fibrillary connective tissue with variably shaped trabeculae of woven bone, which typically lacks osteoblastic rimming[6,9]. Chondromyxoid fibroma and chondrosarcoma show areas of tumor with chondrocytes residing in lacunae of cartilaginous material that helps in differentiating them from OM with calcifications[4].

Small OMs are normally treated by curettage; however, larger lesions necessitate extensive resection. Ability of OMs to recur locally make follow-up of the affected persons necessary after surgical treatment[12]. Effiom *et al.*, reported that even though the exact role of osteocementum-like calcifications could not be established but their presence did not appear to alter the clinical behavior of the tumor[11].

4. Conclusion

The present article contributes to broaden the morphological spectrum of the OM. The presence of osteocementum-like calcifications does not itself mitigate the diagnosis and alter the clinical behavior of the tumor but their presence raises other differential diagnosis for the tumor. Therefore, it should be kept in mind that histopathological findings should not lead to misdiagnosis of the lesion.

5. Acknowledgement

Nil

6. Conflict of Interest

Nil

7. References

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