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Case Report

Sarcomatous transformation of florid cemento-osseous dysplasia: a case report

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ABSTRACT

Florid cemento-osseous dysplasia (COD) represents a rare group of benign fibro-osseous disorders, while osteogenic sarcoma (OS) on the other hand, is a malignant tumour of ominous prognosis. The malignant transformation of a benign fibro-osseous lesion of the jaw is quite uncommon and has few reported cases. The clinico-radiological findings and histopathological analysis of a lesion present in all four quadrants of the jaw of a patient who reported at the outpatient department of a regional dental college is presented here. The lesion underwent sarcomatous transformation over two months and the findings were confirmed by histopathological evaluation. Although florid cemento-osseous dysplasia is a benign lesion of the jaw with a very low propensity for malignant transformation, it should be closely monitored in patients with tumour predisposition syndromes. Further research and molecular studies are required for better understanding of inadvertent changes.

Keywords: Osteosarcoma of the mandible, Florid cemento-osseous dysplasia, Malignant transformation

INTRODUCTION

Fibro-osseous lesions of the jaws comprise cemento-osseous dysplasia (COD), ossifying fibroma (OF) and craniofacial fibrous dysplasia (FD) which are histopathologically similar but have distinct clinic-radiological features. Cemento-osseous dysplasia is quite common and has predilection for middle-aged females of African lineage. The three subtypes of fibro-osseous lesions can be differentiated from each other depending upon anatomic consideration: periapical cemento-osseous dysplasia affects the periapical region of the anterior mandibular region; florid cemento-osseous dysplasia usually affects multiple quadrants. Radiographically these lesions have an initial radiolucent appearance and undergo subsequent mineralization in a centripetal manner leaving a sharply delineated, sclerotic zone after full mineralization.[1-3] Routinely cemento-osseous dysplasia is expansile, asymptomatic and does not require specific management. Radio-graphic features are diagnostic and pathognomonic for this particular entity. Some authors even consider biopsy to be contraindicated since it can result in persistent local infection and a complicated clinical course. Histologically, all subtypes show a monomorphic spindle cell stroma containing an immature matrix formation that consists of woven bone and hypocellular cementum-like material that progressively coalesces to form larger, radio-opaque masses.[4,5]

Whereas FD is known to be caused by a postzygotic activating mutation in the GNAS gene and to rarely undergo malignant transformation, the etiopathogenesis of cemento-osseous dysplasia and ossifying fibroma remains unknown and are not considered precursors of osteosarcoma or other malignant tumours of bone.

CASE REPORT

A 56-year-old woman complained of a swelling in left posterior region of lower jaw which had been present for 3 months (Figure 1). Swelling was insidious in onset gradually it increased in size. Mild intermittent throbbing pain was present in that region. Patient had no history of uncontrolled systemic disorder and deleterious oral habits. On extraoral examination diffuse swelling measuring 4cm in maximum dimension was present extending from mid-pupillary line to the posterior border of the ramus of mandible. The surface overlying the swelling was not shiny, non-erythematous and there was no local elevation of temperature. On palpation swelling was tender and bony hard in consistency.

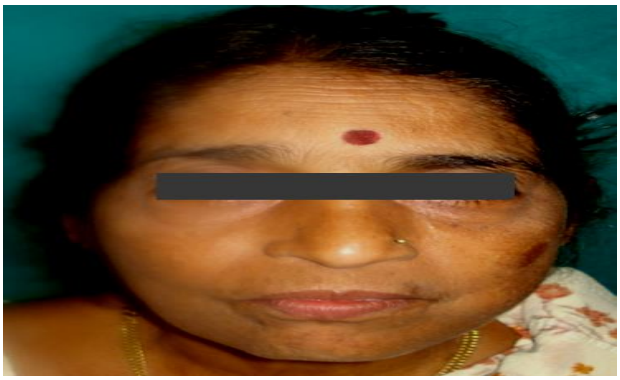

Figure 1: Extraoral swelling in right side of mandible.

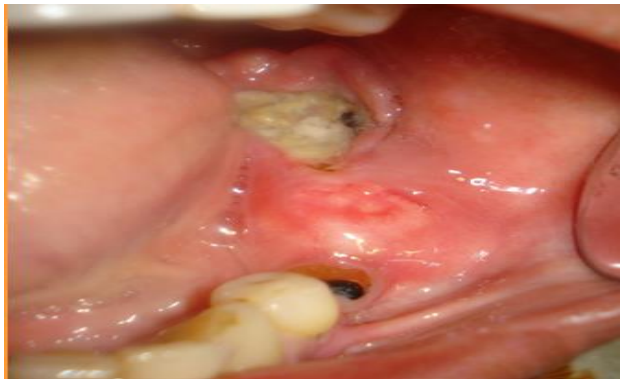

Figure 2: Intraoral appearance of the lesion.

On intraoral examination, there was a 3×3 cm swelling present in left molar region with obliteration of buccal vestibule. Oral mucosa was erythematous in that region with presence of necrotic bare bone. Gingiva around the exposed bone was rolled out, non-indurated. Retained root was present in the adjoining premolar region and there was impingement over the swelling from the upper removable partial denture of the patient. Swelling was tender on palpation and there was absence of perforation in buccal and lingual cortex. Mild bleeding was found during palpation in that region.

Panoramic radiograph showed an irregular cotton wool like radio-opacities in left posterior region of mandible along with diffuse ill-defined radio-opaque flecks in the other three quadrants (Figure 3). Lower occlusal view revealed the presence of ill-defined homogenous radio-opacity in the left molar region with expansion of the buccal and lingual cortices (Figure 4). Cortico-cancellous incisional biopsy specimen was taken from the most representative region and histopathologically it was found out to be florid cemento-osseous dysplasia with no characteristic signs of malignancy. Patient was asked to report at 3 month's interval for periodic follow-up.

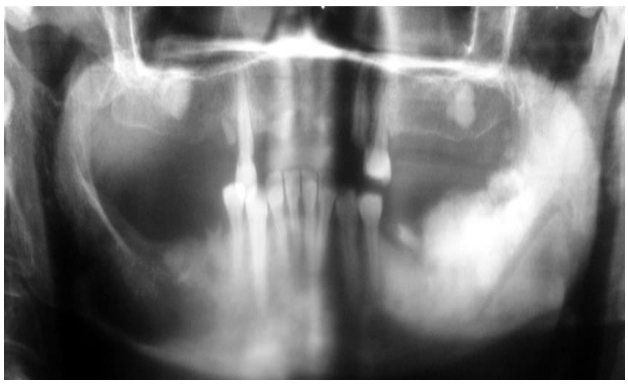

Figure 3: Panoramic radiograph depicting radiopaque masses in all the four quadrants of jaw with a large radiopaque mass in the left mandibular region extending from the first molar region and involving the left mandibular ramus.

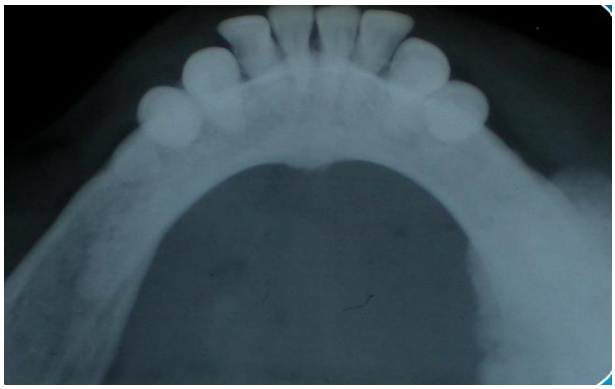

Figure 4: Occlusal radiograph revealing expansion of the buccal and lingual cortical plate on the left side along with presence of homogenous radiodensity.

Patient reported two months later with increased swelling in the region, restriction of mouth opening and elevated pain level in that region (Figure 5). Also the regional lymph nodes were palpable, tender without any fixity to overlying structures as well as overlying skin. Panoramic radiograph revealed increased size of radio-opacity in the left mandibular region without any changes in the other three quadrants (Figure 6). Computed tomography (CT) scans confirmed the known densely mineralized lesions in all four quadrants with the left posterior mandible appearing more aggressively affected and showing new and focal cortical penetration on the buccal aspect (Figures

7 and 8). An incisional biopsy was performed and a specimen was taken from the most representative region and sent for histopathological region. It was diagnosed as low-grade desmoid like intramedullary osteosarcoma of the left mandible (Figures 9).

Figure 5: Patient reported 2 months later with increased swelling, tenderness and palpable, tender and mobile submandibular and submental lymph nodes.

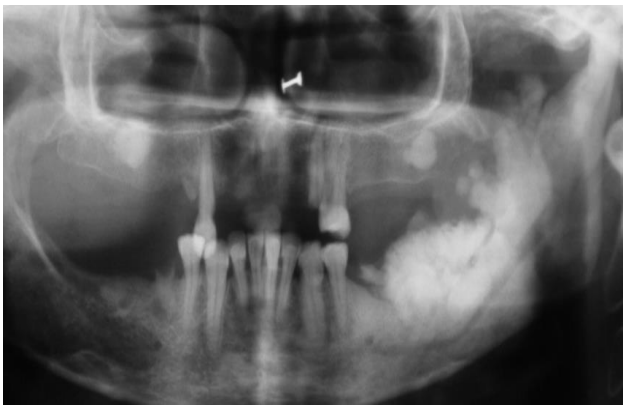

Figure 6: Panoramic radiograph revealed increase in size of the radio-opacity with extension upto the left sigmoid notch.

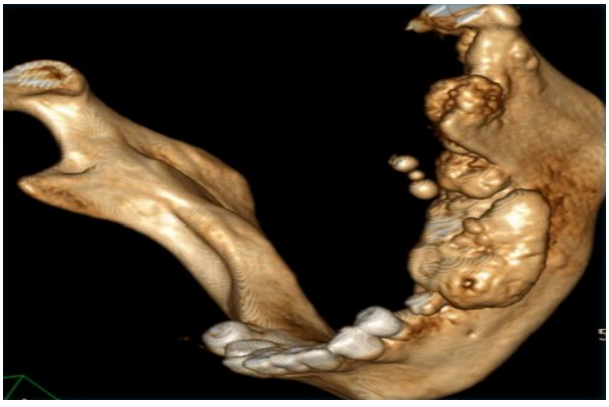

Figure 7: CT scan revealing proliferative changes as well as perfortation of the buccal cortex.

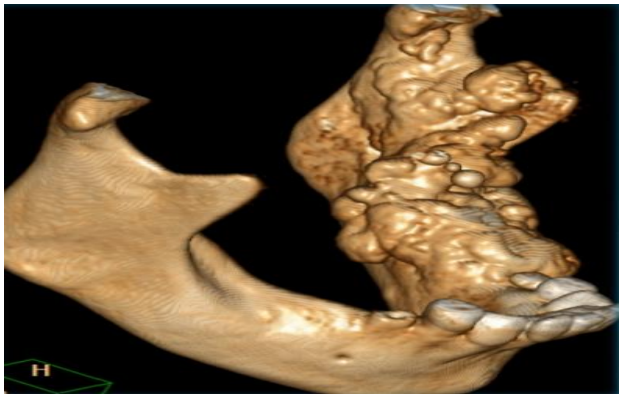

Figure 8: Perforation of the lingual cortex as depicted by 3D-CT.

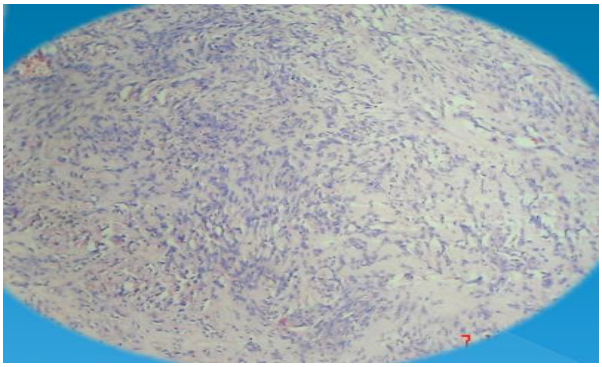

Figure 9: Herring bone pattern typical of osteosarcoma.

DISCUSSION

Cemento-osseous dysplasia is a relatively uncommon non-neoplastic fibro-osseous lesion with any significant propensity for malignant transformation. It is usually an incidental finding on radiographs. COD has been associated with some lesions such as solitary bone cyst, aneurysmal bone cyst and chronic osteomyelitis. The periapical subtype presents as discrete masses of varying radiopacities at the apices of teeth, especially the mandibular anterior teeth which typically remain vital. The focal subtype is typically a solitary, often lobulated lesion presenting bilaterally, while the florid subtype presents as irregular, multiple masses of mixed radiopacities occurring in more than two quadrants of the jaw. The management of COD is conservative unless when symptomatic as a result of superimposed infection as COD is avascular and therefore susceptible to infection.[9,10] Osteogenic sarcoma (OS) is a highly malignant tumour of bone that is associated with a poor prognosis. Its etiopathogenesis id unknown but a few cases have been known to occur in irradiated or previously diseased bone.[10,11] It usually affects the metaphyses of long bones in children and adolescents but the jaws are the fourth most common site involved. Patients with osteosarcoma in the jawbones are usually one to two decades older than patients with peripheral osteosarcoma and have a more favorable outcome because metastatic spread occurs less

often and later in the course of the disease.[6-8] Osteosarcoma of the jaws is uncommon and accounts for about 5% to 10% of all osteosarcomas.[11] This tumour is typically seen in the third and fourth decades of life, has a male predilection and classically presents with a rapidly growing swelling which is usually found in the mandible and painful.[10,11] Schneider et al in 1999, reported a case of a 54-year-old black woman seen in 1979 with "malignant spindle cell tumour" of mandible who was previously diagnosed with florid osseous dysplasia.[12]

The tumour was diagnosed histologically as malignant fibrous histiocytoma, but immunohistochemistry was not available at the time to confirm this diagnosis. In their report, a transition zone between these two lesions was cited though they could not rule out the possibility of a collision phenomenon. Theirs was the first reported case of mandible occurring with florid osseous dysplasia. Their case represents the first reported malignancy occurring with a cement-osseous dysplasia. Melrose and Handlers in 2003 reported a case of a 36-year-old black woman with an increasing, painful, mandibular swelling who had radiographic evidence of florid cemento-osseous dysplasia in the maxilla as well as mandible three years before.[13] The biopsy of the mandibular swelling was stated as high-grade osteosarcoma, while a core biopsy of the lesion on the contralateral side of the mandible revealed features in accordance with cemento-osseous dysplasia. Lopes et al, in 2010, reported a similar case to that of Melrose and Handlers in a clinic-pathologic conference as a 44- year-old black female who presented with a painful swelling of the right mandible which was diagnosed as osteosarcoma in a background of FCOD.[14] Cheng et al in 2002, reported a case of a 72-year-old black female with a week history of left mandibular swelling and numbness of the lower lip.[6] The patient was previously diagnosed with Paget's disease as well as cemento-osseous dysplasia which subsequently underwent sarcomatous transformation over a course of 3-years albeit in a different site of the mandible. A majority of the cases have been associated with florid subtype of cemento-osseous dysplasia which is said to be similar to Paget's disease of bone.

However, FCOD is more localized and predominantly affects the jaws. Based on this comparability, FCOD has been referred to as the "Paget 's disease of the mandible".[14] Sarcomatous transformation of florid cemento-osseous dysplasia is an uncommon entity with few reported cases. Management options include radical surgery combined with adjuvant radiotherapy and chemotherapy. Chondroblastic low-grade tumours have a better prognosis as compared to the high-grade tumours. Although COD generally does not require close monitoring, a closer surveillance should be considered in patients with concomitant tumor predisposition syndromes.

CONCLUSION

Although malignant transformation of florid cemento-osseous dysplasia is rare, there are a few cases which have been reported previously. Further investigations are required at the molecular level to better understand the etiopathogenesis of such diseases.

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