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Case Report

Dentistry Section

Osteosarcoma of Mandible: A Case Report and Review of Literature

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ABSTRACT

Osteosarcoma is a bone tumour and it can occur in any bone, usually in the extremities of the long bones, near the metaphyseal growth plates. Osteosarcomas of the jaws are rare and they are highly malignant neoplasms of the bone whose diagnoses are challenging. In the initial phase, they may present as non-descript, bony swellings with an indolent growth, only to become malignant towards the later stages. Osteosarcomas of the jaw are rare and they differ from osteosarcomas of the long bones in their biological behaviour, even though they have the same histological appearance. The purpose of this study was to report a mandibular osteosarcoma in a 50-year old, which presented as a bony mass which mimicked a periapical infection.

Key Words: Osteosarcoma, Sunray appearance, Codman's triangle, Radical surgery

INTRODUCTION

Osteosarcoma of the jaws (JOS) is a rare, aggressive, malignant mesenchymal tumour which is characterized by the formation of osteoid tissue, which constitutes 5% to13% of all the cases of skeletal osteosarcomas [1,2,3,4]. The estimated incidence of the new cases of JOS per year is 0.07 in 100,000 [1].

Jaw osteosarcomas usually present in the third and fourth decades of life, almost a decade after their presentation in the long bone tumours [1,2,3]. The maxilla and the mandible are equally involved. The mandibular tumours arise more frequently in the posterior body and in the horizontal ramus, whereas the maxillary tumours are discovered more commonly in the alveolar ridge, the sinus floor, and the palate [1,2,3,4,5].

The main clinical manifestations of JOS are pain of variable intensity, swelling of the bone and the adjacent soft tissues, tooth bulging and dislocation, lack of healing and swelling at the site of the tooth extraction, trismus and hypoesthaesia or paresthaesia in the case of the mandibular tumours, and nasal obstruction in the maxillary tumours [1,2,3].

The radiological appearances manifest as mixed, radiolucent/radiopaque lesions, periodontal ligament widening, radiopaque masses with a moth eaten appearance, Codman's triangle and sunburst appearance [3,5].

The aetiology of osteosarcoma remains unknown. However, ionizing radiations (irradiation sarcoma), fibrous dysplasia and Paget's bone disease (Paget's sarcoma) of the bone are considered to be the predisposing factors. Patients with chromosomal aberrations in the p53 and the retinoblastoma genes which are localized to 17p13 and 13q14 and patients with the Li-Fraumeni syndrome or hereditary retinoblastoma have an increased risk for developing osteosarcoma. The current knowledge on JOS has indicated that certain other factors which appeared to co-relate with the occurrence of osteosarcoma included linear bone growth and genetic and environmental factors [4,5,6].

The differential diagnosis of JOS should include chondrosarcoma, Ewing's sarcoma, bone metastasis , fibrous dysplasia ,osteomyelitis, and even lesions that do not usually affect the jaw bones, such as fibrosarcomas, leiomyosarcomas, or rhabdomyosarcomas [7].

The WHO has listed several variants that differed in the location, clinical behaviour and the level of cellular atypia. Classical osteosarcoma is the most frequent variant which develops in the medullary region of the bone and it can be subdivided into the osteoblastic, chondroblastic and the fibroblastic histologic types, depending upon the type of the extracellular matrix which is produced by the tumour cells. The other histological variants include the telangiectatic type, small cell osteosarcoma, the giant cell and the large cell predominant type, etc. [1, 2, 8]

JOS differs from osteosarcoma of the long bones in its biological behaviour, thus presenting a lower incidence of metastasis and a better prognosis [3,4]. Early diagnosis and adequate surgical resection are the keys to high survival rates [6]. The treatment protocols for osteosarcoma include radical or conservative surgery which is complemented by radiotherapy and/or chemotherapy [2,8].

Dental professionals may be the first to detect jaw osteosarcomas in their initial stages. Regardless of its favourable biological behaviour, the patients of jaw osteosarcoma usually exhibit advanced tumour, as it often goes unnoticed by the dental professional, thus stressing on the need for an early diagnosis of the lesion. The aim of this case report was to draw attention to the possibility of diagnosing this tumour based on its clinical and radiographical characteristics before its confirmation by histology.

CASE REPORT

A 50-year old male patient reported to the Department of Oral Medicine and Radiology, Ragas Dental College and Hospital, Chennai, India, with a chief complaint of swelling in his lower right jaw for 15 days. The pain was severe, pricking, and continuous in nature and it did not subside with medication. The pain was

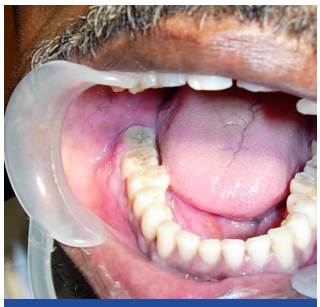
associated with a swelling which gradually increased in size in 15 days. According to the patient, the swelling was evident extra-orally since 10 days. The patient also expressed a difficulty in chewing food on the affected side. His past dental history revealed previous restoration of the 48 tooth after the occurrence of dental caries [Table/Fig-1].

His extra-oral examination revealed a 2 x 2 cm in size, ill defined swelling which was seen at the lower 1/3rd of the right side of the face, which extended superoinferiorly, 3 cm from the right zygoma to 1 cm above the inferior mandibular border and antero posteriorly, 2 cm from the angle of the mouth to 3 cm from the angle of the mandible [Table/Fig-2]. On palpation, the swelling was found to have a smooth surface and it was warm, tender, bony hard and not movable in both the anteroposterior and the lateral directions. The right sub-mandibular lymph nodes were palpable, soft to firm, mobile and tender. Intra orally, a 2 x 2 cm round, well defined swelling which extended from the free gingival groove to the vestibule, which obliterated the buccal vestibule, was seen in relation to the 47, 46, 45, 44 region. Also, a 0.5 to 0.5 small bony elevation with normal overlying mucosa and indistinct margins was seen on the lingual alveolar mucosa in relation to the 47, 46, 45 region. The swelling had distinct margins, a smooth surface and the normal colour of the overlying mucosa. On palpation, buccal and lingual cortical plate expansion was evident and the swelling was found to be tender, bony hard and not movable in the anteroposterior and the lateral directions. The tooth numbers, 47, 46, 45, 44 were tender on percussion [Table/Fig-3].

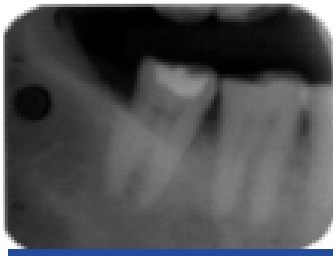
The radiographic evaluation included a intra-oral periapical radiograph (IOPA), a panoramic radiograph and a mandibular cross-sectional radiograph. The IOPA revealed widening of the periodontal ligament space with an irregular absence or attenuation of the lamina dura in relation to 47, 48 [Table/Fig-4]. The panoramic radiograph revealed an ill defined, mixed, radiolucent-radiopaque lesion along the right body of the mandible, denoting irregular areas of osteolysis [Table/Fig-5]. The cross-sectional occlusal radiograph of the right side of the mandible showed bicortical expansion and the presence of radial spicules which spread outside the jaw bone on the lingual side, giving a “sunray appearance” in relation to the 48, 47 teeth region [Table/Fig-6]. The non-contrast, multislice spiral CT scan of the mandible and the face revealed an expansile mass lesion which involved the right side of the mandible [Table/Fig-7]. Based on the clinical and radiological findings, a provisional diagnosis of malignancy of the right body of the mandible was given.

The differential diagnosis included cellulitis which involved the right buccal, vestibular and the submandibular space and a vascular lesion (hemangioma). An incisional biopsy revealed hyperchromatic, pleomorphic cells which were associated with extravasated RBCs. The decalcified tissue section showed vital bone with marrow spaces which exhibited proliferation of the angular cells, some of which were in the lacunar spaces, some in association with amorphous eosinophilic material which was suggestive of an osteoid [Table/Fig-8]. These histological features were suggestive of osteosarcoma of the right mandible. The patient was referred to an oncology centre, and the treatment regimen which was prescribed was radical surgical resection along with a margin of the normal surrounding tissue, followed by radiotherapy.

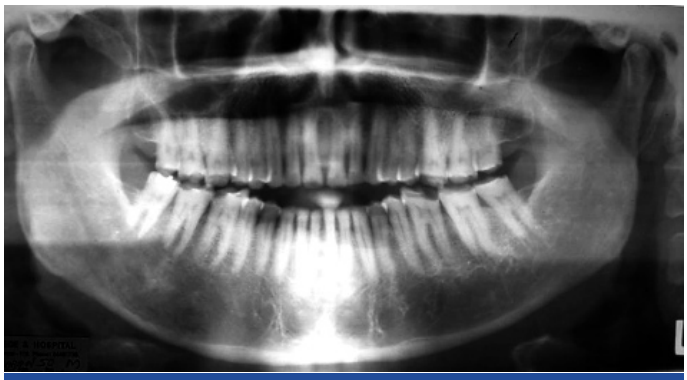
[Table/Fig-1]: Extent of the swelling



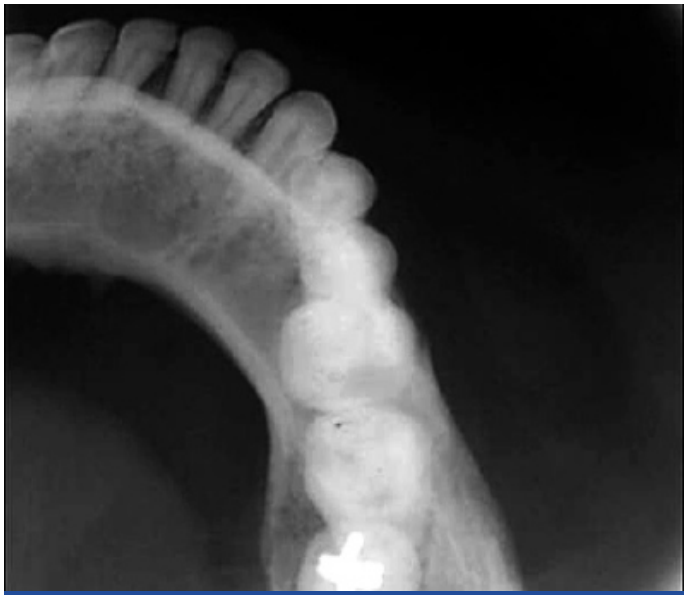
[Table/Fig-2]: Intraoral-Bicortical expansion in relation to 47, 48



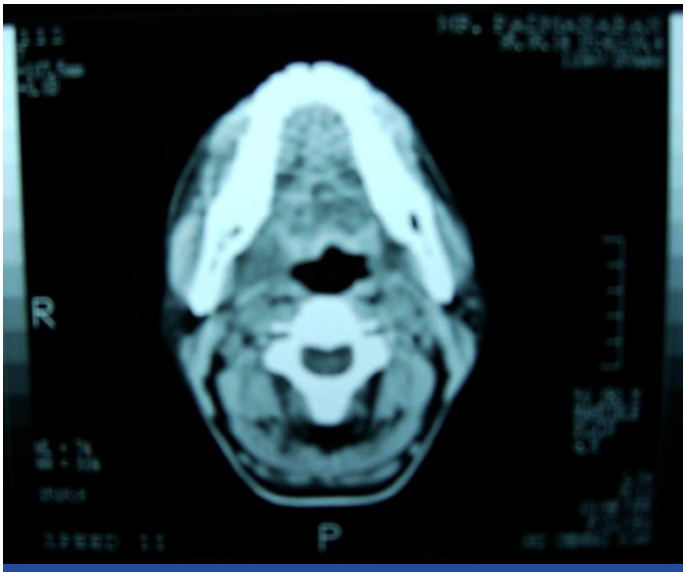
[Table/Fig-3]: Intraoral periapical radiograph showing change in trabacu-lation in relation to 48



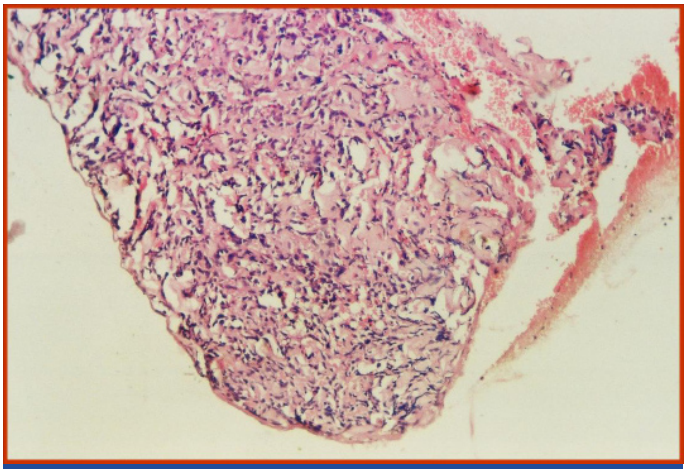
[Table/Fig-4]: OPG showing areas of Radiopacity in the apical region of 47, 48



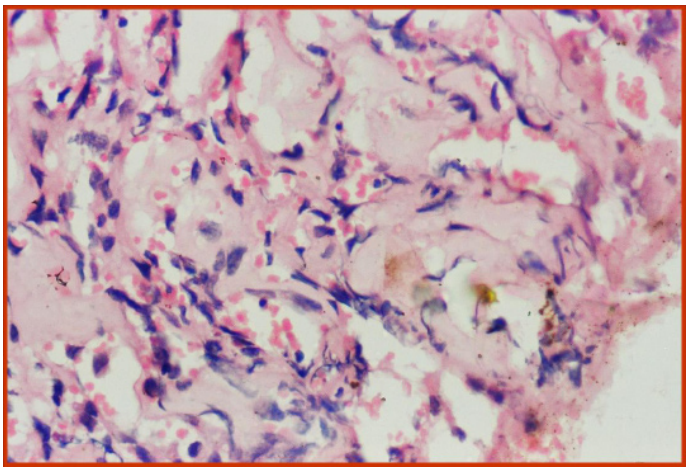
[Table/Fig-5]: Occlusal Radiograph shows “SUNRAY APPERANCE” on the lingual side and bicortical expansion



[Table/Fig-6]: CT showing bony expansion on the right body of the mandible



[Table/Fig-7]: Histopathology at 10X showing hyperchromatic, pleomorphic cells associated with extravasated RBC



[Table/Fig-8]: Histopathology at 40X showing proliferation of angular cells, with amorphous eosinophilic material suggestive of osteoid

DISCUSSION

Although osteosarcoma is generally the most common malignant bone tumour, the lesions of the jaw are rare [3] and despite its histopathologic similarities with osteosarcoma of the long bones, it is biologically different [1]. Therefore, small retrospective studies and case reports are opportunities to report and discuss issues of clinical and diagnostic significance.

Osteosarcomas arise in several clinical settings, which include pre-existing bone abnormalities such as Paget's disease, fibrous dysplasia, giant cell tumour, multiple osteochondroma, bone infarct, chronic osteomyelitis, osteogenesis imperfecta, and a history of radiation exposure [3,9,10]. In the present case, however, the aetiology remained unknown.

The clinical characteristics of the case which has been reported were in agreement with those of previous studies, regarding the age and chief clinical features [1,3,4,5]. Although the radiological and histopathological findings had strongly suggested an osteosarcoma of the jaw, the initial findings of the lesion were probably less suggestive. There is a need to be more conscious while such patients are being diagnosed, as they often go undiagnosed for a significant period of time. This is so, because some of these signs and symptoms can be produced by a number of different developmental infections, benign neoplastic diseases, or malignancies. Further, its presentation is similar to that of osteomyelitis with proliferative periostitis, suppurative osteomyelitis, ossifying fibroma, osteoblastoma, and even fibrous dysplasia, which has often caused an osteosarcoma to be delayed in its diagnosis [6,8]. Several diseases should be included in the differential diagnosis of JOS, since its most common symptom is a jaw swelling and as its radiological appearance may be a radiolucent, radiopaque, or mixed area [9,10].

The diagnosis of osteosarcoma is typically suspected by the radiographic appearance of the affected bone. Ossification in the soft tissue component of the bone, which manifests as a "sunburst" pattern is classic for osteosarcoma, but is not a sensitive or specific feature. Periosteal new born formation with lifting of the cortex leads to the appearance of a Codman's triangle. Garrington et al. mentioned that the roentgenographic evidence of a symmetrically widened periodontal membrane space was a significant early finding in osteosarcoma of the jaw, although the same features had been seen in some chondrosarcomas (Garrington et al). In the present case, the intraoral periapical radiograph didn't show any changes and the occlusal radiograph of the mandible showed the sunburst pattern. The extent of the tumour in both the bone and the soft tissue was best appreciated as was shown by cross sectional imaging techniques such as computerized tomography (CT) or magnetic resonance imaging(MRI). This is particularly important prior to a definitive surgery. A CT scan of an osteosarcoma often shows the formation of irregular endosteal and extracortical bone as well as a destroyed or obliterated cortex. However, CT scan cannot differentiate between osteosarcoma and fibrous dysplasia. Atypical sunray spiculations were seen in this case, which were highly suggestive of osteosarcoma [12-16].

Although MRI is generally accepted to be superior to CT scanning in the evaluation of the local tumour spread, Panicek and colleagues showed that CT scanning and MRI were equally accurate in the staging of the local disease in bone tumours (Panicek et al) [17]. However, in the present case, MRI could not be done due to financial limitations, as the patient belonged to a lower socioeconomic group. In the present case, it was observed that the lesion was mixed (radiolucent-radiopaque) in appearance, which was in accordance with the finding of Clark et al. classification [18].

Histologically, osteosarcomas can be classified according to their cellular differentiation as osteoblastic, chondroblastic and

fibroblastic. In the osteoblastic type, the atypical neoplastic osteoblasts exhibit considerable variation in their shapes and sizes, showing large deeply staining nuclei which are arranged in a disorderly fashion and this type constitutes 60% of the jaw lesions. The chondroblastic type has been described to occur predominantly in the head and neck region and it was found to consist of atypical chondroid areas which were composed of pleomorphic [1,16]. and atypical binucleate cells which had large hyperchromatic nuclei and prominent nucleoli. The fibroblastic type is rare, especially in the jaws. In the myxomatous type, there is atypical myxoid proliferation. A majority of the tumours are heterogenous, reflecting the pleuripotency of the proliferating mesenchymal cells. The other histologic types include a malignant fibrous histiocytoma like osteosarcoma which shows spindle anaplastic cells. In large cell predominant osteosarcoma, there are large cells with prominent nucleoli. Giant cell predominant osteosarcoma is characterized by anaplastic stromal cell producing streams of osteoid, along with giant cells. This type of a tumour may be confused with a giant cell tumour. In small cell or round cell predominant type osteosarcoma, osteoid producing, small malignant cell and primitive bone tissues are the characteristics, whereas in telangiectatic osteosarcoma, anaplastic cells are present along with the osteoid. In our case, histologically, the tumour was composed of hyperchromatic cells and pleomorphic cells which were associated with extravasated RBC. The decalcified tissue section showed vital bone with marrow spaces which exhibited the proliferation of the angular cells, some of which were in the lacunar spaces, some in association with an amorphous eosinophilic material which was suggestive of an osteoid, which was suggesitive of chrondroblastic osteosarcoma [17,18].

The treatment for osteosarcoma has been well established in the long bones, but it is not well understood when the condition involves the mandible or the maxilla [5,18]. It is clear that chemotherapy is beneficial for OS of the long bones, leading to significant changes in the disease-free survival rate (from 20% in the 1960s to 70% in the 1980s). This improvement did not include OS of the jaw, due to its rare occurrence and due to lack of standardized chemotherapy protocols, which made it difficult to evaluate the efficiency of the adjuvant therapy [3]. In most of the cases, the therapy of choice is radical surgical excision, since it provides a 5-year survival rate of over 80%. As for chemotherapy, it seems that it does not have much impact on the survival rates of the patients with OS of the jaws. This can be explained on the basis of the fact that the metastases were rare and late, occurring in only 18% of the cases and that the local recurrence of the lesion was still the leading cause of death [3]. In the present case, the patient had a radical surgical resection of the right mandible along with a margin of the normal surrounding tissue, followed by radiotherapy [17,18].

The prognosis of jaw osteosarcoma is better than that of the long bone osteosarcomas. This could be due to the histologically better differentiation of the jaw osteosarcomas than that of the long bone osteosarcomas [19]. As jaw osteosarcomas occur at higher mean ages, the patients have less chances of developing metastases.

CONCLUSION

It is very obvious that due to the common clinical presentations of pain and swelling, the patients with osteosarcoma usually will report to a dental professional first. Despite their adequate skill and knowledge, it is challenging for the dental professionals, as they usually come across osteosarcoma cases which have reached advanced stages and the prognosis in such patients is very poor. It can be concluded that misdiagnosis is very common in osteosarcoma of the jaw. Besides adding this case of osteosarcoma of the mandible in the dental literature, this article would catch the attention of the dental professionals so that they may approach such cases with greater concern and diagnose them at an early stage, which can lead to a better prognosis.

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