Case Report

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Pathology Section

Chondroblastic Variant of Osteosarcoma of Mandible: Report of a Rare Case

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ABSTRACT

Osteosarcoma (OS), a common malignant tumour of the long bones, is rarely seen in the craniofacial region (5-8%). Though it's aetiology is unknown, previous radiotherapy, Pagets disease, Retinoblastoma and benign bone lesions such as fibrous dysplasia are considered as predisposing factors. It is seen commonly in adults between the third and fourth decades of life, in the Gnathic location mandible. We report a rare case of chondroblastic variant of OS of the right mandible, in a 35-year-old male, who underwent right segmental mandibulectomy with fibular graft reconstruction and is having disease free survival one and half years post surgery. Craniofacial OSs, are considered a separate category in view of their low histologic grade, less frequent metastases and better prognosis. Hence the diagnosis of this variant is important. This case is reported because of its rarity and typical histopathological features.

Keywords: Craniofacial Osteosarcoma, Mandibulectomy, Prognosis

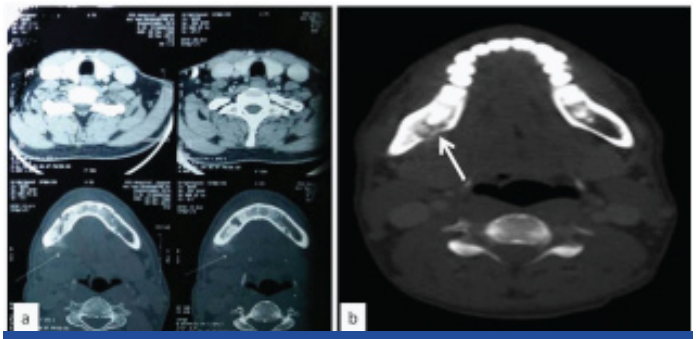
CASE REPORT

A 35-year-old male presented with swelling of the right jaw and history of weight loss of 2 months duration. There was no complaint of associated pain, fever and difficulty in swallowing. There was no history of previous exposure to radiation or any surgery.

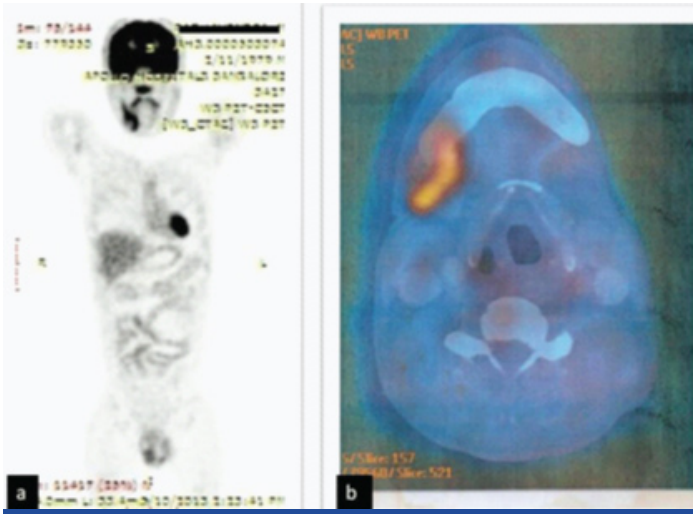
On examination, a firm to hard, non-tender mass measuring 4.0×5.0 cm, fixed to the right mandible was noted with extension into the right submandibular region. Fine Needle Aspiration Cytology (FNAC) was done but was inconclusive. Computed Tomography (CT) scan showed a sclerotic and lytic lesion of the right mandible with cortical breach and soft tissue component having mineralised matrix [Table/Fig-1a,b]. Positron Emission Tomography (PET CT scan) showed heterogenous mass lesion involving the right ramus of the mandible with exophytic partially calcified soft tissue component suggesting Osteogenic Sarcoma [Table/Fig-2a&b]. Patient underwent right segmental mandibulectomy with fibular graft reconstruction.

Right segmental mandibulectomy specimen grossly showed an expansile growth measuring 12.0×6.0×4.0 cm. Cut section showed a solid grey white firm mass measuring 6.0×3.0×3.0 cm seen infiltrating the medullary cavity [Table/Fig-3]. Histopathological study revealed a malignant neoplasm comprising of pleomorphic, polygonal to spindle shaped cells with hyperchromatic nuclei and moderate to scant cytoplasm. Lace like osteoid was seen in between the tumour cells with 3-4 mitosis/10 high power fields. Also, seen were multinucleated tumor giant cells [Table/Fig-4a,b]. Throughout the tumour, extensive cartilaginous differentiation of the tumour cells was seen [Table/Fig-4c]. The cartilaginous component was in the form of lobules having anaplastic cells within the lacunae and hypercellular spindle cells at the periphery [Table/Fig-4d].

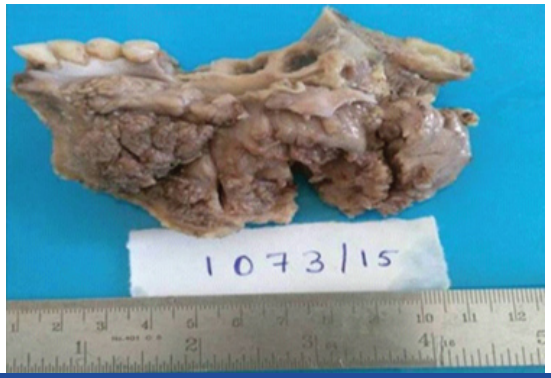
Sections from both the bony surgical margins and 4 lymph nodes retrieved were free of tumour and lymph nodes showed reactive hyperplasia. On follow up, the patient is doing well one and a half year post surgery [Table/Fig-5].



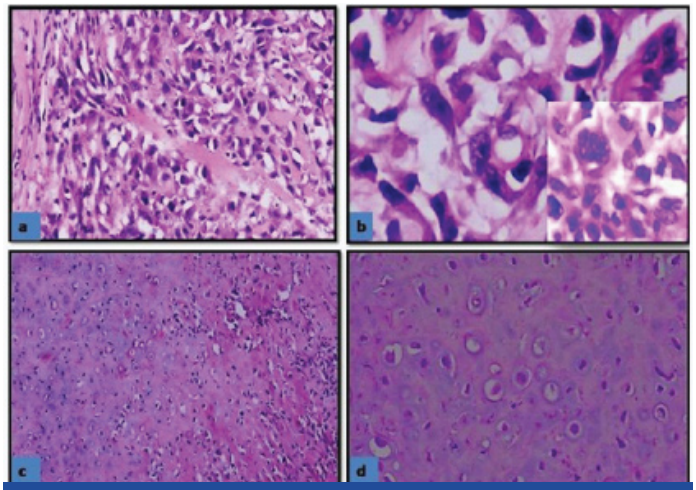
[Table/Fig-1]: Axial Contrast Enhanced CT scan showing breach in the cortex with soft tissue component having mineralised matrix extending into the right ramus of the mandible, favouring diagnosis of Osteogenic Sarcoma.



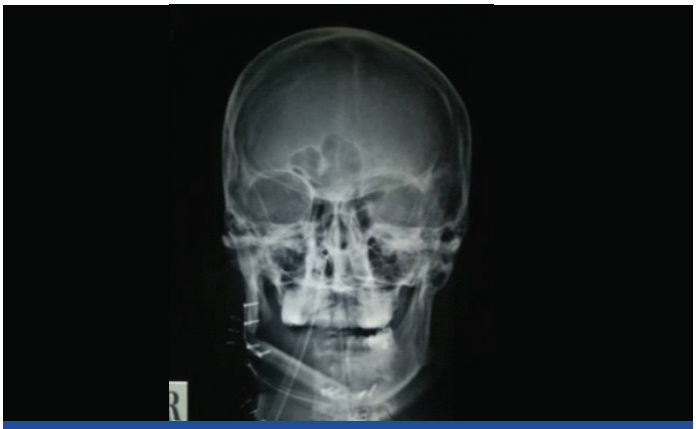
[Table/Fig-2]: PET CT scan showing heterogenous mass lesion involving the right ramus of the mandible with exophytic partially calcified soft tissue component suggesting Osteogenic Sarcoma.



[Table/Fig-3]: Mandibulectomy specimen with cut surface showing friable expansile growth extending into the medullary cavity.



[Table/Fig-4]: (a) Lace like eosinophilic osteoid matrix interspersed amidst the highly pleomorphic tumour cells {Haematoxylin and Eosin (H&E), x100}; b) Tumour cells showing hyperchromatic nuclei, mitoses and the inset shows multinucleated giant cells (H &E, x 400); c) Tumour showing extensive cartilaginous differentiation (H&E, x100); d) Pleomorphic cells within the lacunae in higher magnification (H&E, x 400).



[Table/Fig-5]: Anteroposterior X ray of the skull after mandibulectomy and graft reconstruction.

category [1,2]. However, Paparella et al., recorded maxilla as the common site followed by mandible [3]. According to Slootweg and Muller (1985), age can be an important prognostic factor in the differentiation of OS at different anatomical sites in the body. Some authors believed that older age group have better prognosis due to an increased resistance to the tumour [4].

The most common presentation is local swelling with or without pain [5]. Our case was a 35-year-old male, presenting with painless swelling of the right jaw. He had no prior history of irradiation.

Depending upon the predominant cellular component OSs can be classified into osteoblastic, chondroblastic and fibroblastic variants. Though, chondroblastic sub type is found to be the commonest [2,6]. Some authors have reported a predominance of osteoblastic type [7]. The important tumours whose histological differentiation from chondroblastic osteosarcoma is often difficult are chondroma, chondromyxoid fibroma, chondroblastoma and chondrosarcoma. The other clinico-radiological differentials include Ewing's sarcoma, metastatic tumours, fibrous dysplasia & also osteomyelitis.

In contrast to the prognosis of chondroblastic OSs of long bones, craniofacial OSs of chondroblastic type have a better prognosis. Most of the craniofacial OSs recorded, belong to the intermediate category (52%) closely followed by high grade (42%). Low grade OSs is rare in head and neck region [2]. Craniofacial OSs have varying clinical nature that has an impact on the disease progression, treatment and overall survival [8]. OSs of mandible is less aggressive than those of long bones, since they rarely metastasize. Prior irradiation is found to have influence on the prognosis. Disease recurrence is observed among OSs following irradiation [2].

Usually OSs of the jaw are detected by the dental professionals. The chondroblastic variant of OS has a favourable biological behaviour and therefore early diagnosis and appropriate treatment will benefit the patient [9]. In most of the cases, the therapy of choice is radical surgical excision because it provides a 5-year survival rate of more than 80%. As for chemotherapy, it seems that it does not have much impact on the survival rates of the patients with OS of the

Tumour or tumour like lesions	Age (years)	Sex (M:F)	Bones more commonly affected (decreasing order)	Usual location within the bone	Behaviour
1. Chondroma	10-40	1:1	Hands & feet, ribs, femur, humerus	Medulla of diaphysis	Benign
2. Chondromyxoid fibroma	10-25	1:1	Tibia, femur, feet, pelvis	Metaphysis	Benign
3. Chondroblastoma	10-25	2:1	Femur, humerus, tibia, feet, pelvis	Epiphysis adjacent to cartilage plate	Benign
4. Chondrosarcoma	30-60	3:1	Pelvis, ribs, femur, humerus, vertebrae	Central: medulla of diaphysis; peripheral: cortex or periosteum of metaphysis	Malignant. 5 year survival rate: low grade -78%, moderate grade – 53%, high grade- 22%
5. Mesenchymal Chondrosarcoma	20-60	1:1	Ribs, skull and jaw, vertebra, pelvis	Medulla or cortex of diaphysis	Malignant; extremely poor prognosis
6. Fibrous dysplasia	10-30	3:2	Ribs, femur, tibia, jaw, skull	Medulla of diaphysis or metaphysis	Locally aggressive; rarely complicated by sarcoma
7. Ewing's sarcoma	5-20	1:2	Femur, pelvis, tibia, humerus, ribs, fibula	Medulla of diaphysis or metaphysis	Highly malignant; 20-30%5 year survival rates
8. Bone metastasis	30-60	1:1	Femur, pelvis, vertebrae, tibia, humerus, jaw, skull, ribs	Medulla of diaphysis or metaphysis	Malignant; 22-50% 5 year survival rate

[Table/Fig-6]: Presentation and behaviour of primary bone tumours and tumour like lesions.

DISCUSSION

Craniofacial osteogenic sarcomas (OS) are extremely rare, whose aetiology is unknown. However, previous radiotherapy, Pagets disease, Retinoblastoma and benign bone lesions like fibrous dysplasia are considered predisposing factors seen commonly in adults between the third and fourth decades of life [1]. Takahama Junior et al., in their review series of 25 head and neck OSs, recorded an age range of 9-66 years with mean of 29 years [2].

Craniofacial OSs can be Gnathic and Nongnathic, with mandible being the predominant site followed by maxilla in the gnathic jaws, the reason being the fact that the metastases are rare and late, with low local recurrence of the lesion [10-12]. In addition, early diagnosis is favoured for aesthetical and functional reasons, especially in the maxillofacial region. In this case also, the patient did not have metastasis and the tumour was successfully resected with graft reconstruction.

Ancillary studies have limited role in the diagnosis of OS as the immunoprofile is not specific and histological features are very characteristic. The role of p53, MDM2, CDK4 & PCNA in the

prognosis of Chondroblastic Variant of OS of craniofacial region is controversial [5]. However, CDK4 and PCNA negativity have shown better prognosis [8].

Histopathological types of primary bone tumours and tumour like lesions with their clinical presentations and behaviour are listed in the [Table/Fig-6] [13].

CONCLUSION

Craniofacial OS often poses difficulty in diagnosis to clinicians, radiologists and pathologists due to its overlapping features with other benign and malignant bone lesions. This type being a comparatively rarer entity having low metastatic potential and with early treatment, the clinical outcome needs to be studied extensively.

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