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Chondroblastic Osteosarcoma: Clinical Outcome of Rare Entity

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Abstract
Introduction: Chondroblastic osteosarcoma (COS) is a subtype of osteosarcoma (OS).Osteosarcoma is sub classified into osteoblastic, chondroblastic, and fibroblastic types. 50% are osteoblastic, 17% are fibroblastic, and 33% are chondroblastic. Osteosarcoma of jaw bones is rare and comprises 6-9% of all osteosarcoma and less than 1% of all head and neck malignancies. The maxilla and mandible are involved with about equal frequency. Mandible tumors arise more frequently in the posterior body and horizontal ramus rather than the ascending ramus while Maxilla arises in anterior alveolus and antrum. The average age of onset of jaw lesions is in the 4th decade, with a mean age of 34 years, but cases have been reported in patients of all ages.
Case report: A 42 year old female, complaining of a swelling in right lower jaw since 10 month with no associated symptom like pain, difficulty in swallowing, breathing, wt. Loss, prior radiation, addiction etc. On basis of local examination, radiological & histopathological report, diagnosed as chondroblastic osteosarcoma (COS) of right body of mandible. Patient was managed surgically with adjuvant chemotherapy.
Conclusion: Chondroblastic variant has adverse features as compared to other subtypes of osteosarcoma. Prognosis of high grade & COS variant are poor but as by our reported case, early detection of tumour, its surgical management & timely adjuvant chemotherapy management can help to achieve good quality of life & overall survival benefit.

Key word: *Chondroblastic osteosarcoma (COS), osteosarcoma (OS), mandible, surgery, chemotherapy.*

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I. Introduction

Chondroblastic osteosarcoma(COS) as defined by WHO is a histological entity characterized by predominant presence of chondroid matrix, which tends to exhibit a high degree of hyaline cartilage and is intimately associated with the nonchondroid element (osteoid or bone matrix) [1]. Osteosarcoma is further sub classified into osteoblastic, chondroblastic, and fibroblastic types constituting 50%, 33% and 17%respectively. Osteosarcoma of jaw bones is rare and comprises 6-9% of all osteosarcomas and less than 1% of all head and neck malignancies [2]. The differential diagnosis of COS includes benign cartilage-forming lesions such as chondroblastoma, chondrosarcoma, chondroid chordoma (in some anatomic sites) and pleomorphic sarcoma with a chondroid component [3].

The maxilla and mandible are involved with about equal frequency. Mandibular tumors arise more frequently in the posterior body and horizontal ramus rather than the ascending ramus while Maxilla arise in anterior alveolus and antrum.

The most common signs and symptoms of OS are pain or paraesthesia, rapid growth, swelling and expansion of cortical bone, facial asymmetry, nasal obstruction, displacement and mobility of associated teeth [4]. In case of jaw lesion, the average age of onset of jaw lesions is in the 4th decade, with a mean age of 34 years, though case have been reported in all age groups. The differential diagnosis of COS includes benign cartilage forming lesions such as chondroblastoma, chondrosarcoma, chondroidchordoma and pleomorphic sarcoma with a chondroid component.

Due to the rare occurrence of COS in jaw bones, herein, we report a case of chondroblastic osteosarcoma in posterior body of mandible with emphasis on the clinical, radiological and surgical aspects of the tumor.

II. Case Report

A 42 year old female, reported to department of surgical oncology, AIIMS, Patna, Bihar India complaining of a gradually progressing swelling in right lower jaw for 10 months. She had no associated symptom like pain, difficulty in swallowing, breathing, wt. loss etc. she is not addictive.

On examination, a slight bulge was observed right face. There was no palpable head and neck lymphadenopathy. Intraoral lobulated and pink coloured growth seen in the right lower alveolus in the region of the molar & premolar teeth, involving sublingual space and floor of mouth. It was non tender and bony hard. (Fig 1)

Radiological CECT head and neck examination suggested expansile lytic lesion with adjacent sclerosis and indistinct border noted in the body of right hemi mandible with sunburst appearance in the region of the molar & premolar teeth. Mild periosteal reaction noted along the lingual side. Associated lobulated, heterogeneously enhancing, soft tissue lesion with few high density foci within, noted along the lingual and buccal side obliterating the right sublingual space, with loss of fat planes with the floor of the mouth muscles on the right side. Widening of the right mandibular canal. Edentulous ipsilateral molar and premolar tooth sockets (Fig 2).

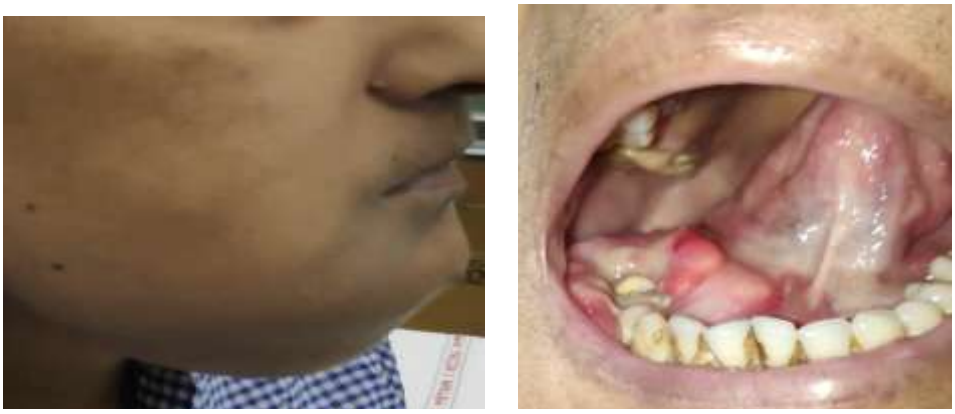

Fig 1(a&b). EXTRA & INTRA-ORALLY EXAMINATION

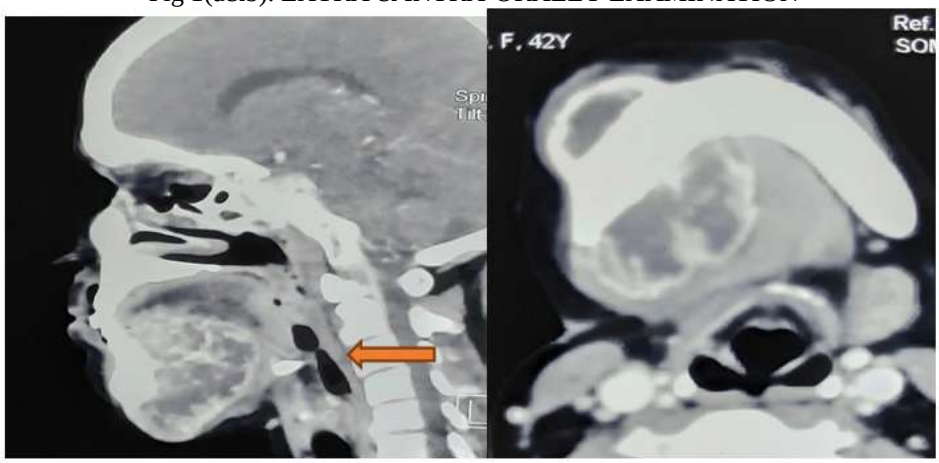

Fig 2(a&b). CECT HEAD & NECK

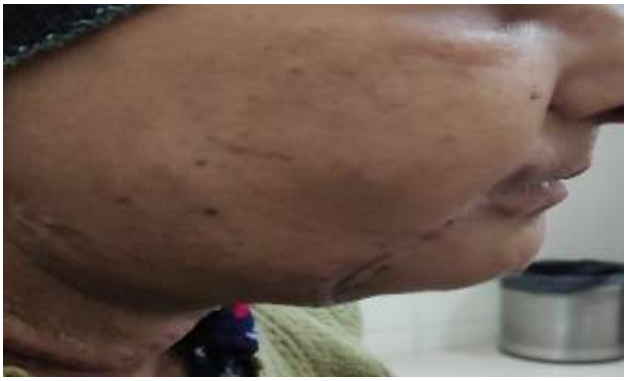

Fig 2. POST SURGERY 15THDAY

Histopathology of intraoral curettage of alveolar fragile soft tissue revealed moderated differentiated chondroblastic osteosarcoma.

Since there was no metastatic involvement, right segmental mandibulectomy with clearance margins of 1.5 cm with right selective neck dissection with PMMC (right Pectoralis major myocutaneous) flap done.

Final histopathology description: sections shows malignant mesenchymal tumor composed of predominantly chondrocytes arranged in lobular pattern with intervening osteoid. Chondrocytes display mild nuclear pleomorphism, hyperchromatic, centrally placed nuclei & abundant cytoplasm. . Binucleated & multinucleated chondrocytes are also noted. No LVI/PNI. All regional node free of tumor. IHC was positive for Vimentin and EMA. As per AJCC TNM classifications (eight edition), present tumour staged as stage III (T3N0M0, G3).

Post-surgical period was uneventful. She underwent adjuvant chemotherapy (cisplatin- doxorubicin). Post adjuvant chemotherapy period was uneventful &she is on regular follow up for last 6 months.

III. Discussion

Osteosarcoma is sub classified into osteoblastic, chondroblastic, and fibroblastic types. 50% are osteoblastic, 17% are fibroblastic, and 33% are chondroblastic. Osteosarcoma of jaw bones is rare and comprises 6-9% of all osteosarcoma and less than 1% of all head and neck malignancies [5]. Chondroblastic osteosarcoma is characterized by the production of chondroid matrix of variable cellularity, most commonly high-grade hyaline cartilage. Osteosarcoma is the second most common malignant bone tumour after multiple myeloma accounting for 15%-35% of all primary malignant bone tumors followed by chondrosarcoma and Ewing's sarcoma.

Pedruzzi *et al*.,[6] conducted a retrospective analysis on osteosarcoma of head and neck in 2006. They found the age group ranging from 13 to 66 years with mean age of 31.5 years. They also reported slight male predominance, with seven cases (53.8%) being men and six cases (46.2%) women. However, according to Chindia (2001), the age range of osteosarcoma cases is variable. Since the reported case refers to a 42-year-old female patient, it is possible to state that she is in agreement with the demographic profile presented in the literature on this tumour.

In the present case, the patient was asymptomatic at the time of diagnosis. Ajura& Lau (2010) [4], analysed 59 cases of osteosarcoma of jaw (OSJ) in their study and observed that most cases (77%) did not report any type of symptomatology, while only 15% of tumour cases presented pain and 8% paraesthesia. Similar character was observed in the study of 30 patients with OSJ by August et al.

Incidence of osteosarcoma secondary to Paget's disease, fibrous dysplasia and radiation induced osteosarcoma is 0.95%, 0.7% and 0.03% respectively. In this present case, however she gives no history suggestive of predisposing factors.

Smith Ac et al reported Maxilla and mandible are involved with almost equal frequency. Pedruzzi *et al*., [6] found that tumour was located in the mandible in eight cases (61.5%) and in the maxilla in five cases (38.5%) in a retrospective analysis study. Paparella et al., [7] recorded maxilla as the common site followed by mandible. Mandible tumors arise more frequently in the posterior body and horizontal ramus rather than the ascending ramus. Maxillary lesions are found in the inferior portion near the alveolar ridge, sinus floor, palate than the superior aspects such as the zygoma and the orbital rim. Clinicopathologically, the, the tumour in present case originated from posterior body of mandible.

CECT is an excellent method in the diagnosis of primary lesion to delineating the location, the extent of the tumour hence surgical planning. Radiographic findings vary from radiopaque to mix to radiolucent. COS constitute the "C" in the FEGNOMASHICmneumonic for radiolucent bone lesions. "Classic" sunray or sunburst appearance due to osteophytic bone production is an important feature. Garrington's sign with tapered resorption of tooth roots maybe present.Cotton balls, wisps or honeycomb pattern is seen.

Nakayama et al. [8] clarify the association between computed tomography (CT) findings, histologic features, and outcome of osteosarcoma of the jaw (OSJ).The present case falls into the group of osteolytic group. The **Lodwick classification** is a system for describing the margins of a **lytic bone lesion** (or **lucent bone lesion**) the present case showed radiographic evidence of type 1C pattern.

The predominant histological variants described are osteoblastic, fibroblastic and chondroblastic varieties. The jaw lesions are predominantly chondroblastic. Chondroblastic osteosarcoma(COS) as defined by WHO is a histological entity characterized by predominant presence of chondroid matrix, which tends to exhibit a high degree of hyaline cartilage and is intimately associated with the nonchondroid element (osteoid or bone matrix). However, Domanski et al. described smaller nucleoli in the chondroblastic variant. Binucleated and

multinucleated cells were rare in approximately 50% of all cases, and nuclear pleomorphism varied depending on tumour grade.

Immunohistochemical studies were used to confirm areas of chondrogenic and/or osteoblastic differentiation. Immunohistochemistry (IHC) will show COS will be positive for Vimentin, EMA, S100 and rarely Cytokeratin while Chondrosarcoma to be positive for S100 and Vimentin and negative for Cytokeratin and EMA (Epithelial Membrane Antigen). Amary et al; in 2011 & Darcy A. Et al; in 2013 reported the use of IDH1/2 mutational profiling in differentially diagnose of chondrosarcoma from chondroblastic osteosarcoma at the molecular level. This suggests that the identification of an IDH1/2 mutation strongly favours a Chondrosarcoma rather than a COS.

Hui-Hui Sun et al [9] found in his retrospective study that Younger onset age, white race, well and moderately differentiated tumors, no metastasis at diagnosis and surgical resection can independently predict better overall and cancer-specific survival of COS. Also Bennett et al, reported chondroblastic variant has better prognosis than others. However Brooder's grading of the tumor based on the degree of cellular anaplasia suggests high grade osteosarcoma to be always associated with poorer prognosis. The present case was staged as stage III (G3- high grade).

The treatment of choice in oral COS is surgical resection. Complete surgical excision with negative margins continues to be the mainstay of treatment. Borba *et al*. affirmed that complete surgical excision with negative margins continues to be the main pillar of treatment. Surgery may be complemented by radiotherapy and/or chemotherapy. Combined surgery and chemotherapy were used most often to manage high-grade tumors and metastatic disease. Chemotherapy can be used as neoadjuvant & adjuvant therapy. Neoadjuvant chemotherapy has been a procedure used for large tumors in an attempt to minimize tumour size and provide a less aggressive surgical approach. The European Osteosarcoma Intergroup established a protocol which consists of cycles of Cisplatin and doxorubicin. The Brazilian Protocol for Metastatic and No Metastatic Osteosarcoma also made a protocol with addition of ifosfamide. Lewis IJ et al in a randomized phase III trial of the European Osteosarcoma Intergroup revealed that intensification of chemotherapy did not improve the overall survival.

Russell et al. [10] published National Cancer Data Base Report on Osteosarcoma of the Head and Neckto assess patient demographics, tumor characteristics, treatment, and outcome for an 11year period extending from 1985 to 1996. 5-year survival rate for patients with mandibular tumors was almost 10 % higher than patients with tumors located in the craniofacial skeleton. Patients with smaller tumors (< 6 cm) demonstrated a > 60% 5-year survival rate, whereas patients with larger tumors had a 5-year survival rate of only 26%. Michael Herman Chui et al reported high-grade osteosarcoma and chondroblastic differentiation are adverse features in his study of Histopathologic Features of Prognostic Significance in High-Grade Osteosarcoma.

IV. Conclusion

Osteosarcoma (OS) of jaw bones is rare and comprises 6-9% of all osteosarcoma and less than 1% of all head and neck malignancies. Chondroblastic osteosarcoma (COS) is a subtype of osteosarcoma. Chondroblastic type has adverse features in compare to other subtype of OS. Prognosis of high grade & COS variant of OS is poor but, as by our reported case, early detection of tumor, its surgical management & timely adjuvant chemotherapy management can help to achieve good quality of life & overall survival benefit.

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