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Research Article

HIGH GRADE VARIANT OF CONVENTIONAL CHONDROSARCOMA IN MANDIBLE: A RARE CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Purpose: High grade variant of mesenchymal chondrosarcoma in mandible is a very rare tumor documented in years. Hence, we have performed analysis of the reported cases of high grade variant of mesenchymal chondrosarcoma with emphasis on its treatment modalities. ***Material and Methods:*** We report a case of a 25 year-old man with pain and swelling which was in rapid onset in nature on right body region of mandible. This review also includes the two similar type of cases and analyzed the data of the patients, lesion site, treatment approach and events of recurrence. ***Results:*** Only three patients of high grade variant of Chondrosarcoma of jaws has been reported till date including this case. The primary treatment in all patients was radical surgery consisting of resection with clear margins. Out of this 2 patients were undergone for radiotherapy. ***Conclusions:*** This article highlighted the potentially aggressive biological behavior of high grade variant of mesenchymal Chondrosarcoma. Inspite of surgical resection with clear margins and giving radiotherapy possible outcome was not achieved. However, due to the rarity, more well-documented cases with long follow-up periods are needed in order to further define the optimal treatment modalities and prognosis.

INTRODUCTION

Chondrosarcoma is a rare tumor presumably derived from mesenchymal cells or embryonic remnants of the cartilaginous matrix. It represents some 17 to 22% of all monostotic bone malignancies.[1]It accounts about 10% to 20% of all malignant bone tumors and it's incidence in the head and neck region is even more uncommon, accounting for less than 10% of all cases, affecting more commonly the mandible, maxilla, paranasal sinuses and nasal cavity.[2]

The majority of Chondrosarcoma of jaws is diagnosed as low grade variant. Only two aggressive high grade variant of Chondrosarcoma has been reported till date.[3] This article reports case of high grade variant of Chondrosarcoma in the right mandible in a 25 year-old man and describe its clinicopathological features, radiological images and treatment performed.

CASE REPORT

A 25-year-old male patient reported to the department with complaint of swelling and pain in right body region of mandible since 1 month. Initially patient noticed small sized swelling over right body region which was aggressively increased upto 3 × 3cm(Fig 1).

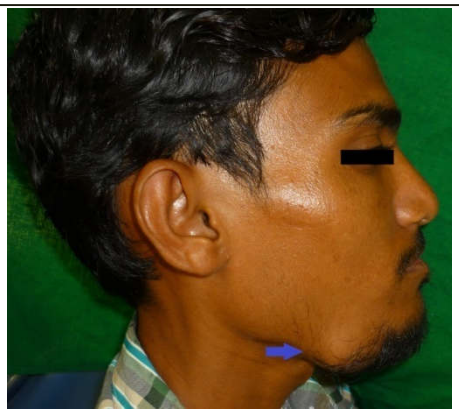

Figure 1 Painless swelling affecting the right side of body of mandible.

The medical history of the patient was non-contributory. The extraoral examination of mid body region revealed a diffuse ill-defined swelling seen on inferior border of mandible. On palpation the swelling was hard, immobile, nontender and non-fluctuant. The overlying and adjacent skin was normal. There was no evidence of lymphadenopathy or neurological signs.

Intraorally diffuse swelling was seen extending from 44 to 48 with obliteration of buccal and lingual vestibule. The overlying mucosa was intact. It was firm in consistency and tender on palpation. The lower right premolarsand molars were vital and showed no mobility.

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Table 1 Overview of published cases on histologically confirmed high grade variant of mesenchymal Chondrosarcoma

Sr No	Author	Age/Sex	Site	Diagnosis	Margins	Treatment	Follow-Up
1	Pontes et al[11]2011	26/M	Left Retromolar Region	Grade III CHS	Negative	Surgery	The Patient Died After 2 Year Of Follow Up
2	Ademar Takahama Jr et al[2] 2012	47/F	Left Maxilla	Grade III CHS	Positive	Surgery And Adjuvant Radiotherapy	After 8 Months The Patient Died Of Local Recurrence
3	This Case	25/M	Rt Angle Of Mandible	Grade III CHS	Negative	Surgery And Adjuvant Radiotherapy	The Patient Died After 1 Year Of Follow Up

An orthopantomogram (OPG) (Fig.2) showed hazy radiopacity with ill-defined borders in the right region of mandible resembling like typical sun ray pattern suggestive of osteosarcoma. Computed tomography (CT) showed an expansile osteolytic lesion involving the right body destroying both the buccal and lingual bone cortices. A significant number of calcified structures were also noted.Under local anesthesia, an incisionalbiopsy was performed. The histopathological report was suggestive of Chondrosarcoma.

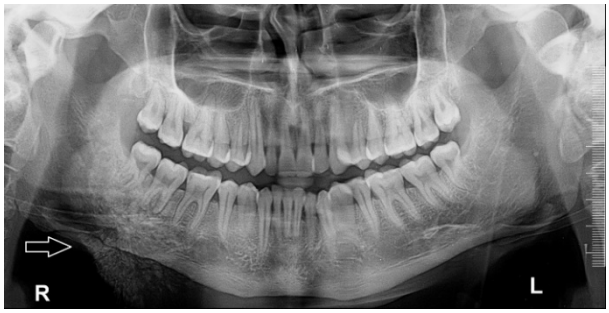

Figure 2 Panoramic radiograph reveling Hazy radiopacity with ill-defined borders resembling like typical sun ray pattern

The patient underwent surgery and using submandibular access, the neoplastic growthwas widely resected by segmental mandibulectomy with safe margins.The postoperative histological(Fig 3) evaluation of the surgical specimen reveals extremely cellular consisting of sheet of oval to spindle shaped, highly pleomorphic, undifferentiated malignant cells showing features like extensive cellular nuclear pleomorphism, hyperchromatic bizarre nuclei, altered N:C ratio, cells showing vesicular nuclei with fragmented nucleoli and mitotic figures. At places malignant chondrocytes have formed lacy stroma of faintly basophilic chondroid tissue. Partial calcification of those malignant chondroid is seen. On grossing the margin of all the sides were free of disease. The high grade variant of chondrosarcoma was final diagnosis. Patient immediately send for adjuvant radiotherapy.

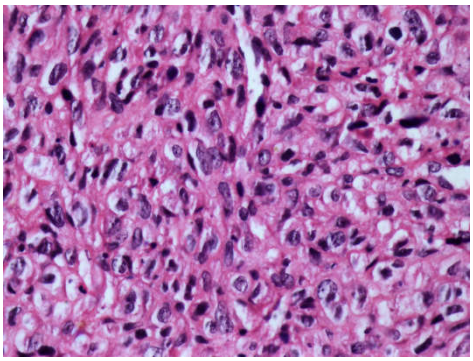

Figure 3 Histologic features obtained, revealing clusters of enlarged chondrocytes with Hyperchromatic nuclei, binucleation. (Haematoxylin–eosin (H–E), 40x)

Patient was kept in strict follow-up. After 6 month patient came to the department with complaint of backache, radicular pain, weakness in both lower limb and difficulty in micturition. Sagittal magnetic resonance imaging (MRI) (Fig 4) showed the presence of metastases affecting lumber vertebrae with compression of bone marrow causing significant pain. USG of liver showed multiple, well defined, hyperechoic lesion of varying sized scattered throughout the liver in largest measuring in IVa of left lobe suggestive of possibly metastasis. The patient died after 1 yr of follow-up.

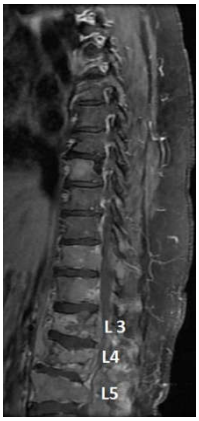

Figure 4 MRI revealing lumbar (L3 L4 L5 vertebras) metastases.

DISCUSSION

Chondrosarcoma is an extremely rare malignant tumor of the head and neck region, representing less than 0.1% of all neoplasms in this location.[3] Patients affected by Chondrosarcoma of the head and neck are usually in the fourth decade of life with a small male preponderance and usually complaint of painless swelling similarly our patient described the complaint.[4]

The radiographic appearance of Chondrosarcoma of the jaws may be varied, but it usually exhibits features consistent with malignancy, primarily presenting osteolytic lesions with poorly defined borders.[5,6] The ill-defined radiolucent areas often contain scattered radiopaque foci corresponding to calcification of the neoplastic cartilaginous tissue.[7,8] In those cases in which Chondrosarcoma involves the teeth, the radiographic appearance may also mimic osteosarcoma and demonstrate a symmetrically widened periodontal space. A hazy radiopaque periphery suggestive of a sunray pattern can also be found in several cases.[5]In our case typical sunray pattern was seen.

Histologically, the Chondrosarcomas can be classified according to the microscopic appearance into conventional, clear cell, myxoid, mesenchymal and dedifferentiated chondrosarcoma.[2] The conventional Chondrosarcomas are characterized by a lobulated malignant cartilage, nuclear pleomorphism, mitosis and binucleated cells.[6] Evans classified conventional chondrosarcoma into three grades (I-II, III) based on cellularity, frequency of mitosis and nuclear dimension. It is also possible to divide chondrosarcoma into two groups: low grade (Evans' grade I-II) and high grade (Evans' grade III) .[7]

Low grade chondrosarcoma (grade I) is very close in appearance to enchondromas and osteochondromas, has occasional binucleated cells and may show atypical cells including binucleate forms (cells with two nuclei instead of one). Calcifications and bone formation can be found, but can also be characteristic of higher grade tumors. Grade II (or "intermediate grade") presents a higher cellular population with a greater degree of nuclear atypia, hyperchromasia and nuclear size. The mitotic rate is low (less than two per 10 high power fields). High grade (grade III) chondrosarcoma has significant areas of marked pleomorphism, large cells with more hyperchromatic, denser and greater nuclei size than grade II, occasional giant cells and abundant necrosis. Mitoses are more than three per field .[7]

The main modality treatment for head and neck Chondrosarcomas is surgical resection of the primary tumor with adequate margin.[8] Radiotherapy has been used mainly in high-grade tumors, unresectable tumors or even as palliative. The efficacy of adjuvant therapies is still not clear, but some authors have noted acceptable outcomes with the use of radiotherapy, which is generally indicated for unresectable tumors and for residual disease after surgical excision.[4] More resistance to irradiation than radiosensitivity of Chondrosarcomas is also reported.[9] Preoperative and postoperative chemotherapy have also been used with beneficial effect in some cases.[10] In our case, surgery was the main modality of treatment being performed on the basis of incisional biopsy report . But after revealing the tumor was high grade we send the patient for radiotherapy immediately.

The incidence of local and distant metastasis in patients with Chondrosarcoma is low occurring in 5% of the cases, and generally occurs as a late stage event.[4] The main cause of death in patients with head and neck Chondrosarcomas is local recurrence, with the tumor locally involving vital structures.[9]

Tumor grade and resectability are the most important prognostic factors for head and neck chondrosarcomas. Tumor site is another important prognostic determinant. Factors indicating poorer prognosis include histologically positive margins and high-grade tumor differentiation (Grades II and III). The prognosis for Chondrosarcoma in general is sobering, particularly in consideration of the mesenchymal variety, for which the 5-year survival value is 42 to 54.6% and the 10-year value is 28%.[1] Conversely, viewed in terms of the grading system, the prospects are somewhat less alarming; the 5- year survival rates for grades I, II, and III have been recorded as 90%, 81%, and 43% respectively, and the 10-year rates have been recorded as 83%, 64%, and 29% respectively.[7]

CONCLUSION

Chondrosarcoma are rare malignant mesenchymal tumors of the head and neck region. Inspite of surgical resection with clear margins and giving radiotherapy possible outcome was not achieved. So Radiotherapy may not be beneficial for the high grade Chondrosarcoma. A long-term study of combined treatment with surgery and adjuvant radiation therapy or chemotherapy is needed to confirm the best approach in the management of these lesions.

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