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MANDIBULAR CHONDROSARCOMA: CASE REPORT WITH VARIOUS DIAGNOSTIC ASPECTS

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ABSTRACT

Chondrosarcoma (CHS) is a slow growing, malignant neoplasm of cartilaginous origin. CHS involving head and neck region is a rarity and represents 0.1% of all head and neck neoplasms. This tumor displays a wide spectrum of clinic-biologic, radiological and pathological features. This report describes a rare case of CHS mandible in a 62 year old woman with relevant review of literature. Patient's chief complaint was a hard, non-tender swelling along the alveolar border of mandible, with fixity of overlying skin. Different imaging findings parallel with histological features help in arriving at diagnosis of CHS Mandible Grade II.

INTRODUCTION

Chondrosarcoma (CHS) affecting the cranio-facial region is a very rare phenomenon, accounting for less than 10% of all CHS [1]. This tumor can arise from any cartilaginous tissue of the body but it usually emerges from the axial skeleton, pelvic girdle, femur, humerus, vertebra, shoulder, sternum, or ribs [2]. CHS has predilection for anterior part of maxilla than the mandible [3]. Though CHS can involve any age group but more frequently encountered between 3rd and 6th decade of life, with a slight male predominance. The tumor is believed to originate from primitive mesenchyme but the exact genealogy of this malignant conditionin cranio-facial region is under controversy [4]. The grading is based primarily on nuclear grade, nuclear staining and cellularity [5].

Grade I: Tumors are moderately cellular and contain hyperchromatic plump nuclei of uniform size. Occasionally binucleated cells are present. The cytology is very similar to enchondroma.

Grade II: Tumors are more cellular and contain a greater degree of nuclear atypia, hyperchromasia and nuclear size.

Grade III: Tumors are more cellular and pleomorphic and atypical than grade II. Mitoses are easily detected. We present a case-report of CHS of mandible in a 62 year old female with a discussion on the diverse clinical, radiological and histologic feature that are usually encountered in CHS of mandible compared to CHS elsewhere in the body.

Aim

The aim of this case-report is to present the entire spectrum of clinic-biologic, radiological and pathologic features of this rare entity affecting the mandible which are suggestive but not diagnostic of disease.

Case-report

A 62 year old female of low socio-economic strata reported to the OPD of Mahavir Cancer Institute and Research Centre, Patna with complaint of a painless swelling involving the right side of face near the angle of mandible. She got operated 13 months back in the Surgical Oncology Department,segmental mandibulectomy with excision of 4 teeth was done. The patient relapsed 11 months after surgery.

Examination revealed a large swelling measuring 8*6*5cms along the alveolar border of mandible with

fixity of the overlying skin. The swelling was non-compressible, hard, non-tender and non-pulsatile on palpation. Intra-orally, the lesion was involving the buccal and lateral aspect of mandible.

OPG and CT Scan of neck and mandible demonstrates an ill-defined heterogeneous enhancing mass lesion with lytic and necrotic areas along the residual mandible, the spiculated periosteal reaction gave the so-called hair-on-end appearance. X-ray chest and USG Abdomen didn't show any significant findings.

Microscopic examination of the resected specimen revealed lobular areas of neoplastic/dysplastic chondroid matrix separated by fibrous bands with focal areas of myxoid change (Figure 1-2). Cells of tumoral lobules showed moderate degree of nuclear pleomorphism and atypia.Binucleated or multinucleated chondroblast in lacunaewere also visualized (Figure 3-4). No area of malignant osteoid was found. Abundant necrosis and occasional giant cells may be seen in high grade CHS [6]. These findings viewed in conjunction with findings of other imaging techniques were found conclusive of Chondrosarcoma Grade II. The patient underwent hemi-mandibulectomy with lymph node dissection. No chemo-radiation was given. Patient didn't return after 9 months of follow-up.

Figure 1. Scanner view (4x) showing lobular areas of chondroid matrix(CM) separated by fibrous bands

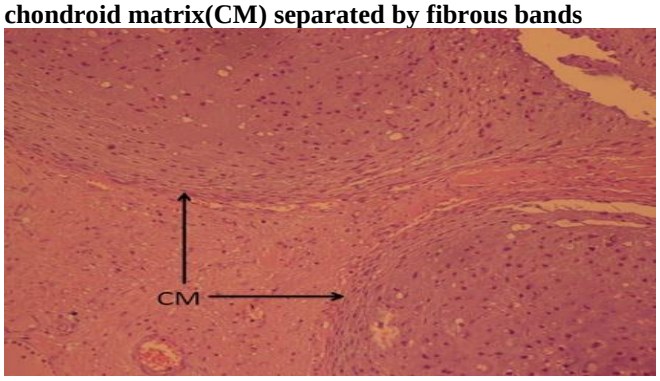


Figure 2. Low power (10x) showing dysplastic chondroid matrix

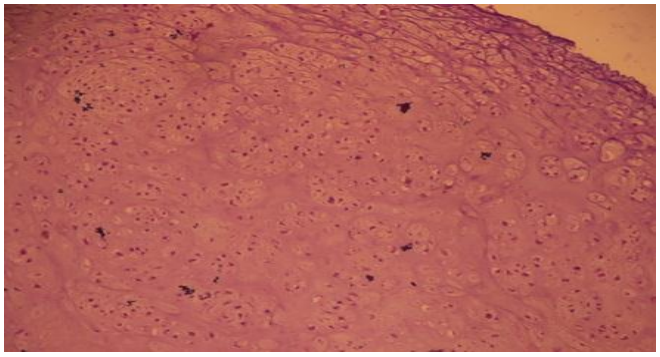


Figure 3. High power view (40x) showing bi-nucleated (BN) and multinucleated (MN) cells

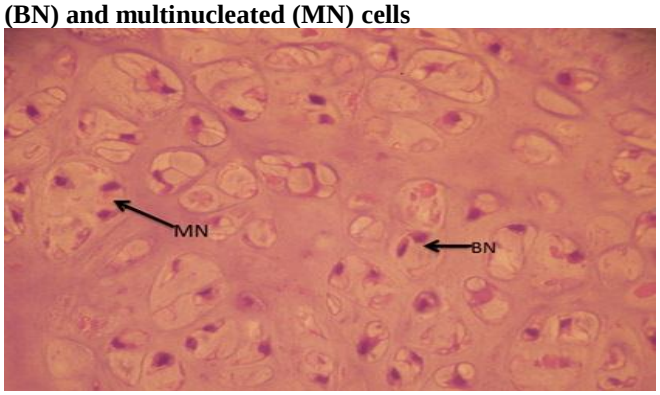
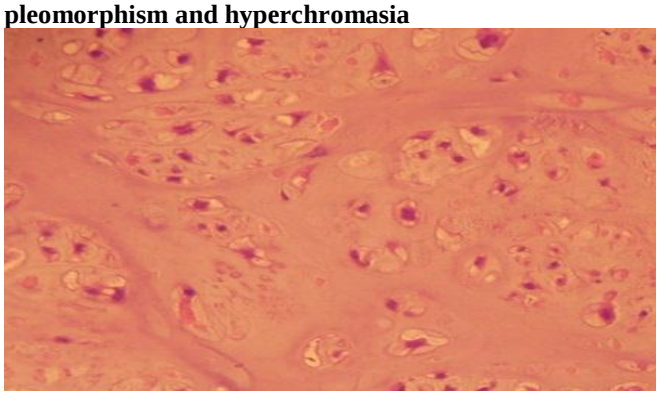


Figure 4. High power view (40x) showing nuclear pleomorphism and hyperchromasia



DISCUSSION

The importance of discussing mandibular CHS lies in its rarity at this site. The clinical picture in CHS is variable, ranging from painless swelling to painful ones. The 62 year old female presented with the chief complaint of a painless swelling with limitation of mouth opening. In the literature Richter et al reported male predominance for CHS of mandible [7].

The conventional radiographic findings in CHS include irregular radiolucent areas with spiculated periosteal reaction along the body of mandible, though sometimes the overlapping complex cranio-facial bones may produce difficulties in assessing the lesion. Chondrosarcoma involving the ramus, head, or even TMJ is an absolute rarity [8]. CT is superior in defining peripheral extend of the neoplasm. Aforesaid findings are highly suggestive of CHS, but may be present in osteochondroma and other metastatic lesions [9,10].

CHS is highly variable with reference to its histopathological findings. CHS is graded on a scale of 1-3. Various studies have found it useful in predicting clinical outcome and prognosis.

Due to its wide range of histopathological features it might be confused with chondroblastic osteosarcoma and enchondroma. But chondroblastic osteosarcoma can be ruled out by the absence of neoplastic osteoid, neoplastic bone or alkaline phosphatase expressing tumor cell [11].

Hypercellularity and permeation of cortical/medullary bone in CHS helps in differentiating from enchondroma.

Prognosis of the patients of CHS is highly variable depending on the size, location, grade of the lesion and positivity of the resection margins. Shafer et al reported that CHS of mandible is a very rare entity and usually have a fatal outcome.

CONCLUSION

Mandibular CHS, being so rare at this site and radiographic simulation to other entities may pose diagnostic challenge. But adequate biopsy allows us to reach on a definitive diagnosis. It requires surgical resection with wide margins. Role of chemo-radiation in CHS is not proven yet. Though high recurrence rates and metastasis in mandibular CHS stresses upon the importance of regular follow-up of the patient.

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