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Oral Pathology

A RARE CASE OF CHONDROSARCOMA OF MAXILLA

Dr. Shatakshi Sharma*

MDS, Oral and Maxillofacial Pathology, Town Dental Advanced Multispecialty Dental Clinic & Oral Cancer Diagnosis Centre, Jammu, J&K. *Corresponding Author

ABSTRACT Chondrosarcoma is an unusual potentially fatal malignancy of any age group. The present paper reports one such case of intermittent grade chondrosarcoma of right side of maxilla in a 35-years old male patient. The lesion initially presented as a firm, painless, nodular growth in molar region and later increased gradually to attain a larger size with a mild pain on palpation, associated with bicortical expansion. Maxillofacial radio-imaging identified a radiolucent lesion with ill-defined borders with mildly displaced maxillary molars. Osteo-degenerative changes were evident and affected the alveolar bone, maxillary antrum walls and floor of orbit. Considering to the age of the patient and the extent of spread, surgery was selected as the treatment of choice in adjunct to radiotherapy and the patient was referred to PGI, Chandigarh without any further delay. This case report explains the clinical, radiological, histopathological investigations employed for chondrosarcoma.

KEYWORDS : Nodular growth, chondrosarcoma, maxillary lesions, malignant oral tumor

INTRODUCTION

Chondrosarcoma is a rare, slow growing, malignancy the origin of which is from the cartilaginous tissue or may be from the osseous tissue derivatives of chondroid precursors that result in the formation of abnormal bone and/or cartilaginous tissue.[1] It accounts for the second most frequent group of bone tumors next to osteosarcoma[2], with only 1-3% of cases occurring in head and neck region[3] and is only 0.1% of all malignant tumors in this region. This tumor affects any bone from within or occurs on its surface [4]and is most commonly seen in association with the bones of the scapula, chest wall and pelvic girdle.[5] The majority of chondrosarcomas in the head and neck region occur in larynx, thyroid cartilage, and arytenoids.[6] However, other sites like maxilla, mandible, nasopharynx, paranasal sinuses, and the base of skull, where cartilage is found may also be affected.[7] Chondrosarcomas of head and neck region, are slightly more predominant in men than when compared to that in women and is mostly seen in third to sixth decade of life.[4] This report presents a case of a intermediate grade Chondrosarcoma involving the maxillary bone, antrum and orbit in a 35-years-old male patient and reviews the clinico-histopathology and treatment of the same.

CASE-REPORT

A 35-year-old male patient reported to the Dental Care Centre, in August 2018 with the chief complaint of an intraoral growth on the right side of his maxilla. The patient noticed the lobular mass approximately 3 months ago. There was no significant observable asymmetry of the face (Figure 1). There was no apparent abnormality during extra-oral examination. Intraoral examination exposed the existence of a painless, discoid swelling about 3 cm x 2cm on the right side of the palate, extending from second premolar to the last molar area up to the maxillary tuberosity on the palatal aspect of right side of the jaw (Figure 2). On palpation, swelling was firm in consistency. It was non compressible, non fluctuant and mildly tender. An evident egg shell crackling was present over some areas of cortical plate. Radiographical investigations showed a mixed lesion seen i.r.t. 16, 17 in a Lateral occlusal view (Figure 4).

Computed Tomography (CT) imaging showed a large, ill-defined, expansile, lytic lesion of size 5.3 cm x4.2 cm x4.2cm involving the right maxillary sinus and alveolar arch of maxilla. It also revealed the inferio-medially destruction of alveolar arch and hard palate with extension of mass in oral cavity. Superiorly, it had caused erosion of floor of orbit. Medially mass was seen to be extending into nasal cavity causing erosion and destruction of nasal turbinates (Figure 5). The differential diagnosis of malignant neoplasms was made, since the lesion, clinico-radiographically, appeared to be aggressive. The differential diagnosis of salivary gland malignancies like muco-epidermoid carcinoma and adenoid cystic carcinoma were chiefly thought of, as these usually arise in relation to the palate. Both of the salivary gland lesions lack bicortical expansile behavior in contrast to the current case. Also included in the differential diagnosis was Carcinoma of maxillary antrum as it is a typical disease in adults and almost always associated with a deleterious habit. Mesenchymal Malignancies such as osteosarcoma and chondrosarcoma were included in the differential although these are rarely encountered in craniofacial region. Both Hodgkin's and non-Hodgkin's lymphoma can also be seen as palatal masses with ulceration. These are mostly present along with cervical lymphadenopathy, which in the current case, was not seen. An incisional biopsy procedure was carried out under local anesthesia, and the collected surgical specimen was histopathologically evaluated (Figure 3). Histopathologic and microscopic investigations revealed a hyper-cellular connective tissue stroma with abundant cartilaginous matrix containing round and ovoid cells in lacunae. These cells exhibit nuclear pleomorphism, atypical nuclei, and nuclear hyperchromatism. Histopathology also revealed a moderate grade mitotic activity with large plump chondroblasts and bi-nucleated chondrocytes seen in a few microscopic fields. The final diagnosis of intermediate grade chondrosarcoma of right maxilla was made. (Figures 6, 7, 8). Patient was referred to PGI, Chandigarh with no further delay.

DISCUSSION

Earlier to 1930, Chondrosarcoma was classified as a variant of osteosarcoma. It was in 1930 that Pheimeister described chondrosarcoma as a separate entity.[8] The site of occurrence of this lesion, in head and neck region differs as per different authors in various publications with maxillary anterior region and posterior mandibular region being more prevalent.[9] The peak age of occurrence in adults is between 3rd and 6th decades of life, but it can be seen in any age, with the youngest patient reported being just 1year 4months old and the oldest one being an elderly man aged 74-years.[10] Chondrosarcoma has a male predilection with a ratio of 1.2 : 1.[3] This tumor has a high rate of local recurrence.[11] Chondrosarcoma is thought to be derived from mature cartilaginous tissue and is considered to be a malignant tumor histogenetically. Exclusive membranous ossification is seen in case of maxilla so the chance of chondrosarcoma occurring in maxilla is hard to explain. But, it is considered to arise from cartilage of incisive papilla, cartilaginous remains in periodontal ligament, para-septal cartilage and focal cartilaginous portion of nasal capsule. Thus, explaining the notion that maxillary chondrosarcomas are derived from cartilaginous differentiation of primitive mesenchymal cells and not from nests of embryonic cartilaginous remenants.[12] The chondrosarcoma of head and neck region present as painless lesions, unlike the high grade chondrosarcoma of the long bones that present with excruciating pains, with the common symptoms of head and neck tumors being reported as swelling/mass (68%), nasal obstruction (32%), epistaxis (32%), and tooth mobility (24%).[13] Expansion of the mass sometimes may cause other symptoms like headache, blurred vision, proptosis, diplopia, facial swelling, with evidence of cervical lymphadenopathy in certain cases.[14] This present case being discussed was diagnosed after a period of 3 months as a painless swelling and mild tooth mobility. A systematic radiological examination was included in the primary investigations and comprised of an occlusal radiograph as well as CT scan. The conventional radiographic findings revealed an irregular intraosseous mixed radiolucent-radiopaque lesion with cortical plate expansion and destruction, widening of PDL

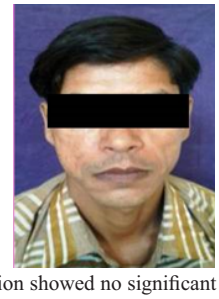
spaces which was similar to the findings observed by previous authors.[2, 3, 15] CT scan shows a more superior image of the peripheral extent of the lesion and detection of calcifications when compared to panoramic radiographs and MRI respectively.[16] CT scan verified an ill-defined cloud-like matrix with calcified whorls and arcs (Figure 5) which was again in comparison with the previous reported cases.[6] In Case of Chondrosarcoma, the final diagnosis can only be made on the basis of Histopathological findings. Lichtenstein and Jaffe are credited with the formulation of a criterion for the histopathological diagnosis of chondrosarcoma.[17] The histopathological variants of chondrosarcoma are classified on basis of the microscopic appearance as a) conventional, b) clear cell, c) myxoid, d) mesenchymal, and e) dedifferentiated.[17] Chondrosarcoma is a malignant tumor that presents histopathologically mature cartilaginous tissue without evidence of tumor osteoid, originating directly from a sarcomatous stroma.[18] The malignant tumor may also exhibit myxoid changes, calcifications, and ossifications.[18] Histological grading of chondrosarcomas is based on their degree of cellularity, mitotic activity, nuclear size, and the composition of the surrounding matrix as grade I, II & III.[15] The present case being discussed showed an intermediate grade conventional chondrosarcoma. As this entity is rarely reported in the jaws and no definite data based treatment protocols are confirmed[6] so chondrosarcomas are generally treated with multiple modal approaches including 1) Wide en-bloc resection[19], 2) Local curettage[20], 3) Cryotherapy[20], 4) Chemotherapy[21], 5) Radiotherapy[22], and 6) Immunotherapy[23]. The treatment should be decided depending on age and sex of the patient also considering the size and extent of tumor. In this case, we opted to refer the patient to a higher medical facility in interest of the needs of the patient as the clinic setup did not permit us to carry out the required treatment protocol and also that the recurrence rate of chondrosarcoma is high if not investigated and treated properly. Regular Follow-up and repeated investigations may be required because of high rate of local recurrence rate (20-60%) and distant metastasis.[24] The prognosis as per previous cases is reported to be good for grade I & grade II chondrosarcomas.[25] 90% of grade I chondrosarcomas have been reported to have a five-year survival rate that lowers to 81% for grade II chondrosarcomas and 43% for grade III chondrosarcomas so early detection may be helpful.[24]

CONCLUSION

Chondrosarcoma is a rare malignant tumor entity of head and neck region. A timely and precise detection of this malignancy helps to formulate an appropriate treatment plan. Considering patient history, the character of tumor, and high rate of recurrence, a suitable multi-step treatment regime must be elected with mandatory, regular, long term follow-up sessions for better prognosis.

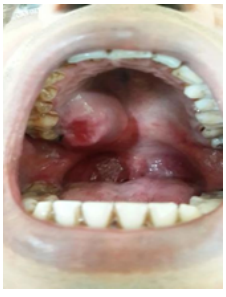
Figures

Figure 1



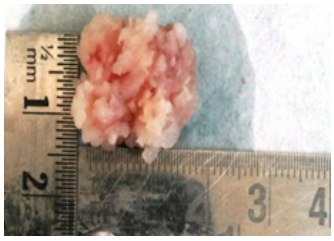
Extra-oral examination showed no significant observable asymmetry of the face.

FIGURE 2



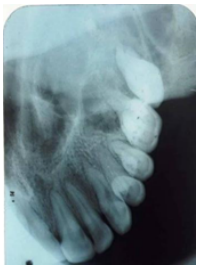
Intraoral examination exposed the existence of a painless, discoid swelling about 3 cm x 2cm on the right side of the palate.

FIGURE 3



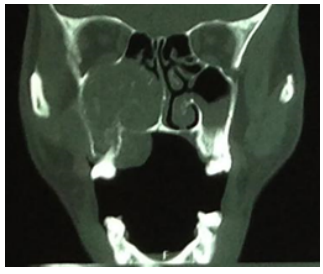
Incisional soft tissue biopsy specimen

FIGURE 4



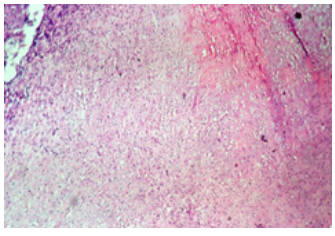
A mixed lesion seen i.r.t. periapical area of 11 to 17 in a Lateral occlusal view.

FIGURE 5



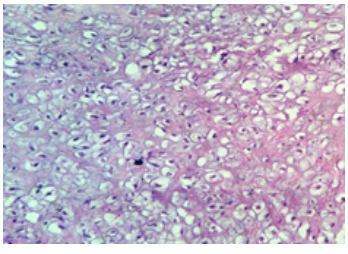
CT scan image showed a large, ill-defined, expansile, lytic lesion involving the right maxillary sinus with the inferio-medially destruction of maxillary alveolar arch and hard palate with extension of mass in oral cavity. Superiorly, erosion of floor of orbit and medially erosion and destruction of nasal turbinates.

FIGURE 6



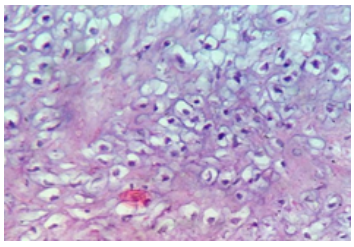
The H&E stained sections show the presence of a hyper-cellular connective tissue stroma.

FIGURE 7



The given H&E stained slide revealed abundant cartilaginous matrix containing round and ovoid cells in lacunae, exhibiting nuclear pleomorphism, atypical nuclei, and nuclear hyperchromatism.

FIGURE 8



The given H&E stained slide revealed a moderate grade mitotic activity with large plump chondroblasts and bi-nucleated chondrocytes seen in a few microscopic fields

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