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KEY WORDS:
Chondrosarcoma, maxilla, high grade, upper alveolus, CT scan.

RECURRENT CHONDROSARCOMA OF MAXILLA – CLINICOPATHOLOGY AND MANAGEMENT-A CASE REPORT

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ABSTRACT

Chondrosarcoma is a rare cartilaginous malignant tumor which occur extreme rarely in head and neck. We are presenting a case report of a recurrent Chondrosarcoma of upper alveolus in a 27 years old male patient presented at our institute with a large proliferative swelling arising from the hard palate. Punch biopsy from the growth was suggestive of high grade Chondrosarcoma. Imaging with MRI showed an altered signal intensity lesion involving hard palate and upper alveolus with erosion of floor and medial wall of left maxillary sinus. The patient underwent bilateral partial maxillectomy with stable post-operative course. Final histopathology was Chondrosarcoma poorly differentiated grade 3. We are hereby discussing the clinicopathological & radiological features of this case along with a review of the Chondrosarcoma.

INTRODUCTION

Chondrosarcoma is an uncommon malignant neoplasm originating from fully developed cartilaginous structure and devoid of osteoid. It is an extremely rare primary tumor of cranio-facial region (10%). The Chondrosarcoma of the craniofacial region may arise from any bone, cartilage or soft tissue structures[i]. This rare tumor has a diverse prognosis after surgical excision depending on grade. This tumors are radiation and chemotherapy resistant. We hereby present a case of recurrent Chondrosarcoma of maxilla. In contrast to osteosarcoma, Chondrosarcomas usually occur in adulthood.

Case report

A 27 year old young male presented to our out-patient department with a recurrent proliferative growth in the anterior part of palate more in left side, coming externally from oral cavity. Patient had a history of small polypoidal mass over palate 3 years back, for which excision biopsy was done and histopathology was suggestive of low grade chondroid tumor. Patient was asymptomatic since then until last 3 months when he developed the rapidly progressing proliferative lesion in the same site. The patient was also complaining loosening of tooth and occasional pain associated. There were neither any significant past history like radiation or family history nor any history of medical co-morbidities or past surgeries.

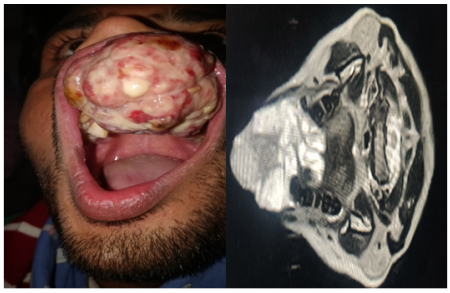

Figure 1: proliferative growth arising from hard palate

On examination there was a cauliflower like proliferating growth on the anterior aspect of hard palate from 1st premolar of left side to 1st premolar on right side. There was no associated trismus, synchronous other lesion, skin involvement or palpable neck nodes. Externally the growth is protruding outside, for which patient was unable to close his mouth completely.

On contrast enhanced CT scan the lesion was 90×55×42 mm with altered signal intensity in upper alveolus with erosion of floor and medial wall of maxillary sinus and abuts inferior turbinate. Magnetic resonance imaging (MRI) was done to asses soft tissue involvement. There were no involvement of posterior wall of maxilla and pterygoids and no cervical lymphadenopathy.

Trucut needle biopsy was done from the growth under local anesthesia. Biopsy revealed high grade Chondrosarcoma. A surgical resection was performed under general anesthesia with Weber Ferguson approach. The tumor was resected en block with bilateral inferior partial maxillectomy after making suitable boney cuts along bilateral hard palate, anterior maxillary wall and alveolus. The posterior part of hard palate was preserved. The procedure was uncomplicated without any need for blood transfusion. Neck dissection was not done in view of no lymphatic metastasis in neck. A temporary obturator was constructed to obturate the surgical defect.

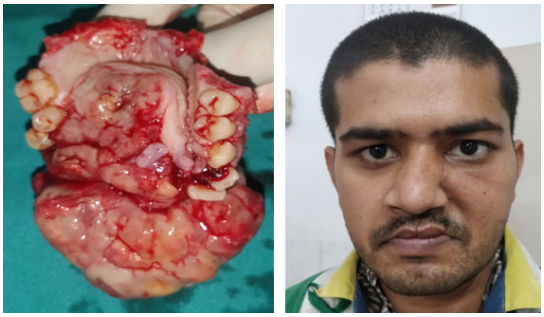

Figure 2: En-block specimen and post-op appearance

Postoperative recovery was uneventful with regular mouth opening exercise. Naso-gastric tube was removed 7 days after surgery and patient started oral diet without any oro-antral regurgitation and slight nasal intonation of voice. 6 months post surgery, a more suitable permanent obturator was inserted.

On histopathological examination, sections stained with hematoxylin-eosin showed many atypical chondrocytes in hyaline cartilaginous matrix in lobular patterns. There was hyper-cellularity, pleomorphism, irregular nuclei and myxoid degeneration with high rate of mitosis. It also had malignant chondroid material, stellate cells, necrotic foci and vascular proliferation. There was lymphatic permeation but no vascular or peri-neural invasion. Macroscopically the tumor was a size of 8×4×4 cm arising from hard palate and maxilla and surface mucosal ulceration with tumor free surgical margins and underlying bony involvement. Immunohistochemistry was positive for S-100. The final histopathological diagnosis was conventional Chondrosarcoma, poorly differentiated, grade-3.

Patients had no risk factor for adjuvant radiation and post op radiotherapy was not given. Patient is on regular close follow-up for 1 year with clinical examination and radiological evaluation 6 monthly. There is no clinic-radiological evidence of recurrence on 1 year follow up.

DISCUSSION

Chondrosarcoma is a malignant neoplasm of the cartilage. These are relatively uncommon but can occur anywhere in the body with a predilection for the long bones. They comprise about 10% of all primary tumors of skeleton. Chondrosarcoma of the head and neck region are very rare constituting 0.1% of all the head & neck neoplasms[8]. Head neck Chondrosarcomas occurs in young age with slight male predeliction[7]. The most common locations in head & neck are maxilla, base of skull, cervical vertebra, nasal cavity. The Chondrosarcoma arises from remnants of embryonic cartilage precursors from nose and septum in the anterior part of maxilla. The most common presenting symptom is a painless mass.

The principal diagnostic modalities are computed tomography (CT) and magnetic resonance imaging (MRI). CT is a good test to detect characteristic stippled to coarse calcifications[9] and bone erosion, expansion of the affected cartilage. MRI is optimal for delineating the soft tissue involvement, these show high signal intensity on T2 weighted images because of high water content in matrix.

The diagnosis of Chondrosarcoma is made histologically. Evans classified Chondrosarcoma into 3 histological grades according to their cellularity, number of mitosis per field and size of cell nuclei. This classification has prognostic implications. Differentiating a grade 1 Chondrosarcoma from enchondroma and Chondrosarcoma from chondroblastic osteosarcoma is difficult[3].

Grade 1 Chondrosarcomas (WHO- atypical cartilaginous tumor) are moderately cellular with abundant chondroid matrix, small round nuclei may be bi-nucleate and absence of mitosis. 10 year survival is 83-95%. Grade 2 are more cellular but with less chondroid matrix. Grade 3 Chondrosarcomas have abundant mitosis, nuclear pleomorphism and high cellularity with absent chondroid matrix. 10year survival is 29-55%.[5] Most Chondrosarcomas of head and neck region are grade 1, but in our case it was grade 3.

The treatment of choice is wide surgical resection with negative margins and preservation of function if possible. Incomplete excision is associated with a high rates of recurrence. Elective neck dissection is not recommended unless the neck is clinically or radiologically involved[4]. Role of radiotherapy in Chondrosarcoma is controversial. Typically Chondrosarcoma is considered radioresistant. Radiotherapy may be used in high grade tumor after surgery as adjuvant therapy or in unresectable tumors. Conventional Chondrosarcomas are chemotherapy resistant. But rare subtypes of Chondrosarcoma: mesenchymal and dedifferentiated subtypes because of their aggressive clinical course may be treated by chemotherapy[2].

The two most important prognostic factors are tumor grade and resectability. Other important prognostic factors are size and location. High grade tumors and histologically positive margins indicate poor prognosis. Maxillary tumors have poor prognosis compared to mandibular tumors because maxillary tumors are more difficult to eradicate. Local recurrence of tumor invading vital structures of head and neck can often lead to death. Overall the prognosis of a low grade chondrosarcoma treated with adequate surgical margins is good. These tumors show a wide variation in the recurrence and metastasis therefore life long follow-up is advised[6].

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