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CHONDROSARCOMA OF MAXILLA: A RARE CASE REPORT

Pathology

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ABSTRACT

Chondrosarcoma (CS) is an uncommon malignant cartilagenous tumor of maxilla. It is an aggressive neoplasm with high recurrence rate and may show distant metastasis. Histopathological diagnosis is still gold standard for diagnosis of CS. However, correlation with clinico-radiological findings is necessary. Here we report a case of maxillary chondrosarcoma in a 57 year old female patient with brief discussion of its clinical, radiological and histopathological findings along with its various treatment modalities.

KEYWORDS

Hyaline cartilage, Enchondroma, Chondrocytes, Malignant osteoid

INTRODUCTION:

Chondrosarcoma (CS) is rare cartilage forming tumor, less than 1% of CS affect head and neck region[1]. In craniofacial region they are slightly more common in male than female. Primarily occur in 3-6th decades of life [2]. Here we present a case of CS of maxilla in a 57 yr old female patient which brief discussion on its pathogenesis, clinical features, histopathology, immunohistochemistry and treatment protocol.

Case report:

A 57 year old female patient visited our tertiary cancer centre in January 2021, with the chief complaint of swelling in upper right cheek area since 3 months. Her general physical examination and family history were not significant. On clinical examination there was a swelling in right cheek region with deviation of right nasal septum towards opposite side. Overlying skin was normal in color and texture. Intraoral examination revealed a nodular growth in upper jaw extending anterio-posteriorly from 1st premolar to soft palate. Latero-medially it was extending from upper gingivobuccal sulcus to midline. The overlying mucosa was normal in color. On palpation, the swelling was tender, non compressible, non fluctuant and soft to firm in consistency. No evidence of lymphadenopathy identified.

CT scan showed an expansile lytic lesion involving posterior part of maxillay bone of size 6x5x5cm. There were few popcorn / cloud like radio-opacity suggestive of calcification. The lesion was involving palatal bone, alveolar process, zygomatic bone and maxillary sinus along with medial deviation of nasal septum. Superiorly, the lesion was bulging the orbital floor however infiltration was not evident (Figure1). Considering the above findings, malignant salivary gland neoplasm, squamous cell carcinoma, osteosarcoma, chondrosarcoma and lymphoma were considered in differential diagnosis.

Right complete maxillectomy along with partial left maxillectomy was done. Histopathological examination revealed, multiple sheets and lobules of mature hyaline cartilage seen, separated by fibro-collagenous stroma. The lobules comprised of disorganized crowded lacunae containing uninucleated and binucleated chondrocytes. Nuclear pleomorphisms, atypical mitosis with conspicuous nucleoli were seen. At periphery, the lobules were merging into spindeloid to ovoid atypical cells, devoid of cartilage differentiation. The tumor has infiltrating interface and permeating into bony trabaculae. At places partial mineralizasion and ossification were noted. No malignant osteoid formation was seen. No perineural invasion/lymphovascular invasion was noted [Figure 2 (a&b)]. Considering the overall findings, final diagnosis of chondrosarcoma grade II was given.

Figure1: Computed tomography shows a radiolucent, expansile, lytic lesion with central radio-opacity involving maxillary sinus.

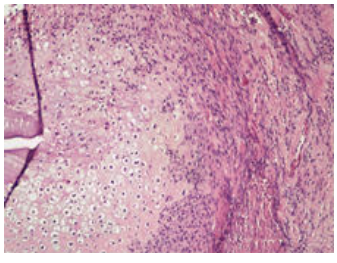

Figure 2(a): H&E stained photomicrograph showing mature hyaline cartilage with pleomorphic chondrocytes. At periphery it is merging with spindeloid cells. (4x)

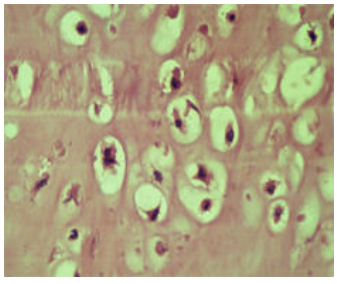

Figure 2(b): H&E stained photomicrograph showing lacunae containing uninucleated and binucleated chondrocytes. (10x)

DISCUSSION:

CS is a malignant tumor which shows diverse morphologic features and clinical behavior. 1-4 % cases of CS occur in head and neck region. Maxilla is more commonly affected than mandible with a ratio of 1.75: 1 [3].The most common symptoms are swelling, nasal obstruction, epistaxis and tooth mobility [4].

Radiologically, they appear as a radiolucent lytic bony lesion with few foci of radio-opacity [5]. Histopathologically, Evan's etal in 1977 graded CS in grade I to III, according to their degree of cellularity, atypia, mitotic activity, nuclear size and surrounding matrix composition [6]. In our case, considering the histological appearance we put it under intermediated grade (grade II).Till date there is no evidence based treatment protocol for CS of head and neck because of its rarerity.[2] Surgical excision is the treatment of choice. Local curettage, cryotherapy, chemotherapy, radiotherapy and immunotherapy are considered as other treatment options[7]. According to WHO, 5 yr survival for grade I lesion is 89% and 53% for grade II and grade III. 20-60% of cases show local recurrence. It can recur from few months to several years. Approximately 20 % cases metastasize to lungs.[3]

CONCLUSION:

Chondrosarcoma in maxilla is a rare entity. A thorough clinical and radiological investigation along with histopathology is required to reach the final diagnosis. Lifelong follow up is mandatory since it shows high recurrence rate.

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