Case Report:

Mesenchymal chondrosarcoma of maxilla- a rare case report

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Abstract:

Mesenchymal chondrosarcoma(MC) is a rare histological variant of chondrosarcoma(CS) that can arise from both bone and soft tissue. It is usually seen in younger age group compared to conventional CS. MC reveals unusual clinical behavior, characteristic histopathological features, and poor prognosis with late recurrences. This report describes a unique case of MC in a 52year old female who presented with swelling and pain over left side of the face. CT scan showed an expansile lytic lesion in the left maxillary sinus. Incisional biopsy confirmed the diagnosis of maxillary MC. The patient underwent left total maxillectomy with postoperative chemo and radiotherapy.

Key-words: Chondrosarcoma, Maxilla, Mesenchymal chondrosarcoma

Introduction:

Mesenchymal chondrosarcoma(MC) is one of the most unusual neoplasms, first described by Bernstein and Lichtenstein in 1959 as a distinct variant of chondrosarcoma (CS).[1] MC can arise from soft tissue/ bone in the ratio of 1:2 to1:6. This tumor has a female preponderance with male: female ratio of 1:4.[3] MC is more common in second and third decade of life.[4] Facial bones and ribs are the commonest bones to be involved in this disease.[2] MC is characterized by sheets or clusters of highly undifferentiated small ovoid cells that are intermixed with small zones of neoplastic cartilage.[2]MC usually present as painless swellings. Pain may be a late stage feature, and regional lymphadenopathy is very rare.[5] These are usually aggressive neoplasms with a high tendency for late recurrence and distant metastases.[6]The review of literature shows that a very few cases of maxillary MC have been reported so far in the English literature. Hence we report and discuss this infrequent case of mesenchymal chondrosarcoma of the maxilla in a 52 year old female patient.

Case History:

A 52year old female reported to the department of ENT with a complaint of swelling and pain over left side of the face since 1month. The patient was prescribed antibiotics but she returned after 1week with proptosis of left eye. On examination, there was facial asymmetry (due to diffuse swelling in the left middle 1/3rd of the face) with slight obliteration of nasolabial fold. (Figure 1)

Skin over the swelling was normal. There was no local rise of temperature. On palpation, it was firm to hard in consistency. Intraorally, no swelling was noted. CT scan (Plain and I.V contrast study) showed a 3.5x3.3cm mass noted in the left maxillary sinus which is isodense on plain CT with few hyperdense areas. The mass shows heterogeneously intense enhancement with contrast. The mass was seen

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eroding the floor of the orbit and extending superiorly and located extraconally just abutting the inferior rectus muscle and pushing it upwards. The anterolateral, posterolateral, medial walls of maxilla, anterior portions of all turbinates were eroded by the mass. (Figure 2)

An incisional biopsy was done from the eroded left lateral wall of the nose in the middle meatal area, which showed a dimorphic pattern composed of highly undifferentiated small round cell component arranged in hemangiopericytomatous pattern, intermixed with chondroid islands. (Figure 3)The patient underwent left total maxillectomy and the excisional biopsy report concluded to be a mesenchymal chondrosarcoma. Later the patient was treated with chemo and radiotherapy.

Discussion:

Mesenchymal Chondrosarcomas(MC) are infrequently reported in the literature due to their rare incidence. They constitute only 1% of all the chondrosarcomas(CS). [7]Both skeletal and extraskeletal lesions have been reported in the head and neck region.[2] When these tumors arise in the head and neck region, they appear to have a predilection for the maxillofacial skeleton.[7] Only 45 cases of MCs of jaws have been reported in literature till now, 23 in maxilla and 22 in mandible. We are reporting the 24th case of MC in maxilla. [8]

They are usually misdiagnosed as odontogenic fibroma, chondromyxoid fibroma, fibrosarcoma and angiosarcoma on incisional biopsies. MCs show no specific clinical signs and symptoms. The predominant symptom was usually a painless mass or swelling (53%). However, painful mass (16%) as in the present case has also been reported. Few patients developed neurological disturbances such as facial paresthesia and lip pareses.[9]CSs of the head and neck occurs most frequently in the third to sixth decade of the life as compared to MC, which usually occurs in second to third decade of life.[8]In contrast, the present case was reported in the sixth decade.

The most common radiographic appearance of MC of jaw is radiolucent shadow, which makes its differentiation difficult from the rest of sarcomas.[2]In the present case there was a wide radiolucent osteolytic shadow in the left maxilla with randomly scattered opaque mottled areas.The histopathology of MC is distinct regardless of the site of origin. They are characterised by a biphasic pattern, with a sheet-like or hemangiopericytoma like proliferation of small undifferentiated spindle or round cells surrounding islands of chondroid differentiation.[10] No mention has been made in the literature on enzyme histochemistry of MCs. Immunohistochemistry is advised for difficult cases. Chondroid areas are positive for S-100 protein, and neuron-specific enolase is focally positive for primitive mesenchymal cells.[10]The most effective therapeutic modality is wide surgical excision.[11] Postoperative radiotherapy and chemotherapy offer a good prognosis and eradicate micrometastases that have not been previously detected.[3]These tumors tend to recur locally after resection and metastasize even years after surgery. The most common site of distant metastasis is the lung.[12] Hence, even after complete surgical resections with negative tumor margins, a long term follow up is essential for chondrosarcomas.

Conclusion:

MC is a very rare tumor reported in the maxilla. Adequate biopsy is enough for making the diagnosis. MC requires surgical excision with wide margins. Postoperative chemotherapy or radiation may be a

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choice of treatment. They have high potential for late local recurrence and distant metastases.

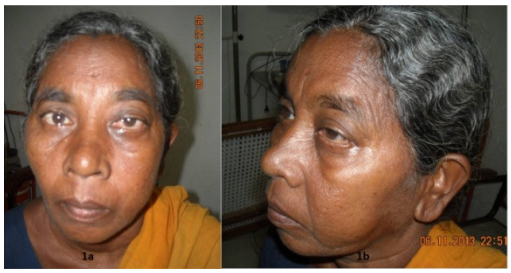

Figure 1a: Diffuse swelling in the left middle 1/3rd of the face with slight obliteration of nasolabial fold.
1b.Note the proptosis of left eye

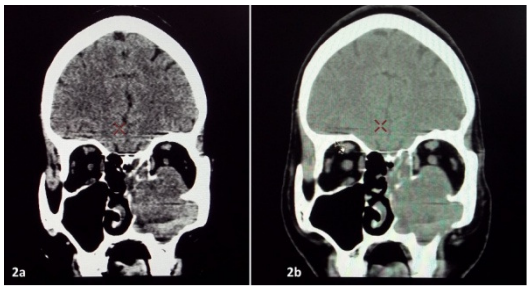

Figure 2a. Plain CT showing radiolucent isodense mass with few hyperdense areas in the left maxillary sinus
Figure 2b. Contrast CT showing heterogenous intense enhancement of the mass

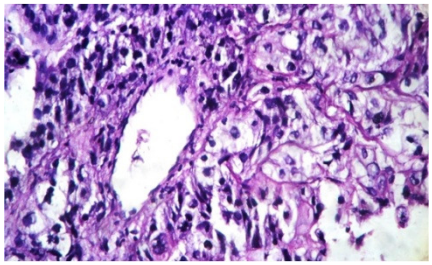

Figure 3: Biopsy showing dimorphic pattern composed of highly undifferentiated small round cell component arranged in hemangiopericytomatous pattern, intermixed with chondroid islands.(H&E,400x)

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