MESENCHYMAL CHONDROSARCOMA OF MANDIBLE - A RARE CASE STUDY AND LITERATURE REVIEW
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Abstract: A case report of 36 year old gentleman with no comorbid presented with swelling in left side of face for 2 months. Clinically, ulceroproliferative lesion of size 14 x 6 cm in left side of mandible and maxilla with marked facial asymmetry and left mandibular nerve paraesthesia. Computed Tomography of local part revealed large irregular heterodense mass obliterating oral aperture with bony destruction of mandible and ipsilateral maxilla, hard palate and soft palate submucosally. Histologically, tumour shows characteristic bimorphic appearance of small primitive appearing round cells with focal cartilaginous differentiation with immunohistochemical correlation suggestive of Mesenchymal Chondrosarcoma. Since lesion was inoperable, planned for Radiation therapy and single agent Doxorubicin. He developed grade IV neutropenia following first cycle of doxorubicin. Hence, further chemotherapy deferred. He received total dose 60 Gy of radiation. He presented a month later with paraplegia. MRI spine revealed an extradural mass compressing spinal cord at D2 vertebra. He received palliative radiation of 30 Gy. However, he succumbed to disease within a month. Mesenchymal chondrosarcoma is a rare tumour with aggressive course and rapid progression as seen in this case. 

Keyword : Mesenchymal chondrosarcoma, mandible, bimorphic histologic pattern

INTRODUCTION:
Mesenchymal chondrosarcoma is a rare neoplasm involving bone and soft tissues. It corresponds to 1% of all chondrosarcomas(1). In 1959, it was first described by Bernstein and Lichtenstein as a distinct variant of chondrosarcoma (CS)(2). It usually appears in the second and third decades of life (3). This neoplasm affects females more frequently than males (F/M = 1.4/1)(4). They can occur in any part of craniofacial compartment where cartilage is found like maxillofacial skeleton, skull base, and nasopharynx(5,6,7).

These neoplasms are characterized by bimorphic histologic pattern a) sheets or clusters of highly undifferentiated, small, round to ovoid cells, b) zones of neoplastic hyaline cartilage. This type of neoplasm shows aggressive local behavior as well as a high metastatic potential. Due to these features and the high risk of recurrence, the prognosis is poor (4). Many investigators believe that these tumours have their origin from cartilage forming mesenchyme and hence the name Mesenchymal Chondrosarcoma.

CASE REPORT:
A 36-year-old man with no comorbid presents with rapidly growing foul smelling, necrotic, ulceroproliferative lesion, measuring 14 x 6 cm on the left side of the face for 2 months duration. On examination a large foul-smelling necrotic ulceroproliferative lesion occupying the left half of the face. Clinically, mandible appears to be infiltrated. The lesion protrudes through the oral cavity and he presented with difficulty in closing his oral cavity. He presented with marked facial asymmetry (Figure - 1).

Figure 1 : Local extent of disease before treatment
There was left mandibular nerve paresthesia with numbness over left half of the face. There was no significant cervical lymphadenopathy. Introraaly, the lesion appeared as large, lobulated lesion with an ulcerated surface and the lesion occupied the entire oral cavity.

Figure 2 : Computed Tomography showing tumour extent
Computed tomography (CT) scans of local part depicted a large irregular, heterodense lesion obliterating the oral aperture. There is obvious bony destruction of the mandibular alveolus with infiltration to the ipsilateral maxilla, the hard palate, and also infiltrating the soft palate submucosally, as it is evident by a bulge in the left lateral wall of the oropharynx [Figure 2]. Serum alkaline phosphatase, chest CT, and bone marrow aspirate were normal.

Figure 3 : Characteristic bimorphic histological pattern with well differentiated neoplastic chondrocytes above and small primitive appearing round cells below (A) High power (B) Low power
Histological evaluation revealed a characteristic bimorphic histological appearance, with areas of well-differentiated atypical chondrocytes along with abnormal small primitive appearing round cells with scanty cytoplasm and round to oval hyperchromatic nuclei. Mitotic activity was increased and varied from 1-3 per high power field. Chondromyxoid areas were also seen [Figure 3].

Figure 4: Immunohistochemistry (A) Showing vimentin positivity by primitive-appearing small round cells, (B) Showing NSE positivity by primitive-appearing small round cells, (C) Showing CD 99 positivity by primitive-appearing small round cells, (D) Showing S 100 positivity with neoplastic chondrocytes Immunohistochemically, the cartilaginous component demonstrated positive with S100, and round cells positive for vimentin, CD99, and neuron-specific enolase [Figure 4]. The histological features were suggestive of MC. The lesion was deemed inoperable in view of its large size, its rapid progression and presence of extensive skin and soft tissue involvement. He was hence planned for radiotherapy and single agent doxorubicin chemotherapy. Patient underwent elective tracheostomy prior to starting treatment and was put on ryles tube to maintain adequate nutrition. Patient developed Grade IV afebrile tracheostomy prior to starting treatment and was put on ryles tube agent doxorubicin chemotherapy. Patient underwent elective tracheostomy prior to starting treatment and was put on ryles tube to maintain adequate nutrition. Patient developed Grade IV afebrile neutropenia following first cycle doxorubicin and hence further chemotherapy not contemplated.

Figure 5: Clinical response after radiation therapy. He was treated with external beam radiation to locoregional site to a total dose of 60 Gy using cobalt beam therapy. Patient tolerated radiation well, except for grade I – II oral mucositis. Partial response achieved (Figure 5). Patient discharged with advice to review after 6 weeks.

Figure 6: MRI spine showing extrudal mass at D2 vertebra. Unfortunately, he presented with paraplegia of two days onset, 4 weeks after completion of treatment. Magnetic Resonance Imaging of the spine revealed an extrudal mass at the level of D2 vertebra compressing the spinal cord [Figure 6]. He was then treated with palliative external beam radiation using 8 MV X-ray beam therapy to D2 spine to a total dose of 30 Gy. His general condition continued to deteriorate and he succumbed to disease within a month.

DISCUSSION:
Mesenchymal Chondrosarcomas can occur in both osseous and extra-osseous sites in a ratio of 2:1. Among skeletal Mesenchymal Chondrosarcomas 3 to 25% occur in the maxillofacial region (8). Most common sites are maxilla and mandible. Extraskeletal tumors arise from the orbit, meninges, nasal, paranasal mucoza, and the parapharyngeal space (9). The most common location in the maxilla is in the premolar and molar area and for mandible is in premolar-molar area but the symptoms Coronoid and condylar processes may also be involved (10). Commonly present with a painless mass or swelling (53%) although a painful mass is also (16%) reported (11). Bleeding from the lesion, nasal obstruction, epistaxis, paraesthesia, facial nerve palsy, hard of hearing are also observed. Dental complaints may be the initial symptoms. Injudicious dental extractions or biopsy can provoke rapid growth (4).
Routine blood investigations including liver, renal function test with bone biochemical analyses are performed for preoperative assessment and for evaluations of distant metastasis. The typical appearance of a cartilaginous lesion on plain radiographs is discrete calcification and they may be radiolucent on radiographs, which may show punctuate or punctate calcifications. They vary depending on the amount of mineralization. CT of local part helps in visualization of tumour extent, bony destruction (12). MRI of local part is the investigation of choice for assessing the exact extent of lesion, the extent of soft-tissue involvement, making it an important tool for preoperative planning. MRI also helps in diagnosing recurrence at a surgically treated site. Bone scan, CT chest, CT abdomen are used for systemic staging of the tumor before surgical treatment. Histologically, characteristic bimorphic appearance with areas of well differentiated cartilage along with neoplastic abnormal chondrocytes and small round cells with scanty cytoplasm and round to ovoid hyperchromatic nucleus. Immunohistochemically, the small round cells are positive for Vimentin, CD 99, Neuron specific enolase and the neoplastic chondrocytes are positive for S-100. In general, prognosis for Mesenchymal Chondrosarcoma is poor because they have high tendency for both local and distant spread (13). Metastasis is hematogenous and the most common site is the lung. Five-year survival rates for craniofacial tumours are 40%-60%, and at least 60% of patients have recurrences within 5 years of initial treatment (10). Takahashi et al. reported that 6 out of 14 patients with mandible tumors died after an average survival time of 28 months and that survival time is longer with maxillary than with mandibular lesions (12). The most effective treatment modality is wide surgical excision (13). Mandible-wide local excision with a tumor free margin of 2 to 3 cm is recommended (7,10). According to Nakashima et al. (5), extensive resection has less recurrence and a better survival rate than limited surgical excision (2). Although there have been reports of resolution of this tumor with chemotherapy and radiation alone (3), the benefit from chemotherapy and radiation is as yet unclear (7). However, experience of these methods is limited and there is no evidence that these therapies improve the prognosis (14). However, postoperative radiotherapy and chemotherapy offer a good prognosis and eradicate micrometastases that have not been previously detected.

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