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A RARE CASE OF MESENCHYMAL CHONDROSARCOMA OF THE MANDIBLE MIMICKING A SOFT TISSUE SARCOMA

Oncology

Dr R. Balasubramaniam* M.S. Mch., Chief, Dept of Surgical Oncology, Madras Medical College and Rajiv Gandhi Government General Hospital, Chennai *Corresponding Author

Dr S. Pradeep MDS, Fellow in Head and Neck Oncosurgery, Special trainee, Dept of Surgical Oncology Madras Medical College and Rajiv Gandhi Government General Hospital, Chennai

Dr. Kalpa Pandya MDS, Fellow in Head and Neck Oncosurgery, Special trainee, Dept of Surgical Oncology Madras Medical College and Rajiv Gandhi Government General Hospital, Chennai

ABSTRACT

Chondrosarcomas account for 0.1 % of all head and neck malignancies. Mesenchymal chondrosarcoma is a malignant tumour known to have a biphasic histopathology consisting of undifferentiated small cells and areas of cartilage formation. It is known to be highly aggressive and is associated with poor prognosis. In head neck region, maxilla is the most common bone affected, followed by mandibular body, ramus, nasal septum and paranasal sinuses We describe a rare case of mesenchymal chondrosarcoma of the mandible mimicking a soft tissue sarcoma.

KEYWORDS

chondrosarcoma, mesenchymal chondrosarcoma, mandible

INTRODUCTION

Chondrosarcoma are commonly found to be present in flat and long bones. They account for 10-20 % of all bone tumours and 0.1 % of all head and neck malignancies.(1,2) Mesenchymal Chondrosarcoma (MS) was first described by Lightenstein and Bernstein in 1959 as a high grade rare type of chondrosarcoma.(3) It is a malignant tumour known to have a biphasic histopathology consisting of undifferentiated small cells and areas of cartilage formation. It is known to be highly aggressive and is associated with poor prognosis.(4)

We describe a rare case of mesenchymal chondrosarcoma of the mandible mimicking a soft tissue sarcoma.

CASE REPORT

A 45 years old patient came with the chief complaint of extensive swelling with respect to right side of jaw, since 4 months. The patient reported a rapid increase in size of the swelling. On examination, patient presented with a swelling approximately 7 X 6 cm in size on the right cheek, with small ulcerated area approx. 5 mm in size. Intraoral, we noticed an ulceroproliferative growth approx 7 X 6 cm in size extending from 31 to 48. Bone expansion with intact mucosa could be seen in the region from 31 to 33. (Fig 1)

Fig 1: Patient with an extensive extraoral swelling with reduced mouth opening making intraoral examination difficult

Patient presented an enlarged node with reference to level IB region. The biopsy was reported as Ameloblastoma. Contrast Enhanced CT scan showed heterogeneous lesion with reference to right mandible with breach of the buccal cortex. Lesion extended from left canine up to posterior region of the body of the mandible, going superiorly up to the condyle of the mandible. The temporomandibular joint space appeared free. (Fig 2)

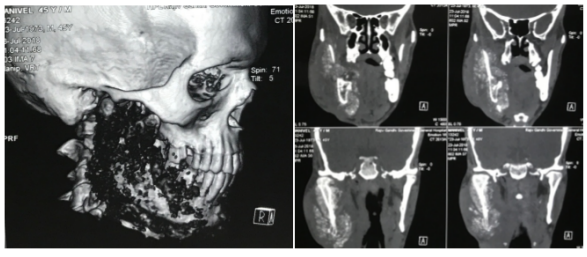

Fig 2: Contrast Enhanced CT scan showed heterogeneous lesion with reference to right mandible with breach of the buccal cortex. Lesion extended from left canine up to posterior region of the body of the mandible, going superiorly up to the condyle of the mandible. The temporomandibular joint space appeared free

Patient was taken up for composite resection and reconstruction with a pectoralis major myocutaneous flap.The lesion was excised in a monobloc fashion along with a hemimandibulectomy. The defect was reconstructed with a pectoralis muscle myocutaneous flap. (Fig 3)

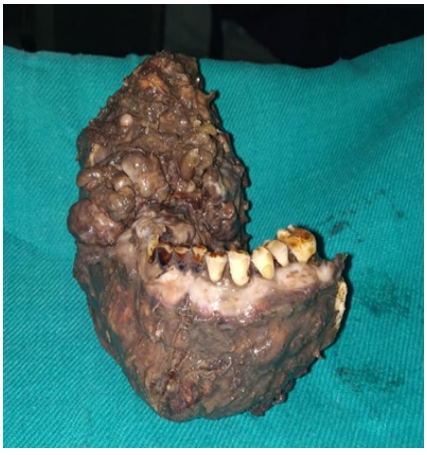

Fig 3 The lesion was excised in a monobloc fashion along with a hemimandibulectomy

Histopathology was reported as Mesenchymal Chondrosarcoma. Microscopy revealed neoplasm composed of sheets for undifferentiated round to oval cells with elongated hyperchromatic nuclei and prominent nucleoli. Cartilaginous differentiation was also seen with extensive calcification and new bone formation. Immunohistochemistry study was reported as negative for p63, EMA and s100.

DISCUSSION AND REVIEW OF LITERATURE

Chondrosarcomas have several variants which include clear cell, dedifferentiated, myxoid and mesenchymal. Mesenchymal chondrosarcoma comprise of only 1 % of the chondrosarcoma and is the most aggressive.(5) This tumour is more commonly found in the 2nd and 3rd decades of life unlike other variants which are more commonly seen in the older age group. Our case reported falls in the fifth decade of life which is slightly unusual. There are varied observations regarding gender predilection, with some authors reporting equal predilection(4) while some showing female preponderance.(2)

Two thirds of the MS arise in the bone while the rest arise extraosseously. Chondrosarcoma are thought to be derived either from the reserve cells from cartilages or from mesenchymal cells displaying chondroblastic differentiation, (6) In cytogenetic studies, no specific translocations are found in MS. However Nauman described a translocation of chromosome 13 and 21 in some cases of both osseous and extra-osseous MS suggesting a similarity between both.(7) Sainati described a translocation between chromosome 11 and 22.(8)

In head neck region, maxilla is the most common bone affected, followed by mandibular body, ramus, nasal septum and paranasal sinuses.(9) Our case showed the tumour involving symphysis, parasymphysis, body, ramus and condyle of the mandible.

MS in maxillofacial region present early compared to other sites due to a more apparent location. They present with facial swellings, nasal blockage, bone expansion, disturbance in functions such as swallowing, speech and breathing. MC arising from bone tend to first expand and then erode the bone once it reaches the cortex.(10) Generally in oral cavity, they present as smooth swelling.(11) However they also have a tendency to invade structures and are known to be vascular. In our case, at the patient presented to us with an extensive fast growing ulcer proliferative mass, due to which we suspected it to be a soft tissue sarcoma. Also it was a highly vascular and did cause extensive bleeding at the time of tissue biopsy.

Histological MS consists of a biphasic pattern comprising of solid sheets of round cells and chondroid matrix.(10) However, sometimes deficient chondroid matrix in the sample makes it difficult to identify it as chondrosarocma. Molecular biology of the tumour is being recently studied under great detail with a view of distinguishing it from other soft tissue sarcoma. Type II collagen has been found to a useful marker to distinguish it from small cell sarcomas like Ewings sarcoma and synovial sarcomas. Also s-100 which is consistent marker in other chondrosarcoma is negative in MS.(12)

The accepted treatment for chondrosarcoma is surgical modality involving en bloc excision of the tumour with wide margins. Pseudopods projecting from the tumour make it mandatory to take adequate margins. Some authors have reported the use of pre-op radiotherapy to decrease the bulk of tumours which are inoperable at the time of presentation.(10) In a systematic review, Xu et al concluded that surgery is mandatory in case of MS. Adjuvant treatment in the form of radiotherapy and chemotherapy may decrease local recurrence but may net reduce the overall survival. They have also recommended that post operative radiotherapy may improve local control in cases of positive margins.(4)

The prognosis of patients with MS with is poor compared to other chondrosarcoma. However in case of head and neck, the prognosis is relatively better than other sites. The 5 and 10 year survival rates f MS in general have been reported as 55% and 27%.(5) However, in case of head and neck, Vencio et al have reported 5 and 10 year survival rates of 64% and 55%.(2)

CONCLUSION

Thus mesenchymal chondrosarcoma is a rare aggressive tumour which warrants early detection and prompt management. Our reported case is being planned for adjuvant treatment and a close follow up on account of the aggressive nature of the disease.

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