Dedifferentiated chondrosarcoma of the maxilla

ABSTRACT

A 20-year-old male reported with right-sided facial swelling, epistaxis and right sided proptosis of two months duration. Computerized tomography scan of the face revealed a mass in the right maxillary sinus. The patient underwent surgery and the postoperative histopathology was suggestive of dedifferentiated chondrosarcoma. The present report discusses this rare presentation and the issues in its management.

KEY WORDS: Chondrosarcoma, dedifferentiated, maxilla

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INTRODUCTION

Chondrosarcoma is a bone tumour that usually arises in peripheral long and flat bones. These tumors have a high propensity for local recurrence. Occasionally a chondrosarcoma undergoes dedifferentiation and then assumes a very aggressive course. It is important to recognize dedifferentiation early, since it can be an indicator for more aggressive treatment of the tumor. We are discussing a 20-year-old male who presented with facial swelling and was diagnosed as dedifferentiated maxillary chondrosarcoma. This pathology has been reported only once in the maxillary area in literature.

CASE REPORT

A 20-year-old male presented with complaints of right-sided facial swelling, epistaxis and right-sided proptosis of two months duration. On examination there was a gross bulge on the right side of the face. The mass was filling the right-sided nasal cavity as well. CECT of the face revealed a heterogeneously enhancing mass in right maxillary sinus with extension to the right nasal cavity orbit and ethmoid and sphenoid sinuses [Figure 1].

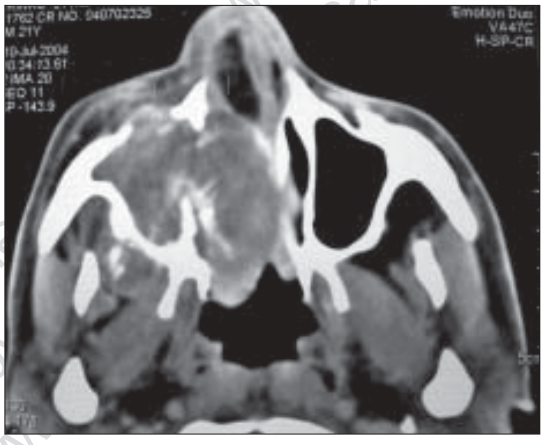

Figure 1: Axial CT showing the mass in the maxillary cavity

The patient underwent radical maxillectomy. Histopathology was suggestive of a 12 x 9 x 5 cm dedifferentiated chondrosarcoma, Grade III [Figures 2 and 3]. The medial resected margin of the specimen was involved by tumor while the rest of the margins were free. The patient received local radiotherapy to the face to a dose of 60 Gray in 30 fractions over six weeks and this was followed by six cycles of chemotherapy with ifosfamide, carboplatin and adriamycin. The patient was clinically controlled and was disease free six months after completing the adjuvant treatment.

DISCUSSION

Chondrosarcoma accounts for 10-20% of malignant primary bone tumors. Occurring predominantly in younger patients, these tumors are locally aggressive with high propensity for local recurrence. Chondrosarcomas usually arise in peripheral long and flat bones, most commonly involving the femur and having a predilection for secondary ossification centers. Secondary chondrosarcomas are most prevelant in the ilium.

In 1971, Dahlin and Beabout first fully described the concept of dedifferentiated chondrosarcoma.[1] Dedifferentiated chondrosarcoma is a highly

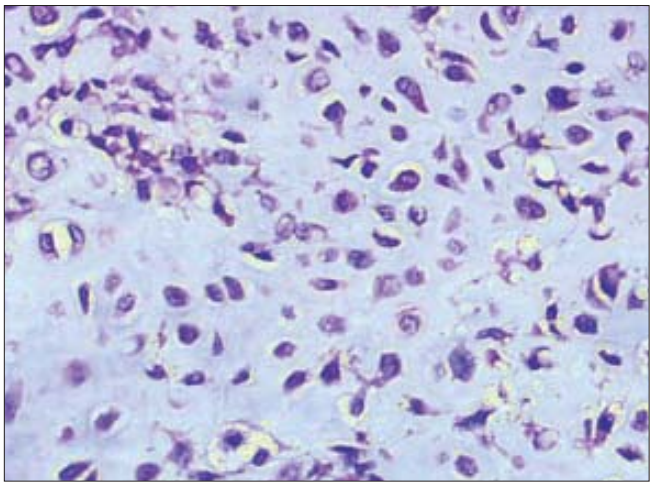

Figure 2: High power view of the cartilaginous component with features of high-grade chondrosarcoma (H and E, x400)

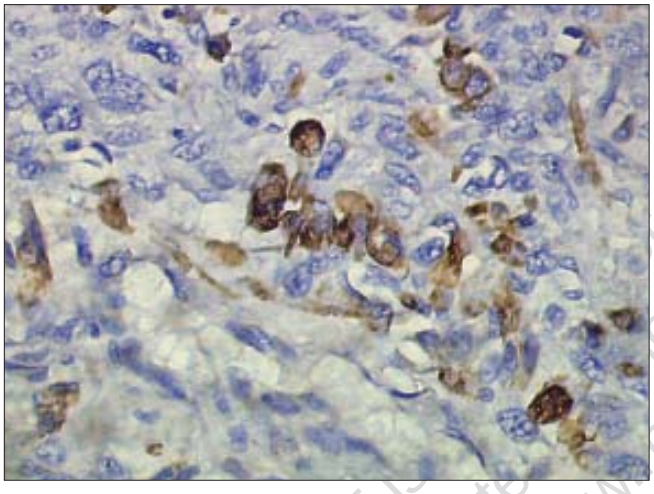

Figure 3: Immunohistochemistry for desmin was positive in cells with dense eosinophilic cytoplasm (IHC, x400)

malignant variant of chondrosarcoma. Approximately 11 per cent of chondrosarcomas can be expected to dedifferentiate into more anaplastic lesions. From a pathologist's perspective, multiple sections of a low-grade chondrosarcoma should be seen to avoid overlooking an anaplastic zone. Usually, however, the area of dedifferentiation is gross. The presence of two tumour portions of different mesenchymal differentiation lineages in these neoplasms gives rise to questions that do not have any definitive answer at present. Whether and how this tumour splits up is unanswered at this stage. There is probably a common monoclonal origin of both tumor portions.[2] The importance of recognizing dedifferentiation as early as possible lies in resecting with a wider or more radical margin to prevent local recurrence.[3]

Dedifferentiation is not peculiar only to chondrosarcomas. It has been reported in other tumors such as giant cell tumours, parosteal osteosarcomas and adult astrocytomas. The cause of dedifferentiation is largely unclear. In one of the largest series, Frassica *et al* analyzed the clinicopathological features and treatment of seventy-eight lesions of dedifferentiated chondrosarcomas.[2] The ages of the patients ranged from nineteen to eighty-two years. Eleven of the lesions developed in the site of a previously resected low-grade chondrosarcoma. Dedifferentiation was from low-grade chondrosarcoma to osteosarcoma in forty-two patients, to fibrosarcoma in thirty-three and to malignant fibrous histiocytoma in three. Widespread metastatic disease within two years after resection was a frequent finding. The over-all five-year-survival rate was 10.5 per cent. None of these patients had maxilla as a primary site.

Bruns *et al* reported 13 patients with extremity dedifferentiated chondrosarcomas with a mean age of the patients at diagnosis was 59.8 years.[4] In 11/13 cases, surgery was performed. Mostly, limb salvage with tumor resection and implantation of a megaprosthesis was done. Adjuvant chemotherapy was given to five patients. Recurrence was detected in 5/11 of the patients At follow-up 11 patients were dead of the disease, one dead of unknown reason and one alive with disease. The mean survival time was 9.7 months. Similar finding were reported for extremity chondrosarcoma by Kalil *et al*.[5]

The only case of dedifferentiated chondrosarcoma of the maxilla has been reported by Paume *et al*.[6] They discussed a 19-year-old girl who presented with a fascial swelling. The recurring tumor, diagnosed initially from the provoked pain, appeared on the scan image as a heterogeneous mass containing numerous calcifications. Histology showed, within a well-differentiated chondrosarcoma, an anaplastic sarcomatous zone of the malignant fibrous histiocytoma type. The details of the treatment given and the outcome of the treatment, was however not reported.

The management of the patient of dedifferentiated chondrosarcoma begins with a detailed preoperative staging of the case. The principal treatment of the condition is surgery. Surgical resection of the lesion with wide or radical margins should be attempted wherever possible. Adjuvant radiotherapy and chemotherapy should be administered in appropriate cases. The role of the adjuvant treatment is not very clear since there was no difference in survival rates of patients with or without adjuvant therapy two separate series.[3,7] Frassicat *et al* reported 5% overall five year survival rates with surgery and RT, 22% with surgery and chemotherapy and 10% with surgery alone. However surgery and chemotherapy arm had only nine patients and so the authors did not recommend routing chemotherapy use in these tumors.[3] Forty-two patients with dedifferentiated chondrosarcoma were reported by Dickey *et al*. Twenty-seven patients received neoadjuvant therapy; of these, twenty-three received chemotherapy only, two received radiotherapy only and two received combined therapy. The median survival time was 7.5 months and the five-year rate of disease-free survival was 7.1%. With the numbers available, there was no significant difference in the rate of disease-free survival with respect to the use of chemotherapy ($P = 0.54$).

To summarize, dedifferentiated chondrosarcomas have a poor overall prognosis but it is prudent to perform a wide excision with clear margins. The role of adjuvant chemotherapy is unclear in these cases. Our patient was disease free after six months but it would be inappropriate to put a treatment guideline since we are reporting only the second case of maxillary chondrosarcoma in world literature.

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