

An Extensive Sinonasal Osteosarcoma Mimicking Chondrosarcoma

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SUMMARY

Osteosarcoma is a rare tumour in the sinonasal region. Early diagnosis is essential for adequate management and better prognosis and this requires a meticulous histopathological examination. Reported is a case of osteosarcoma misdiagnosed as chondrosarcoma and treated by surgery followed by radiotherapy. However, appropriate diagnosis and pre-operative chemotherapy would have been significant in the prognosis. The pitfall of accurate diagnosis and the subsequent treatment is discussed in order to find the ways to maximize five years survival which is not more than 25% in this type of lesions.

KEY WORDS:

Sinonasal, Osteosarcoma, Chondrosarcoma, Diagnosis, Management

INTRODUCTION

Osteosarcoma of the sinonasal region is comparatively rare and accounts for about 6.5% of all osteosarcomas¹. The diagnosis depends upon the identification of woven bone on histopathological examination. It is an aggressive tumor, prone to both local and distant failure². Five years survival is not more than 25% which may be improved to 60% when a chemotherapy is initiated³.

Several epidemiologic risk factors have been related to the development of osteosarcoma, including a history of ionizing radiation exposure, fibrous dysplasia, retinoblastoma or prior exposure to thorium oxide, a radioactive scanning agent. Four percent of all patients with osteosarcoma had a history of previous radiation therapy for other tumours or conditions².

Osteogenic sarcoma is a non-epithelial malignant tumor which closely mimics chondrosarcoma in clinical presentation and on histological examination in the sinonasal region. Approach to treatment slightly varied in these two tumours. In osteosarcoma it certainly improves the long term prognosis if radical surgery is combined with chemotherapy preoperatively or as adjuvant³. In contrast, surgery followed by radiotherapy is the treatment of choice in chondrosarcoma³. The problem arises when there is a predominant chondroblastic element which may be misdiagnosed as chondrosarcoma.

CASE REPORT

A 31 years old male Malay teacher presented with a progressively enlarging mass of six months duration associated with dysphagia and deterioration in vision for two months. Examination showed a mass on the right cheek extending from the lower eye lid to the mandible. The mass was non-tender and firm. Another mass was seen protruded from the mouth measured eight by ten cm in size (Fig. 1). The mass occupied the whole oral cavity and has a raw surface which was firm in consistency and foul smelling. Computed tomographic scan showed a fungating mass arising from the right anterior wall of maxillary sinus, extending into the nasal cavity, ethmoid sinus, orbital cavity and infratemporal fossa. The anterior wall of the right maxillary sinus was eroded and the floor of the right antrum was breached by the tumour mass (Fig. 2a and 2b). The mandible was not involved and there was no intracranial or distant metastasis. Tissue biopsy taken from the oral mass revealed spindle cell tumour with islands of cartilage and without osteoid cell suggesting chondrosarcoma. After a proper consultation and discussion with the patient, excision of the tumour was performed by a radical maxillectomy. This created a huge defect which was repaired by an osteomyocutaneous latissimus dorsi free flap based on the thoracodorsal artery. The arterial system was anastomosed to the superior thyroid artery while the venous system was anastomosed to the internal jugular vein. The final histopathological examination of the resected specimen showed features of increased mitosis, spindle shaped tumour cells, chondroid differentiation and island of osteoid indicating the diagnosis of osteosarcoma.



Fig. 1: The mass protruding from the persistently open mouth. (Note: Consent obtained from patient for publication.)

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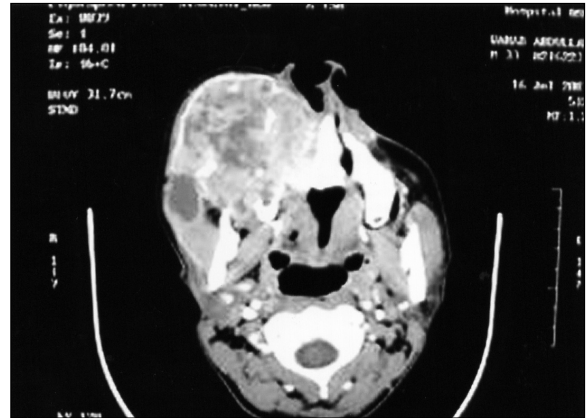
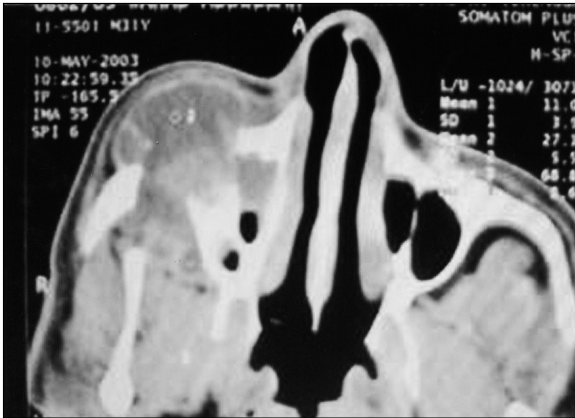


Fig. 2: CT-Scan showed (a) the mass eroding the right anterior wall of the maxillary antrum and (b) the floor of the right antrum was breached.

He recovered well postoperatively and discharged home on day eight post-operation. On follow-up he developed another recurrence after six months. He was given palliative chemotherapy.

DISCUSSION

Osteosarcoma is an uncommon tumour which occurs most frequently in the long bones of adolescents and young adults during the period of maximal growth². The sex distribution is equal and most lesions in the head and neck arise in the mandible and maxilla in patients in the third and fourth decades of life². Osteosarcoma with a predominant chondroblastic element and absence of malignant osteoblastic tissue with mature bone formation or osteoids is difficult to be distinguished from chondrosarcoma. A meticulous search for woven bones is important to obtain the correct diagnosis³. Demonstration of alkaline phosphatase and negative staining for alpha -1 on immunohistochemistry are further evidences in support of osteosarcoma³.

The treatment of choice for osteosarcoma is chemotherapy followed by surgical resection⁴. Imaging modalities of plain x-ray, computed tomographic scan (CT Scan) and magnetic resonance imaging (MRI) are valuable to establish tumour extension and assist in the planning for operative resection. Both resection of the tumor with free surgical margins and appropriate evaluation of the surgical defect for the most suitable reconstruction will contribute to a good outcome for the patient.

Except for its pattern of failure, osteosarcoma of the facial bones has a natural history similar to that of osteosarcoma of the extremities. There is a much lower risk of distant metastasis in the head and neck but a high rate of local recurrence. This may be due to the difficulty in achieving wide surgical margins in the head and neck for anatomic and cosmetic regions. The literature suggests that radical surgery alone frequently results in local failure².

The optimal management for patients with osteosarcoma of the head and neck is unclear. The rarity of the lesion precludes adequate patient enrollment in prospective

randomized studies². The treatment and prognosis of patients with osteosarcomas have changed radically since the introduction of adjuvant polychemotherapy into the various treatment protocols. In addition to prolonged adjuvant chemotherapy, most treatment programs for osteosarcoma now also incorporate preoperative (neoadjuvant) chemotherapy. Preoperative chemotherapy provides early treatment for patients with systemic, occult micrometastases and a detailed histologic evaluation of their response to chemotherapy in the resected specimen by grading the degree of tumor necrosis. This histologic evaluation generates prognostic factors for local recurrence and distant metastases, with the possibility of switching to other chemotherapeutic agents in the adjuvant treatment of poor histologic responders. Moreover, preoperative chemotherapy causes shrinkage of the tumor by necrosis which makes conservative surgical resections feasible and allows time to plan definitive local therapy for primary tumors⁴.

Unfortunately for this patient the treatment given was surgical resection followed by radiotherapy as the initial biopsy revealed a chondrosarcoma. Although the patient was relieved of the massive mass which was extremely foul smelling and the inability to close his mouth due to the protrusion of the mass, he developed recurrence in six months. The recurrence was treated with a palliative chemotherapy. In conclusion, histopathological interpretation requires meticulous examination in the diagnosis of sinonasal osteosarcoma and chondrosarcoma. Approach to the treatment and prognosis depends upon early and correct diagnosis.

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