Collision Tumor of Maxilla: A Diagnostic Dilemma

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Abstract Most malignant tumors arise from the primary tissue except when showing extreme dedifferentiation. This can be identified by examination of tumor cells and their products. Occasionally two distinct tissues are recognized within a malignant tumor. In mixed malignant tumors both carcinomatous and sarcomatous elements are present either because of simultaneous malignant change occurring in the epithelium and its nonepithelial stroma or there is a sarcomatous transformation of stroma of a carcinoma. Willey's in 1960 stated that in these circumstances the resulting tumor must be classified as a "carcino-sarcoma". These separate tumors arise in nearby structures and may grow to infiltrate in each other resulting in a tumor mass. The present article is a report of a diagnostic dilemma in an extremely rare carcinosarcoma of maxilla in a 24 year old male which was first on biopsy reported as odontogenic myxoma. After resection (right maxillectomy under GA using Weber Fergusons approach with Diffenbach's extension) was diagnosed as Squamous Cell Carcinoma arising from lining of the maxillary antrum associated with Osteogenic Sarcoma (chondroblastic differentiation) of maxilla with negative margins. The tumor was restaged to T2NoMx and further managed as per National Comprehensive Cancer Network Guidelines. Immunohistochemistry later re-diagnosed it to be an Osteogenic Sarcoma maxilla with chondroblastic differentiation. 'T' stage remains the most reliable predictor of survival and loco-regional control. Complete surgical resection for all 'T' stages (except T4b, any N) followed by postoperative

therapy remains a corner stone of treatment of maxillary sinus tumors.

Keywords Osteogenic sarcoma · Carcinoma · Immunohistochemistry · Maxillectomy

Introduction

Osteosarcomas (OS) are primary bone tumors of mesenchymal origin, which are quiet uncommon in the craniofacial region accounting for 4 % of total osteosarcomas. About 60 % of the tumors occur in second decade of life and 10 % occur in third decade of life. It is rarely found in very young and the reason for its predominance in second decade corresponds to the growth spurts. Interestingly, the incidence of OS of jaws is more so in the fourth decade of life. It has a slightly higher incidence in males with a ratio of 1.25:1 [1, 2]. The etiology of OS is bizarre, however risk factors do exist, such as rapid bone growth (as suggested by the fact that there is increased incidence during growth spurt, and its typical location at the metaphyseal end of long bones). As far as environmental risk factors are concerned radiation seems to be the only cause. There may exist some genetic risk factor as in familial cases where there is deletion of chromosome 13q14, thereby inactivating the retinoblastoma gene (RB gene), other risk factors include the bone dysplasia, like the Paget's disease, fibrous dysplasia, hereditary multiple exostosis, etc. Li-Fraumeni syndrome and Rothmund-Thomson syndrome are also associated with OS. It is evaluated that 3 out of 16 patients have risk factors for OS [3]. The most common symptom of OS is swelling, followed by loosening of teeth and hypoesthesia. Radiographically, the lesion shows a bizarre form of radiolucency and radio opacity occurring together,

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this hallmark is rarely found in initial cases. However widening of periodontal ligament space is pathognomonic [4]. Computed tomography and MRI are the mainstay for the diagnosis of OS and its relation to its surrounding structures. Histopathologically, osteosarcomas are composed of malignant spindle cells which produce foci of osteoid or immature bone. In the jaws, about half of the lesions demonstrate a cartilaginous differentiation. In general, osteosarcomas of the jaws tend to be better differentiated than their long bone counterparts, with some tumors exhibiting a deceptively bland histological appearance. Therefore, correlation of the histological features with the clinical and radiographic findings is essential for the diagnosis. Immunohistochemistry forms an integral part in the diagnosis of OS [5]. Although, OS of maxilla shows less signs of haematogenous spread and better prognosis than its other counterpart, it becomes important for its early diagnosis and prompt treatment.

Case Report

This 24 year old male patient reported to our department with a complaint of slow growing painless swelling of right upper jaw since last 6 months (Fig. 1). Associated teeth were mobile and two teeth exfoliated since last 2 months. Clinically the swelling was diffuse extending up to right zygoma. Oral examination revealed a firm non tender swelling extending from right maxillary cuspid to tuberosity. Tooth number 17 and 18 were missing and other associated teeth were mobile. Cervical lymph nodes were not enlarged. An incisional biopsy carried out by a local practitioner diagnosed him as a case of odontogenic myxoma and he was referred to our institution for further management. As per the institutional protocol incisional biopsy was repeated which confirmed it to be a case of odontogenic myxoma (Fig. 2). CT scan was suggestive of

Fig. 1 Extraoral and intraoral preoperative photographs

Fig. 2 Micro photograph (×40) showing mixed tissue

Fig. 3 Axial scan showing the lesion

soft tissue growth in the right nasal cavity and maxillary antrum which had eroded the anterolateral wall but the orbital floor, posterolateral wall and pterygoid plates were intact (Fig. 3). Consent for surgery was taken. Under general anesthesia right maxillectomy was carried out using Weber Fergusons approach with Diffenbach's extension (Fig. 4). The lesion was excised in toto with a healthy margin. The postsurgical histopathology report was suggestive of Squamous Cell Carcinoma arising from lining of the maxillary antrum associated with osteogenic sarcoma (chondroblastic differentiation) of maxilla with negative margins (Fig. 5). The tumor was restaged to T2NoMx and further managed as per National Comprehensive Cancer Network (NCCN) Guidelines. Immunohistochemistry studies revealed it to be an osteogenic

Fig. 4 Operative photograph and specimen

Fig. 5 Micro photograph (×40) showing chondroblastic differentiation

Fig. 7 Post operative photograph

Fig. 6 T⁹⁹ bone scan

sarcoma maxilla with chondroblastic differentiation. A T⁹⁹ bone scan study was done which ruled out any metastasis (Fig. 6). After 4 weeks the individual was scheduled for radiotherapy followed by chemotherapy (Fig. 7). Regular follow up visits have been advised as per the schedule given in NCCN guidelines.

Discussion

Osteosarcoma is a very rare tumor of occurrence in the craniofacial region. The diagnosis of the lesion is challenging and needs a multi-pronged approach with the use of radio diagnosis, histopathology and clinical features. The affected patients usually present with pain and swelling in the concerned area as in our case, other clinical features included loosening of teeth, widening of PDL space (Grittman's sign), occasional ulceration and purulent rhinorrhea [2, 6]. Radiologic evaluation is a must in such cases as the clinical signs and symptoms are vague and do not pin point to a specific lesion. A tissue biopsy is the only way of establishing a diagnosis, yet an incisional biopsy may lead to a wrong diagnosis as in our case and there is greater need for immunohistochemistry in these cases. In general, osteoblastic tumors are most common but the chondroblastic variant still persists in case of jaw tumors [4]. In spite of all investigations, a differential such as

chondrosarcoma, Ewing's Sarcoma, Osteiod Osteoma, Odontogenic Myxoma may be considered. In this case we considered Odontogenic myxoma [1]. Although a final diagnosis of OS was established, the treatment plan in our case did not change as it was maxillectomy either ways as per NCCN guidelines. Radiotherapy following surgical excision in case of carcinosarcoma of maxillary sinus had poor prognosis [7]. In high grade OS of jaw bone chemotherapy can be instituted following surgery [8]. We follow the NCCN guidelines for management of all the malignant tumor cases. Reconstruction of the surgical defect depends upon the course of the disease and treatment outcome. We have provided an interim surgical obturator and the patient is awaiting definitive rehabilitation.

It is concluded that the chances of misdiagnosis in OS is quite high and a proper investigative back up is needed to justify the treatment plan. It is article like this that would help to throw light on approaches to diagnosis and treatment plan. For maxillary carcinomas and sarcomas it is the 'T' stage that remains the most reliable predictor of survival and loco-regional control. Complete surgical resection for all 'T' stages (except T4b, any N) followed by postoperative adjuvant radio and chemotherapy to rule out

metastasis remains a corner stone of treatment of maxillary sinus tumors.

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