Case Report

Histopathological Variants of Jaw Osteosarcoma

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Osteosarcoma (OS) is a highly malignant bone forming tumor characterized by frankly to subtly anaplastic stromal cells with evidence of direct formation of osteoid and/or primitive bone by these cells. Incidence is lower in jaws as compared to long bones (4-13%). They have different biological behaviour but same histopathology. The mean age of occurrence is third to fourth decade, almost a decade older than that of osteosarcoma of long bones. The most common presenting features are pain and swelling, with a radiographic presentation of sunburst appearance. The histopathology of osteosarcoma is highly variable, ranging from the more commonly seen osteoblastic type to the rare variants like myxomatous and telangiectatic types among others. The histopathological diversities of jaw osteosarcoma are being presented in the following three cases.

Introduction

Osteosarcoma (OS) is a malignant mesenchymal tumor characterized by formation of osteoid tissue 1. It is most common primary malignant bone tumor, accounting for approximately 20% of sarcomas but only 5% osteosarcomas occur in jaws 2. Osteosarcoma of jaw is uncommon and despite its histopathologic similarities with long bones osteosarcoma, it is biologically different 1. Jaw osteosarcomas usually present in third & fourth decades of life, almost a decade after their presentation in long bone tumors 3. Maxilla & Mandible are equally involved. Mandibular tumors arise more frequently in posterior body and horizontal ramus, whereas maxillary tumours are discovered more commonly in alveolar ridge, sinus floor, and palate 3.

The most common presenting features are increase tumour volume, pain, ulceration and neurological disorders 2. Radiological appearances manifest as mixed radiolucent/radiopaque lesion, periodontal ligament widening, radiopaque masses with moth eaten appearance, codman triangle and sunburst appearance 4.

Osteosarcomas arise in several clinical settings, including pre-existing bone abnormalities such as Paget’s disease, fibrous dysplasia, giant cell tumor, multiple osteochondroma, bone infarct, chronic osteomyelitis, osteogenesis imperfecta, and with history of radiation exposure 3.

WHO lists several variants that differ in location, clinical behaviour and level of cellular atypia. The classical osteosarcoma is the most frequent variant which develops in medullary region of bone & can be subdivided into osteoblastic, chondroblastic and fibroblastic histologic types depending upon the type of extracellular matrix produced by tumor cells 2 Other histological variants include telangiectatic type, small cell osteosarcoma, giant cell and large cell predominant type etc.

The purpose of this paper is to highlight the diverse clinical and histopathological presentations of jaw osteosarcomas through the three cases, came across in our practice.

Case Reports

Case 1

A 29 year old male patient reported to the dentistry department with an intraoral swelling in the lower posterior mandibular region. The swelling was ovoid measuring 5x 6 cm extending from left lower premolar region to anterior border of mandible. Orthopantomogram showed a mixed radiolucent radiopaque lesion extending from left 2nd premolar to distal root of 2nd molar with loss of normal trabecular pattern (Fig 1). CT scan showed extension of lesion into the soft tissue mass with destruction of cortex in ramus area. The biopsy material submitted for histopathology showed a mesenchymal lesion mainly composed of chondroid areas along with few osseous areas (Fig 2). Tumour cell were pleomorphic, plump to oval and spindle shaped. Osteoid consists of homogenous irregularly distributed material. The
chondroid areas showed abundant pleomorphism, atypical binucleated cells and large hyperchromatic nuclei with prominent nucleoli (Fig 3). This case was diagnosed as chondroblastic variant of osteosarcoma.

Case 2

A 23 year old male patient reported to the department with the chief complaint of swelling involving buccal and lingual cortical plates in relation to first and second molar. X-ray showed radiolucent lesion with ill defined borders in relation to first molar tooth involving furcation area and periapical area along with widening of periodontal ligament. The biopsy material submitted for histopathology revealed an aggressively proliferating mesenchymal tumour, showing a tumour osteoid (fig 4) and prominent loose myxoid areas (fig 5). It was diagnosed as Osteosarcoma with myxomatous areas.

Case 3

A 42 years female patient reported with chief complaint of pain and swelling in lower right posterior region. Clinically there was exophytic growth on the alveolar ridge in relation to lower right region extending from 1st molar to 3rd molar region. It was hard in consistency. The biopsy material revealed a tissue lined with stratified squamous epithelium. Subepithelial tissue showed pleomorphic oval to spindle shaped cells entrapped in the osteoid. Tumor cells were laying down early osteoid. Mitotic figures were also frequent (Fig6). The histopathological diagnosis was osteoblastic variant of osteosarcoma

Discussion

Worldwide, OS is considered to be a rare lesion occurring in about 1 per 100,000 persons per year; about 6% to 7% of all OS occur in the maxillofacial region 5. OS of the head and neck are considered by most clinicians to be distinct from OS that arise in the long bones. It is reported to affect older patients and follows a different clinical pattern. In the present study there was slight male predilection 2: 1, all the three cases were in third decade, a finding similar to earlier statistics that jaw lesions arise a decade later than those in the long bones.

OS may arise de novo or subsequent to previous irradiation. It can also be seen in pre-existing Paget’s disease of bone and fibrous dysplasia. Isolated cases of trauma have been stated as contributory factors. All the cases in the present series appear to have developed de novo; as no history of any predisposing factors could be elicited from these three patients. All the cases in the present series arose in posterior mandible 5, which is compatible with earlier reports 6. Radiographically osteosarcoma has a variable presentation with spectrum ranging from osteolytic through mixed osteolytic, osteoblastic to predominantly osteoblastic 7.

Both genetic and locally active epigenetic factors influence whether a particular cell progresses down as osteogenic or chondrogenic pathway. In experimental studies when fibroblastic cells derived from marrow stroma were implanted in vivo in diffusion chamber, a bone-like tissue formed peripherally, with chondroid or fibroblastic areas centrally. These finding suggest that oxygen and nutrient gradient may have a role on cell differentiation to osteoblastic or chondroblastic pathway 1. Alternatively it could be simple various stages of enchondral ossification

Histologically osteosarcomas can be classified according to the cellular differentiation as osteoblastic, chondroblastic and fibroblastic. In osteoblastic type atypical neoplastic osteoblasts exhibit considerable variation in shape and size, show large deeply staining nuclei and are arranged in disorderly fashion and these constitute 60% of jaw lesions. The chondroblastic type has been described to occur predominantly in head and neck region. In this type, tumor consists of atypical chondroid areas composed of pleomorphic and atypical binucleate cells, having large hyperchromatic nuclei and prominent nucleoli. The fibroblastic type is rare, especially in the jaws 8. In myxomatous type there is atypical myxoid proliferation. The majority of tumor is heterogenous reflecting the pleuripotency of proliferating mesenchymal cells 9. Other histologic types include malignant fibrous histiocytoma like osteosarcoma which show spindle anaplastic cells. In large cell predominant osteosarcoma there are large cells with prominent nucleoli. Giant cell predominant osteosarcoma is characterized by anaplastic stromal cell producing streams of osteoid along with giant cells. This type of tumor may be confused with giant cell tumor. In small cell or round cell predominant type osteosarcoma, osteoid producing small malignant cell and primitive bone tissue are characteristic where as in telangiectatic osteosarcoma, anaplastic cells are present along with osteoid4. All the cases in present series showed diverse histopathological patterns ranging from chondroblastic to osteoblastic to myxomatous variants.

Clark et al. reported that the patients with
chondroblastic osteosarcomas of head and neck had a better overall survival rate than the patients with

Fig. 1 Orthopantomogram showing a mixed radiolucent radiopaque lesion (arrows) extending from left 2nd premolar to distal root of 2nd molar with loss of normal trabecular pattern.

Fig. 2 Photomicrograph showing osteoid production (arrow head) with chondroid areas having typical binucleate appearance. (H&Ex100)

Fig. 3 Photomicrograph showing anaplastic cells (arrowhead) in the area of chondroid differentiation (H& Ex400)

Fig. 4 Photomicrograph showing tumor osteoid and myxomatous areas. (H&Ex100)

Fig. 5 Photomicrograph showing tumor producing osteoid (arrow head) and pleomorphic osteoblasts. (H& Ex40)
Fig. 6 Photomicrograph showing dense large areas of osteoid with pleomorphic and bizarre osteoblasts (H&E x100)

osteoblastic or fibroblastic tumours. The prognosis of jaw osteosarcomas is better than that of long bones osteosarcomas. This could be due to histologically better differentiation of jaw osteosarcomas than long bone osteosarcomas. As jaw osteosarcomas occurs at higher mean age, the patients have less chance of developing metastases.

Conclusion

Jaw osteosarcoma presents a wide spectrum of clinical, histological and radiological features. Therefore all these features have to be correlated to reach a conclusive diagnosis. It has a better prognosis if diagnosed and treated at an early stage.

References