CASE REPORT

PERIOSTEAL OSTEOSARCOMA PRESENTING IN JAW BONE; UNUSUAL SITE OF AN UNUSUAL ENTITY

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Periosteal Osteosarcoma (PO) is a rare surface-based bone tumor of intermediate grade, comprising mostly of chondroblastic tumor cells and accounts for 1–2% of all osteosarcoma cases. PO is almost often located in the long bones and affects patients mostly in their second and third decades of life. Mandible is a rare site. We report a rare case of mandibular periosteal osteosarcoma in a 26-year-old female patient. She presented with fracture and swelling left side of mandible. Segmental mandibulectomy was performed. The radiological findings and histologic features led to the diagnosis of Periosteal Osteosarcoma. There is no evidence of local recurrence after 05 months of follow up. This treatise highlights importance of radiological and histological identification of such an unusual entity at such unique primary site of occurrence.

Keywords: Periosteal osteosarcoma; Surface based bone tumor; Osteosarcoma; Mandible

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INTRODUCTION

Osteosarcomas arising from the cortical surface of bone can be subdivided into parosteal, periosteal, and high-grade surface osteosarcoma. Periosteal Osteosarcoma (PO) is a rare primary malignant bone tumor arising from the surface of long bones, especially femur and tibia, comprising less than 2% of all osteosarcomas. To date, less than 15 cases of PO affecting the jaw bones have been reported, with most cases following an evolution time of months to a year, and they favored a benign course following resection.[1] The biological behavior of PO is less aggressive and it portends lesser degree of metastasis.

CASE REPORT

A 26-year-old female with no known comorbid, presented first time to her primary physician with fracture of left mandible bone. She gave history of swollen jaw noticed few months ago. Local examination showed a left sided jaw swelling extending from mid of tragus to the inferior border of mandible bone; hard, not tender to touch and well demarcated. Overlying skin was intact. Core biopsy of the swelling was performed and signed out as "Osteo-cartilagenous lesion with fracture site changes". Later on, she underwent segmental mandibulectomy and reconstruction. Pre surgical radiological images showed a cortical, broad based largely circumscribed lesion showing chondroid matrix with mild internal mineralization, hyperintense in T2. Signs of medullary involvement were not present. (Figure-1) Grossly, the mandible bone showed a relatively circumscribed, firm, nodular mass that measured 4.5×2.2 cm. (Figure-2) Histological examination of the hematoxylin & eosin-stained slides showed a sub periosteal lesion comprising of poorly delineated lobules of atypical hyaline cartilage with intervening bands and sheets of sarcomatous cells exhibiting areas of osteoid formation with variable mineralization. (Figure-3) The sarcomatous element composed of atypical fibroblastic cells showing moderate pleomorphism, stellate nuclei with occasional nucleoli and moderate cytoplasm. Upto 3–5/10 high power field mitoses were counted. Overall features including radiological findings were consistent with periosteal osteosarcoma. No pre surgical treatment was received. There were no signs of recurrence after 05 months of follow up.

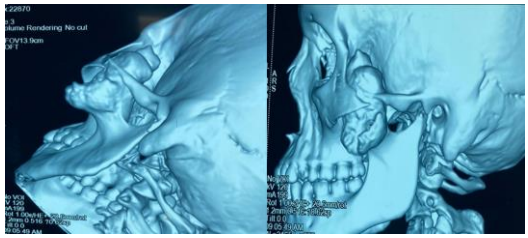

Figure-1: (B) 3D CT scan imaging shows a broad based expansile tumor involving coronoid process of mandible with cortical erosion and fracture of zygomatic arch.

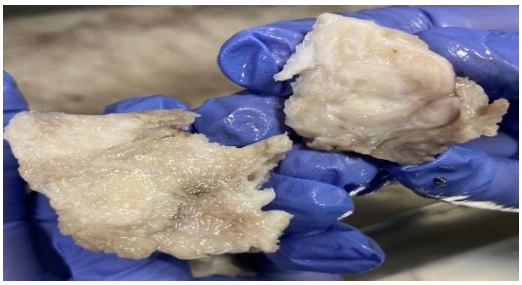

Figure-2: Chondroblastic tumor with glistening grey surface diffusely destructing the bone cortex.

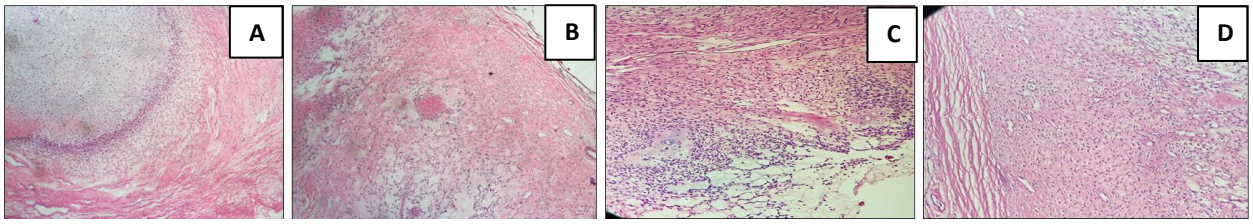

Figure-3, A-D: H & E stained sections show a subperiosteal lesion (A, B) showing chondroblastic and fibrous sarcomatous components(C). The sarcomatous area shows moderately pleomorphic stellate cells with occasional osteoid formation (D).

DISCUSSION

Osteosarcoma is an aggressive malignant bone forming tumor and it represents the commonest sarcoma arising from bones. The estimated percentage of osteosarcoma presenting in the jaw ranges from 2% to 13%. 50–60% of tumors involve the mandible bone compared to the maxilla.[2] Of these, PO accounts for less than 2% of all osteosarcoma cases presenting all over the body. It primarily involves the long bones of limbs. The unique reported sites of rare occurrence include maxilla, mandible, metatarsal, metacarpal bone in a pediatric patient, calcaneum, sphenoid and clavicle.[3] Occurrence of PO in jaw bones is much rare so that only a handful of cases are available for review in literature.

Periosteal osteosarcoma is an intermediate grade osteosarcoma. It originates from periosteum with rare signs of medullary involvement[i]. It occurs in a wide age range; mean age 35 years (range, 16–65 years), presents almost equally in both genders, and occurs more in mandible compared to maxilla (commonly in posterior region). Time lapse from occurrence until diagnosis varies in months to years, mean time until presentation is 06 months according to some authors.[1] In contrast, parosteal osteosarcoma, also a surface-based bone tumor, shows slight female predilection, with a longer evolution time until diagnosis, and good prognosis after adequate surgical treatment.

Histologically, POs show a mixture of cartilage and bone forming areas. There are lobules of cytologically atypical cartilage with intervening bands of primitive sarcomatous cells in which osteoid formation is evident. Periphery of the tumor shows more primitive appearing undifferentiated mesenchymal cells with nuclear atypia and mitotic figures. Such histology offers a differential diagnosis of small round blue cell tumors, especially Ewing sarcoma, or low-grade central osteosarcoma. Cartilagenous areas may show transition to osteoid matrix. MDM2/CDK4 amplification or IDH mutations are not present in this tumor.[5] A specific immunohistochemistry is not recommended or proposed to date. However, other differentials of small round blue cell tumor, Rhabdomyosarcoma and Neuro-endocrine carcinoma can be excluded through correlation with radiologic findings along with application of relevant immune histochemical markers and molecular study. Complete resection with wide margins is the treatment of choice for PO. The benefit of chemotherapy, though it is frequently used, is yet doubtful and much controversial, and several studies showed that it has no impact on overall outcome or survival.[6] According to the current literature, the local recurrence rate is 5.6–40%, and the rate of metastasis is 11.1–22.2%. Of this, lung and pleura are the commonest sites of metastasis.[7] Due to the rarity of this tumor, there is no evidence-based study for chemotherapeutic treatment regimen in metastatic cases. Such as, for example, in case of lung metastasis of colon cancer, palliative and complete resection of the lesions is usually the recommended treatment.

CONCLUSION

PO of the jaw appears to have a slightly more favorable prognosis than that in long bones, although a larger series of cases needs to be evaluated before a definitive conclusion concerning the behavior of these juxtacortical bone tumors of the jaw can be made. There is a dire need to identify a potentially predictive biomarker for PO patients who may thus benefit from systemic chemotherapy and have an improved survival and function. Moreover, identification of this entity is crucial in sparing these patients from more aggressive and radical treatment options recommended for conventional, high-grade osteosarcomas.

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Ethical review

Not required

Consent

Informed consent was taken from the patient for the publication of this case via telephone.

AUTHOR CONTRIBUTIONS

Anam Ghauri: Writing; original draft, literature review, slides review, took photomicrographs of slides. Afifa Ghauri: Conception of study, literature review, pictures of radiologic films, critical review. Both authors read and approved final manuscript.

Availability of patient data

All data regarding the case has been included in this article.

Competing interests

The authors declare that they have no conflict of interests.

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