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Gnathic Osteosarcoma in a 15 Year Old Male in Calabar- Nigeria

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Abstract

Osteosarcomas are malignant bone forming tumours and the second most common primary bone tumours after multiple myeloma. The imaging features of less common subtypes of primary osteosarcoma are variable and frequently overlap with those of multiple benign and malignant entities, creating substantial diagnostic challenges. This is a case report of a 15 year old male who presented a dental clinic with five months history of a solid left jaw mass. Clinical examination revealed a stable, moderately cachectic, afebrile and mildly pale with obvious facial asymmetry. Computerized Tomography showed facial asymmetry with a large irregular hyperdense destructive mass involving the bony cortex and the medulla of the left mandible with speculation of its outer cortex giving the classical sun burst appearance. A diagnosis of gnathic osteosarcoma was made. Misdiagnosis of osteosarcoma of the jaws, as tumors or cysts of odontogenic or non-odontogenic origin leads to alternative treatments such as enucleation, marsupialization, curettage or segmental resection. This will led to the dissemination of the tumor. Thus, histological and radiological knowledge of such highly malignant tumors is essential for early and adequate treatment.

Introduction

Osteosarcomas are malignant bone forming tumours and the second most common primary bone tumours after multiple myeloma.[1] They account for about 20% of all primary bone tumours and occur in primary and secondary forms, each with different epidemiology and distribution.

Primary osteosarcoma mainly occurs in young patients (10-20 years) with 75% occurring before the age of 20; which is logical because the growth centers of the bone are more active during puberty to adolescence period.[2] For unknown reasons, there is a slight male predominance.[1,2]

Secondary osteosarcoma**,** is seen in the elderly, usually secondary to malignant degeneration of Paget's disease, extensive bone infarcts, post-radiotherapy for other conditions, osteochondroma, and osteoblastoma.[3]

The current World Health Organization classification of bony osteosarcoma includes eight categories: conventional, telangiectatic, small cell, low-grade central, secondary, parosteal, periosteal, and high-grade surface.[4] Although the conventional and secondary osteosarcoma are histologically indistinguishable, diagnoses of the conventional form and secondary form are made on the basis of typical radiographic appearances (that is, a destructive mass with cloudlike radiopacity in long bones and a mass arising from a preexisting abnormality such as Paget disease, respectively). The imaging features of less

common subtypes of primary osteosarcoma are variable and frequently overlap with those of multiple benign and malignant entities, creating substantial diagnostic challenges.[4,5]

The conventional high-grade form is the most common, which was incidentally found in the index case.

Histological findings associated with conventional radiography are distinctive and allow for differentiation between the subtypes. Macroscopically osteosarcomas are bulky tumours where a heterogeneous cut surface demonstrates areas of hemorrhage, fibrosis and cystic degeneration. Their extension within the medullary cavity is often much more extensive than the bulky part of the tumour would suggest. Areas of bone formation are characteristic of osteosarcomas, with the degree of bone formation varying widely.[12,3]

Microscopically poorly formed trabecular bone is seen with (in the typical high grade conventional subtype) cellular pleomorphism and mitoses and variable amounts of fibrocytic and chondroblastic appearing cells may also be encountered. [3,4]

They typically occur at the meta-diaphysis of tubular bones in the appendicular skeleton. Common sites include: femur: (40% especially distal femur);tibia: (16% especially proximal tibia); humerus: (15%).Other less common sites include; fibula, innominate bone (oscoxae), mandible (gnathic osteosarcoma) ,maxilla and the vertebrae.[5,6]

Case Study

F. L is a 15 year old male junior secondary school (JSS) three pupil who presented at the dental clinic patient with five months history of a solid, left jaw mass. The mass was intermittently painful and seen to be progressively increasing in size, precluding optimal opening of the mouth and proper mastication. There was no history of tooth ache, allergies or systemic illness. Patient is also said to have lost weight significantly.

On examination, patient was stable, moderately cachectic, afebrile and mildly pale with obvious facial asymmetry. A large, non-tender, solid mass was seen in the body of the left mandible extending to the para-symphyseal region and the left condyle. There was no differential warmth. Enlarged cervical and submandibular nodes were visualized. On inspection of the oral cavity, halitosis was noted with a suggestion of minimal dental anarchy. A preliminary diagnosis of osteosarcoma of the jaw was made. The mass was biopsied and the results proved that this was a case of Pleomorphic osteosarcoma. Plain radiography was not done for the patient, rather the managing team requested for computerized tomography which showed facial asymmetry with a large irregular hyperdense destructive mass involving the bony cortex and the medulla of the left mandible with spiculation of its outer cortex giving the classical sun burst appearance. (Fig. 1); the lesion also extended to the ipsilateral maxilla.

The 3 dimensional tomographic image of the jaw shows a dense mesh work of dysplastic bone (Fig. 2). A chest x-ray was done for the patient to exclude metastasis. This was essentially normal. The index child was booked for surgery two weeks later and a subtotal mandibulectomy and latissimusdorsi flap reconstruction for the face was done. Post up condition was satisfactory. Patient has long been discharged to the out -patient clinic to commence chemotherapy at a later date.

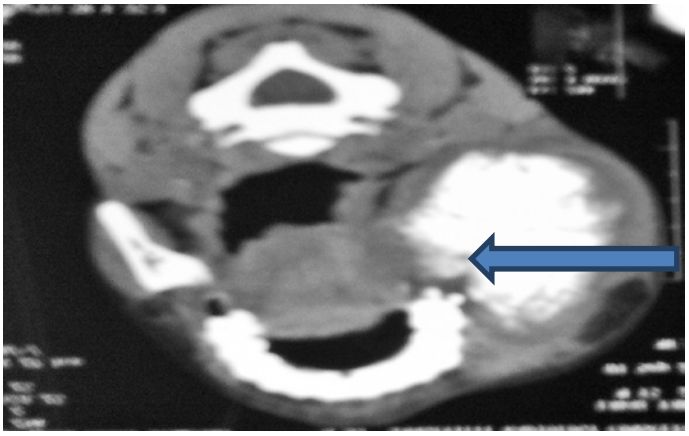

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Fig. 1: Axial non-contrast enhanced CT of the face, showing an extensive, destructive, irregular, hyperdense left mandibular mass involving the bony cortex and the medulla with extension into the adjacent soft tissue. Spiculation of the outer cortex of this mass gives the classical sun burst appearance seen in osteosarcoma (blue arrow).

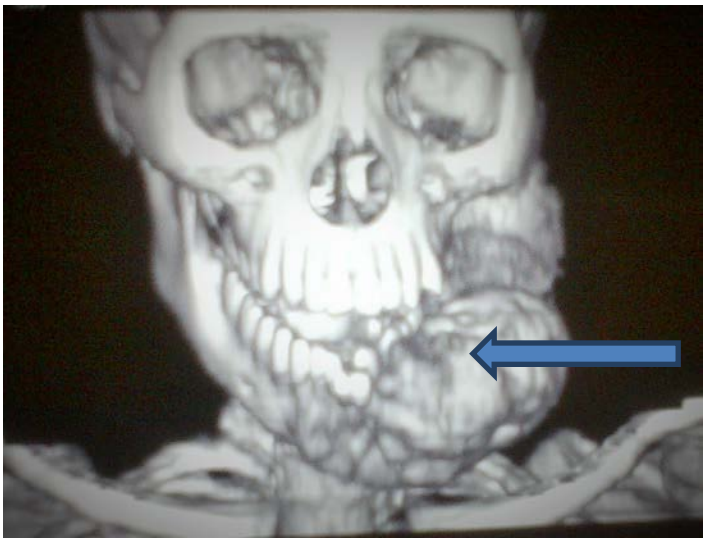

Fig. 2: 3-D computerized tomographic imaging of the left jaw shows an extensive area of bony destruction involving the left maxilla, body and ramus of the mandible up to the margins of the left mandibular condyle in keeping with osteosarcoma (blue arrow).

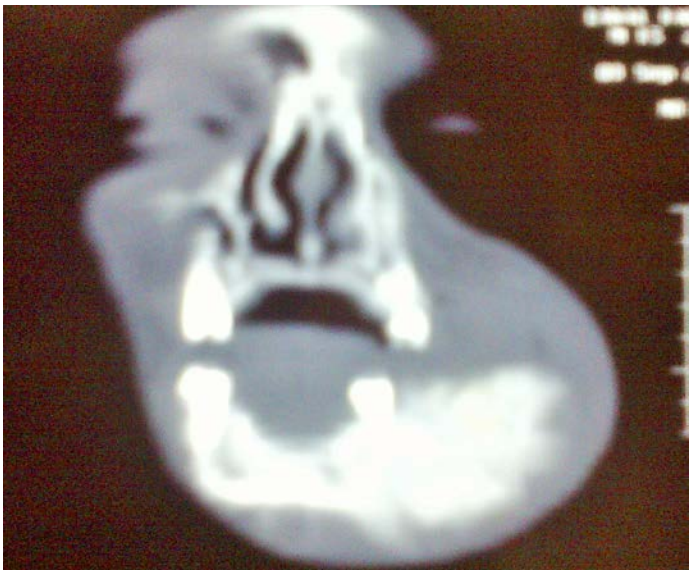

Fig. 3: Coronal non-contrast enhanced CT of the face showing facial asymmetry and gross osseous destruction with sun burst appearance.

Discussion

Osteosarcomas of the jaw bones are very rare, aggressive, malignant mesenchymaltumour which are characterized by the formation of osteoid tissue, with an incidence of 0.7 per million.[4] They differ from osteosarcomas of the long bones in their biological behaviour, even though they have the same histological appearance.[4,5] Lesions of the mandible and maxilla constitute 6% to 9% of all osteosarcomas.

Jaw osteosarcomas constitute the third most common malignancy in children and adolescents. 900 new cases are diagnosed in the United States per year and 400 of these are in patients younger than 20 years old; about 80 percent of these tumors are non-metastatic at time of presentation.[5.6] Peak incidence is in persons aged 10-20 years .Male to female ratio is 3:2.[5,6] The index case is male and 15 years old.

Osteosarcoma of the jaw has no known etiology., rather genetic alterations of certain genes has been postulated such as the *genes p53, Rb, met, fos, sas, mdm2, cdk4, and prim1*[4] *Osteosarcoma affects the* mandible (gnathic osteosarcoma) and the maxilla with equal affectation, however, the males showing a predilection for occurrence in the mandible and females in the maxilla.[7, 8,9]

The mandibular tumours arise more frequently in the posterior body and in the horizontal ramus, whereas the maxillary tumours are discovered more commonly in the alveolar ridge, the sinus floor, and the palate.[8,9,10] The index case was seen to arise from the body of the mandible with extension into the maxilla.

Serum alkaline phosphatase (ALP) may be raised (particularly with advanced disease). These serve as disease markers for pre and post-surgical evaluation.[4]

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The characteristic clinical presentation of osteosarcoma of the jaw is swelling, compared with pain in long bone lesions with the most common symptoms included swelling with or without paresthesia and limitation of mouth opening. Other features include periodontal ligament invasion, tooth mobility or displacement.[5,7,8] The index patient had limited mouth opening due to the presence of the tumour.

The preferred modalities for evaluating osteosarcoma of the jaw are plain radiography computerized tomography and, magnetic resonance imaging.[1-5]

The characteristic radiological features are sun-burst appearance, periosteal lifting with formation of Codman's triangle, new bone formation in the soft tissues along with permeative pattern of destruction of bone and other features for specific types of osteosarcoma.[4,6,8] The index case showed an expansive, osteolysis with the classical sun burst appearance as seen in Fig 3. Thetelangiectatic variety of bone is characterized by Osteolysis and expansion while more of osteoblastic appearance is seen in the sclerosing type of osteosarcoma.

After chemotherapy the tumor becomes well defined, capsulated, and more mineralized. Chest X-ray can detect metastasis in form of cannon ball appearance or nodules in the lungs.[7] Chest x-ray in the index patient was normal. A normal chest radiograph however does not preclude pulmonary metastasis. A Computerized tomographic imaging of the chest is superior in this regard.

Computerized Tomography scan delineates the bony anatomy; bony architecture like cortical integrity, pathological fracture and assessment of ossification and calcification (chondroidcomponent) are also more accurately demonstrated.[8,9] However, the soft tissue component and medullary extent is best defined by Magnetic resonance imaging.[10]

Magnetic resonance imaging (MRI) is the most accurate tool for determining the extent of the tumour, within and outside the bone. It precisely delineates the soft tissues, the medullary canal, joint and the growth plate involved; presence of skip lesion in the same bone and across the joint in other bone, proximity and/or encasement of the neurovascular bundle by the tumor.[8.9] In addition, the response of chemotherapy is being judged by MRI. As the neo-angiogenesis decreases with chemotherapy, necrosis occurs and the tumor shrinks with better capsulation. This is done by performing a contrast enhancement and diffusion MRI.[11,12] MRI is also being coupled with Positron Emission Tomography for detection of the systemic involvement of the tumor, local recurrence, and metastasis after treatment. [11] In view of the nonspecific findings of an MRI, it should always be correlated with the patient's x-ray.

Differentiation of osteosarcoma from other bony lesions like Paget’s disease, fibrous dysplasia, multiple myeloma, metastatic tumors and odontogenictumour is based more on microscopic than radiological evidence. [5,7]

Osteosarcoma of the jaw could be treated by a combination of surgical excision and chemotherapy. This was done for the index patient.

Conclusion

Misdiagnosis of osteosarcoma of the jaws, as tumors or cysts of odontogenic or non-odontogenic origin leads to alternative treatments such as enucleation, marsupialization, curettage or segmental resection. This will led to the dissemination of the tumor. Thus, histological and radiological knowledge of such highly malignant tumors is essential for early and adequate treatment.

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