Case Report

A Case of Ewing's Sarcoma of the Clavicle

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ABSTRACT

Aim: The Clavicle is a rare primary site for Ewing sarcoma (ES). We report one case of patient with clavicular ES under our tracking and review the related literatures on imaging of this rare tumor.

Methods and Material: A 22 year old female came with complain of swelling at clavicular region on right side in the radio-diagnosis department .However no pain or fever and the swelling were not tender. Patient is initially investigated with plain radiograph than with CT and MRI.

Results: We report here a case of ES of the clavicle in an adult female because of its rare site and difference from typical radiological appearances elsewhere.

Conclusions: Ewing's sarcoma of the clavicle is rare lesion with non - specific imaging findings different from that seen in other long bones. The MRI helped to characterize the lesion by defining the soft-tissue components of the tumor and the involvement of the adjacent bones.

Keywords: Ewing's Sarcoma; Adults; Clavicle

INTRODUCTION

It is evident that Ewing's Sarcoma is the second most common childhood bone cancer [1]. It occurs more often in the femur and pelvis however any bone can be involved. According to previous studies the age at diagnosis usually younger than 30 years, especially 10 - 15 years. It is more common in male with predominance 3:2, and 95% of Ewing sarcomas occurs in white patients [2,3].Ewing Sarcoma rarely affects the clavicle bone[4,6]. Clavicle has its oncological characteristics with flat bones and not with other long bones. Majority of clavicular tumor are malignant [4], and the diagnosis may be missed due to low level of suspicion. Clavicle has different features than other long bones [5]. It is the only long bone which located in horizontal axis in its anatomical position. It ossifies by membranous ossification and doesn't have a definite medullary cavity. There are one secondary center ossification and two primaries. It is the first bone to ossify in the embryo (fifth month). The clavicle can be resected without causing significant disability and it is in subcutaneous plane throughout its length. Supraclavicular nerve occasionally pierces the clavicle[7].

Because of rarity of the tumours in this bone and the relative lack of literature on the management of tumours in this bone, we report the imaging findings, distribution, clinical features and histopathological correlation with primary tumours and tumor-like conditions involving the clavicle.

CASE REPORT

A 22-year-old female presented to our department with an enlarging, palpable, painless mass in the region of the right sterno-clavicular joint. There was no history of trauma, and there was no restriction of shoulder joint movement.

The patient underwent X-rays which showed irregular bony destruction in its center part with increased bone mineral density and irregular periosteal reaction, and adjacent soft tissue swelling shadow in the 1/3 diaphysis of the clavicle(Figure1). However, the sterno-clavicular joint is normal. A thorax computed tomography demonstrate osseous destruction with obscure boundary, irregular bone cortex increased proliferation sclerosis, Onion peel reaction or lace sample layered, periosteal reaction in the distal end of the right clavicle (Figure 2,3), However there were no pulmonary nodules are seen. Magnetic Resonance Imaging (MRI) reported a lobulated, expansile abnormal signal signal intensity mass lesion involving medial end of clavicle and adjacent soft tissues appear hyperintense on T2 and STIR, isointense on T1 with destruction of cortical surface and adjacent irregular periosteal reaction . On Post contrast, heterogeneous avid enhancement was noted(Figure 4,5,6,7). A needle biopsy revealed small round blue cells consistent with Ewing's Sarcoma.

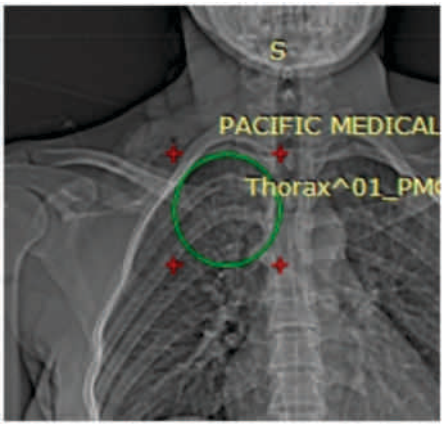

Figure 1: AP X-ray Chest

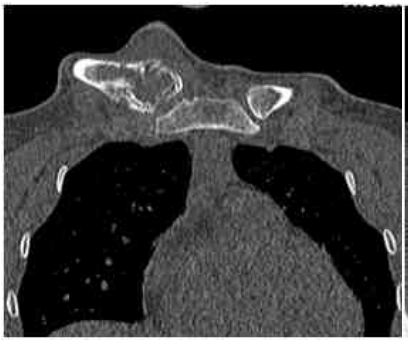

Figure 2: CT Thorax coronal view

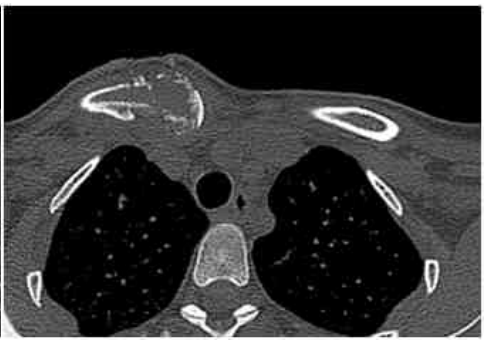

Figure 3: CT Thorax Axial View

Figure 4: MRI Coronal T2

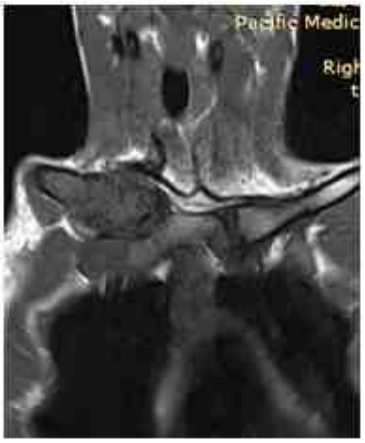

Figure 5: MRI Coronal T1

Figure 6: MRI Axial T1

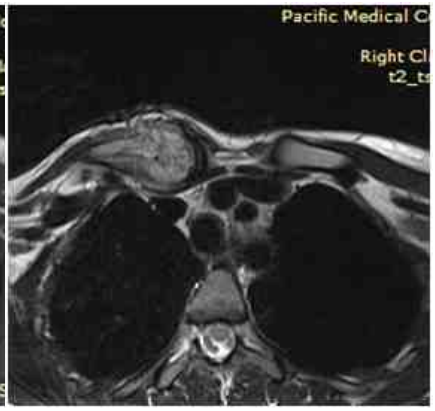

Figure 7: MRI Axial T2

DISCUSSION

Ewing's Sarcoma is a poorly differentiated neoplasm composed of small-round-blue cells; it was originally described in 1921 by James Ewing [8]. Often presenting as a painful, rapidly growing soft-tissue mass 5–10 cm large, it typically arises from the medullary cavity with invasion of the Haversian system and has been described in virtually every bone in the body. It comprising about 4-6% of all primary bone tumors however reported as occurring at all ages. The maximum occurs within the first two decades. It is the most common primary neoplasm of bone after osteosarcoma in second decade. The most common site is in the diaphysis of long bones, less common in ribs, pelvis and vertebrae [12]. Involving head and neck in Ewing's sarcoma is very uncommon, approximately 1% to 4% of cases [13]. Males are predominantly affected than females; the ratio is 3:2. Ewing's sarcoma is more common in whites; blacks and Asians are unusually affected unlike osteosarcoma. [9]

Mostly patient come with complain of pain and local swelling, hyperthermia, anemia, increased erythrocyte sedimentation rate, dilated veins and leukocytosis. Previous trauma history is reported in many cases [13]. The typical imaging features of Ewing's sarcoma are a permeative destruction of bone along with a large soft tissue mass. It can cause a periosteal reaction with the characteristic lamination (onion skinning) or vertical spiculation in one half of the cases. Most usual imaging features reported in Ewing's sarcoma of the clavicle are as follows: a poorly defined permeative lesion with or without associated sun-ray spicules of the periosteal bone, localized honeycomb appearance, cortical erosion, and a soft tissue mass next to bony destruction, displacement or destruction.[10].

If CT is not used both intraosseous with adjacent soft tissue involvement is underestimated. In our study CT shows both the soft tissue mass and the intraosseous origin of the tumor is seen. Magnetic resonance imaging is the choice of imaging to evaluate the extent of the primary lesion, to monitor the response of chemotherapy and to follow up. To rule out skeletal metastasis bone scintigraphy is necessary and 201-thalium scanning is sensitive in the monitoring of the treatment response [14].

The Imaging differential diagnosis of Ewing's sarcoma of the clavicle consists of "osteogenic sarcoma, neuroblastoma, lymphosarcoma, eosinophilic granuloma, osteomyelitis, plasmacytoma and metastatic carcinoma". There is involvement of large soft tissue mass aided interpretation of Ewing's tumor from osteomyelitis and eosinophilic granuloma [10]. Age of the patient ruled out neuroblastoma, which is seen in less than 5 years age group. However, radio-diagnosis is not a totally reliable guide to interpretation and histopathological examination is compulsory to confirm the nature of the tumor.

Clavicular Ewing's Sarcoma having a better prognosis than long bone or pelvic location, since diagnosed earlier [15]. Treatment rates of have improved from 10% to 75% with the use of a of local therapy and extended multi-drug chemotherapy, to decrease both the incidence of local disease recurrence and the development of pulmonary and skeletal metastases. Complete surgical resection is preferred in Ewing Sarcoma local control in those instances when surgery can be performed with minimal loss of function and disfigurement [8]. To conclude, although the classical feature i.e. "onion peel type periosteal reaction described in Ewing's sarcoma elsewhere, may not be present, a lytic expansile lesion of the clavicle with a soft tissue mass and intramedullary origin should suggest in young age the possibility of Ewing's Sarcoma as a differential diagnosis".

CONCLUSION

We have described a rare case of Ewing's Sarcoma of Clavicle showing excessive fibro-osseous response which is not a frequent presentation. Because of its high metastatic potential, Ewing's sarcoma demands early intervention. Evaluation of lesion using plain radiographs, CT, MRI, biopsy followed by histopathology and immunohistochemistry are necessary for early diagnosis.

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