Case report

Multifocal osteosarcoma

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Background: Osteosarcomatosis (also known as multifocal osteosarcoma or multiple sclerotic osteosarcoma) describes a condition in which there are multiple intraosseous foci of osteosarcoma at the time of presentation.

Case report: In a patient of multifocal osteosarcoma the simultaneous appearance of multiple skeletal tumors without pulmonary metastatic lesions was seen. However symptomatic lesion was large in comparison to satellite lesions. Case is reported and general subject of multifocal osteosarcoma is reviewed.

Introduction

Osteosarcomatosis (also known as multifocal osteosarcoma or multiple sclerotic osteosarcoma) describes a condition in which there are multiple intraosseous foci of osteosarcoma at the time of presentation. Previous investigators have considered osteosarcomatosis to represent multicentric primary neoplasia. More recently it has been suggested that all cases of osteosarcomatosis represent rapidly progressive metastatic disease[1].

Case report

A ten-year-old boy presented with swelling, pain and restricted movements in the region of left knee joint and history of fever from last one months duration. On examination knee was held in flexed position with ill-defined soft tissue mass and tenderness adjacent to upper tibia. Physical examination revealed increased body temperature (39.0 degree C). Routine blood tests showed Anemia and leucocytosis.

Radiographs of left knee region demonstrated extensive epiphyseal and metaphyseal osteosclerotic lesions in tibia. Metaphyseal and diaphyseal lesions were also seen in femur. Metaphyseal lesion of tibia showed evidence of aggressive periosteal reaction in form of Codman's triangle with adjacent soft tissue mass. Poorly defined intramedullary dense lesions were noted in femoral diaphysis. These findings were alarming and all indicated towards possibility of a malignant bone tumor with soft tissue invasion. So further imaging studies CT and MRI were done.

CT scan of upper tibia confirmed the presence of densely sclerotic lesion of upper tibia with hyperdensity of adjacent muscles indicating invasion by tumor. CT scan of chest was normal.

T1 weighted images of MRI demonstrated multiple hypointense lesions in metaphysis, epiphysis of tibia and femoral metaphysis and diaphysis indicating a diffuse sclerotic pathological process. Soft tissue mass near upper tibia is very well delineated which was hypointense on T1 weighted images and showed a mixed signal intensity on T2 weighted images.

Biopsy report from Tibial lesion was Osteogenic sarcoma.

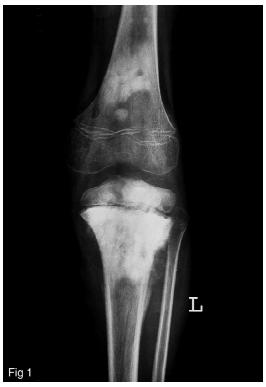
Considering the imaging findings diagnosis of multifocal osteosarcoma was made.

Comments

Multifocal osteosarcoma is an uncommon condition hence this case is reported. Classical imaging features are shown in Radiograph, CT and MRI. These findings are similar to the findings mentioned in review of literature of 262 patients.

Discussion

Osteosarcoma is the commonest primary neoplasm of the bones in children and adolescents. The commonest form is the high-grade intramedullary variant also called conventional osteosarcoma. Long bones are typically involved. The most frequent sites are distal femur, proximal tibia and proximal humerus. Metaphysis is involved in majority of cases[2,3]. Primary involvement of diaphysis and epiphysis is very rare. Clinical presentation includes swelling, pain and enlargement of extremity. Radiographs typically show a mixed sclerotic and lytic lesion in metaphysis of afflicted bone. This pattern is accompanied by aggressive periosteal reaction (sunburst, codman triangle, hair-on-end) and by soft tissue swelling. Metastasis of osteogenic sarcoma have been reported to occur in lungs, Lymphnodes, spleen, kidneys, pancreas, liver pleura, heart, skin and brain and small bowel and



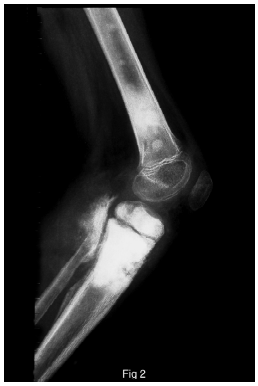


Fig. 1 **Fig. 2**

Fig 1 and 2: *Radiographs of left knee demonstrated multiple extensive epiphyseal and metaphyseal osteosclerotic lesions in tibia. Metaphyseal and diaphyseal lesions were seen in femur. Metaphyseal lesion of tibia showed evidence of aggressive periosteal reaction in form of Codman's triangle with adjacent soft tissue mass. Poorly defined intramedullary dense lesions were noted in femoral diaphysis. These findings were alarming and all indicted towards possibilty of a malignant bone tumor with soft tissue invasion.*

muscles. In several cases there is more than one lesion at the time of presentation. (Multicentric osteosarcoma)[2]. A slight male predominance exists among young patients (<18 yrs) and a much greater male predominance is present approximately 3:1 among older patients (>18yrs age)[4] Osteosarcomatosis is uncommon accounting for approximately 3-4 %of osteosarcoma cases. However multifocal skeletal involvement by osteosarcoma has been found at autopsy in as many as 48% cases. Although osteosarcomatosis is believed to be more common in skeletally immature patients. Hooper et al reported a series of 29 cases in which equal numbers of skeletally immature and mature patients were seen. Younger skeletally immature patients tend to have rapidly appearing usually symmetric lesions where as older patients typically have fewer, asymmetric sclerotic lesions. In 97% cases of those cases reported by Hooper et al a radiologically dominant skeletal tumor was seen. The radiological features of dominant lesion include ill-defined margins, aggressive looking periosteal reaction, adjacent soft tissue extension and cortical disruption. Although lesions usually contain cloud like osteoid, purely lytic dominant lesions may be seen. In contrast to dominant lesion the secondary foci are often smaller, more sclerotic, better defined, and lack periosteal reaction or cortical destruction[1]. From the review of literature only 262 cases have been reported. The common features identified are multiple radio-dense lesions that present simultaneously or within 5 months of initial presentation with or without pulmonary metastasis single dominant lesion with multiple, symmetrical or not smaller lesions and a uniformly rapid fatal prognosis[3]. Multifocal osteogenic sarcoma is also reported in cases of Paget's disease. The most serious complication of Paget's disease is sarcomatous degeneration. Multifocal sarcomatous degeneration occurs mainly in polyostotic Paget's disease and can arise in any site. Compact and confluent sclerotic foci superimposed upon characteristic Pagetic changes and accompanied with soft tissue mass are highly suggestive of malignant transformation[5].

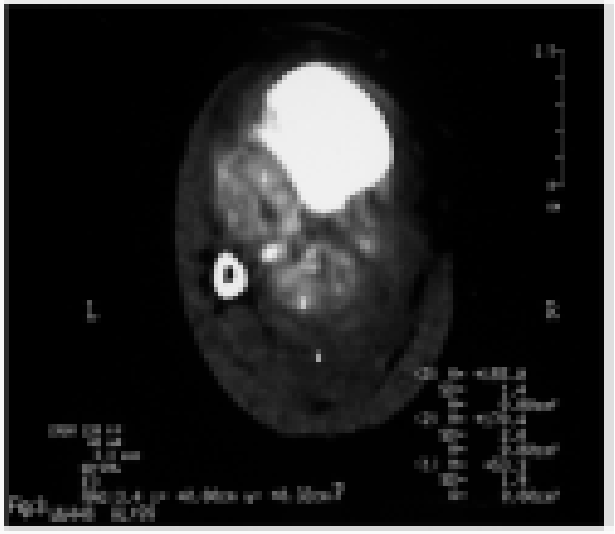


Fig. 3: *NECT scan of upper tibia confirmed the presence of densely sclerotic lesion of upper tibia with hyperdensity of adjacent muscles indicating invasion by tumor.*

T1 weighted longitudinal images depict the intramedullary extent of tumor and identify skip lesions. Determination of tumor soft tissue extent, muscle infiltration, relationship to major neurovascular structures and evaluation of adjacent joint is accomplished by true T1W and heavily T2W axial plane images[6]. Berquist et al has correlated histologic features with MR imaging features.cortical destruction is seen as increased signal intensity on T2 W images. Marrow involvement/skip lesions as decreased signal intensity on T1W images. Periosteal elevation as decreased signal intensity at margin on T2Wimages.Osteosclerotic zones as decreased signal intensity on T1WI and T2WI.Most malignant bone tumors have inhomogeneous signal intensity both on T1WI and T2WI.The degree of inhomogeneity and signal intensity varies with tumor matrix (chondroid, osteoid, or fibrous) and the presence of haemorrhage, necrosis or both[7].

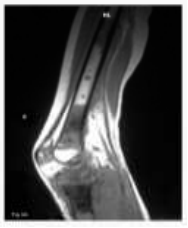
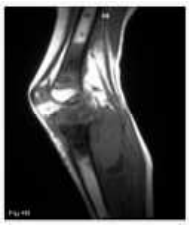
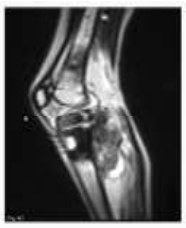




Fig. 4 A ***Fig. 4 B*** ***Fig. 4 C***

Fig 4 A, B & C: *NE Sagittal T1and T2 weighted images showing multiple hypointense lesions in left Tibia and femur with intervening normal bone marrow and a large soft tissue mass adjacent to tibial lesion posteriorly shows heterogenous signal intensity.*

In CT scan imaging all images should be photographed with both bone and soft tissue window. Bone windows will best display calcified tumor matrix and cortical involvement. Soft tissue window settings are needed to evaluate marrow space for tumor invasion (which appear as loss of the normal low density fat.) Extraosseous soft tissue extension and the relationship of tumor to major neurovascular structure[6]. The radiographic differential diagnosis of multicentric osteosarcoma include metastatic carcinoma, diffuse skeletal angiomatosis and multifocal chronic recurrent osteomyelitis. Metastatic carcinoma that give rise to osteoblastic lesions such as Breast and prostatic cancers are uncommon in young patients. Skeletal angiomatosis may radiographically show multiple blastic, lytic or mixed foci in the axial and appendicular skeleton. Chronic recurrent multifocal osteomyelitis is a rare condition in childhood[8]. In conclusion, we present a patient with multifocal osteosarcoma. This lesion illustrates a pattern of a dominant lesion [left upper tibia] and multiple skeletal and soft tissue foci at the time of presentation.

References

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