Multifocal osteosarcoma, a case report

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Citation

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Abstract

Multifocal osteosarcoma is identified when two or more tumors simultaneously appear in the skeleton. Approximately 1-3% of osteosarcoma patients involve multiple bones[1]. It is controversial whether these tomors are of multicentric origin or represent osseous metastases[2].

CASE REPORT

A 5 year old child presented with rapidly growing swelling and pain in the lower end of the right leg since 6 months. She also had generalized weakness, loss of appetite and loss of weight.

Examination showed a 10 x 8 cm swelling in the middle and lower 1/3rd of the right leg with ulceration and hyperpigmentation.

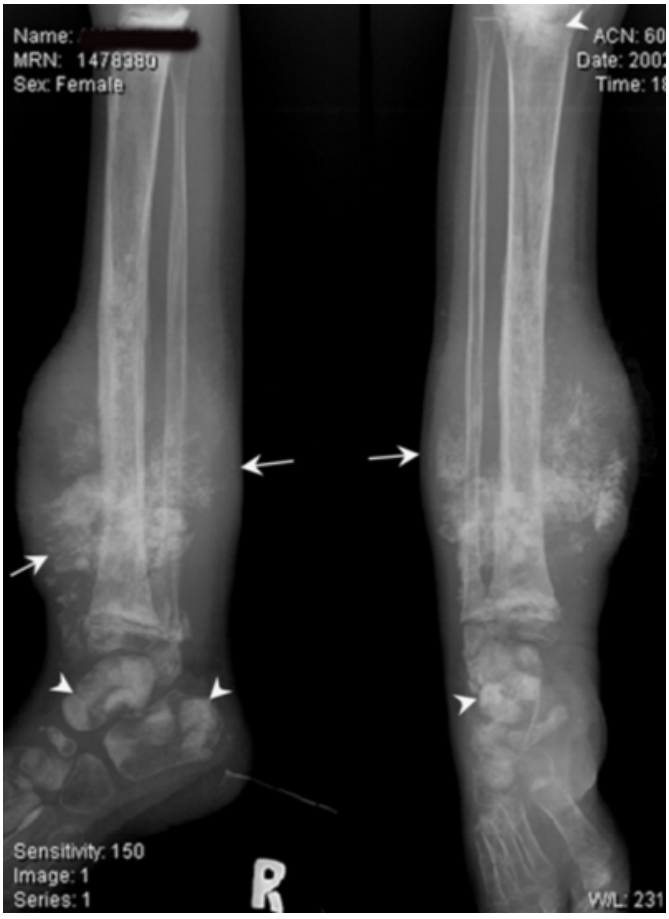
Laboratory investigations revealed raised alkaline phosphatase and lactate dehyogenase levels and rest unremarkable.

RADIOLOGICAL FEATURES

Plain radiographs revealed permiative destructive lesion with lytic and sclerotic areas noted in the lower metadiaphyseal region of the right tibia. Interrupted periosteal reaction was noted with tumor matrix ossification and extension of the tumor into the soft tissues (Fig 1).

Figure 1

Fig 1: Anteroposterior and lateral radiographs of the dominant lesion in the right tibia showing characteristic features of osteosarcoma (arrow) and the multiple sclerotic foci in the upper tibial metaphysis and in the tarsal bones (arrowhead).



In addition, multiple focal sclerotic lesions were noted in the upper and lower metaphyseal ends of the right tibia, right tarsal bones, upper metaphysis of left humerus, lower

metaphysis of right femur, lower metaphysis of the left tibia and in the left iliac creast (Fig 1 & 2). The lesions in the long bones were adjacent to the growth plate, few of them were well-defined and few of them showed relatively wide zone of transition. Chest radiograph was unremarkable.

Figure 2

Fig 2a, Radiograph of the pelvis including thighs and knee joints

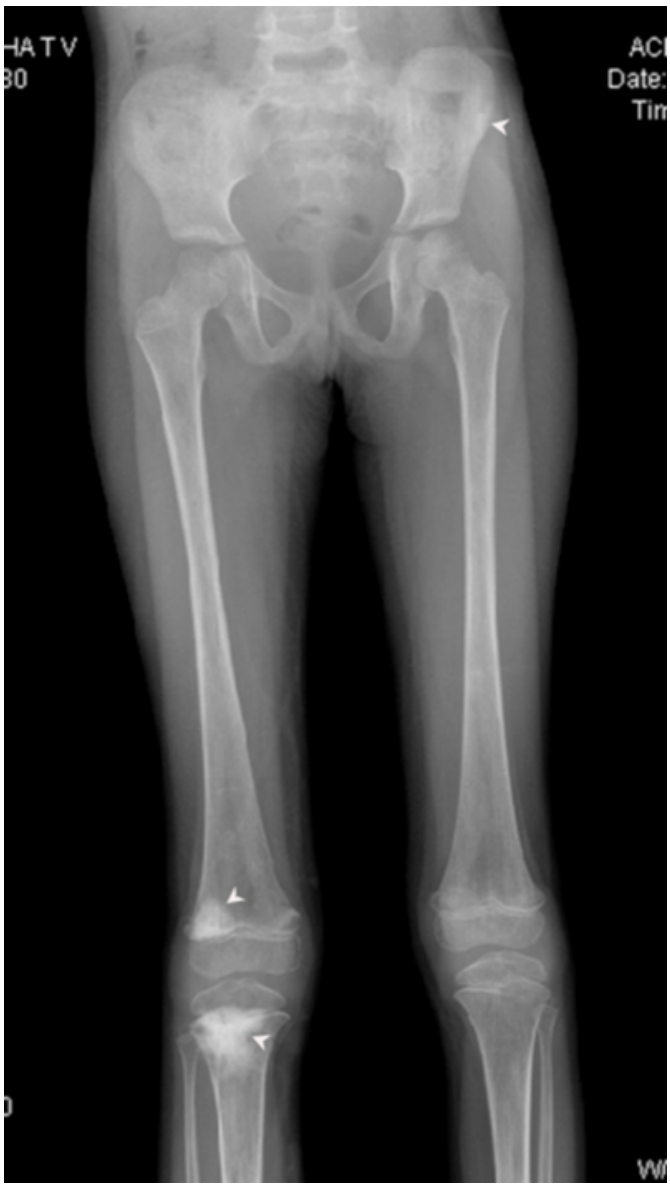


Figure 3

Fig 2b, Radiograph of the left leg, frontal view

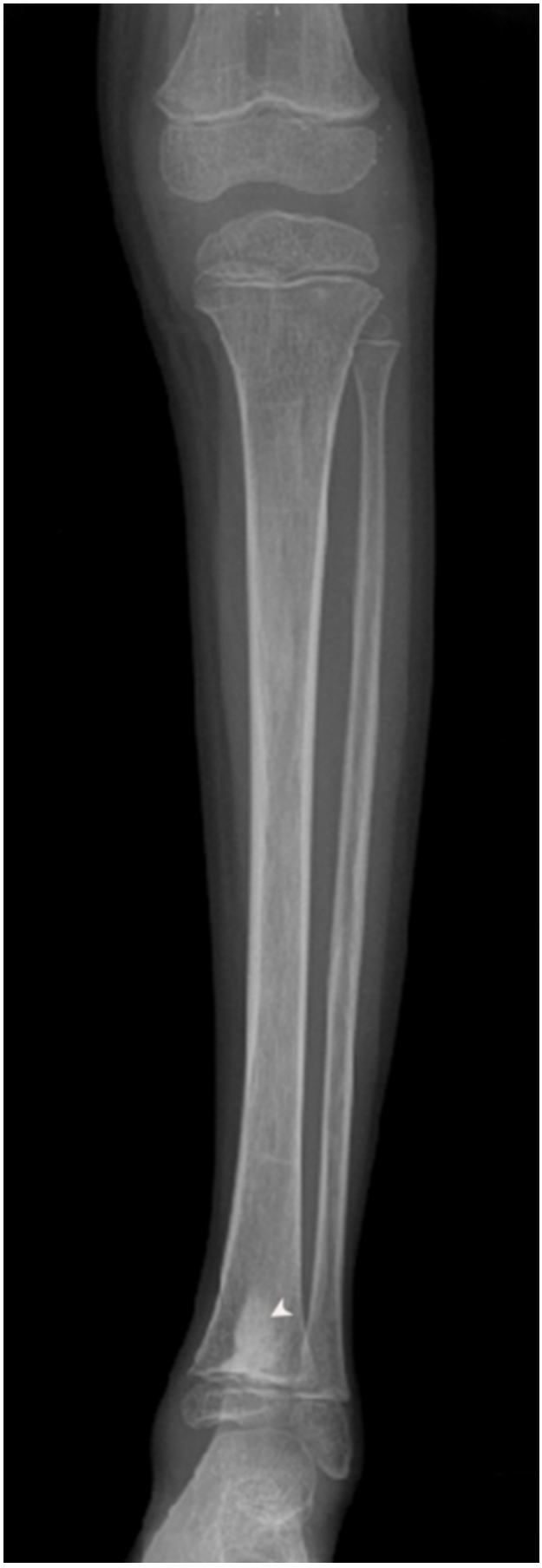


Figure 4

Fig 2c Radiograph of the chest, frontal view

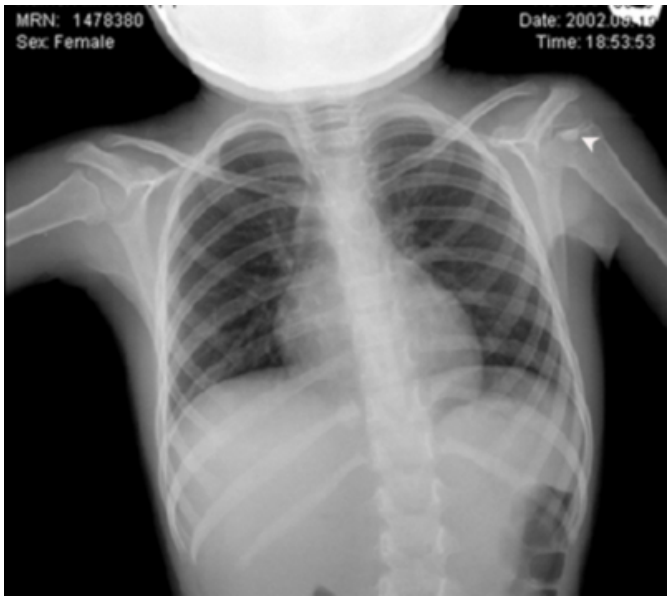


Fig 2a, 2b, 2c: Multiple sclerotic foci in the metaphyseal ends of multiple bones. Lungs are unremarkable.

Bone scan shows increased uptake corresponding to sclerotic areas on radiographs.

Tissue biopsy from the dominant lesion revealed osteogenic sarcoma.

DISCUSSION

The differential diagnosis of the multiple sclerotic metaphyseal lesions of the long bones would include:

Osteosarcomatosis or multiple osteosarcomas occurring synchronously (ie. Within 5 months) in children or adults.

Multiple lesions that occur metachronously (ie. After 5 months)

Unicentric lesion with second neoplastic lesion developing in the same bone or adjacent bone due to transarticular spread (skip metastasis) or metastasis to the distant bone with or without pulmonary metastasis.

Osteosarcomatosis is considered a rare distinct form of the disease that predominates in children. It is characterized by the presence of multiple sclerotic lesions which are distributed bilaterally, usually at the metaphysis of the long bones with or without a dominant lesion. It could be due to multicentric in origin or unicentric origin with multiple intraosseous metastases. The similarity in size, radiological features and histology, absence of dominant lesion and lack of pulmonary metastasis favors multicentric origin of these lesions. Presence of dominant lesion and pulmonary metastasis favors unicentric origin with metastasis. Serum alkaline phosphatase levels and lactate dehyogenase levels are useful in following the clinical evolution of multifocal osteosarcoma[3].

CLASSIFICATION SYSTEMS

AMSTUTZ classification[4]:

Multiple synchronous lesions occurring in <18 years

Multiple synchronous lesions occurring in >18 years

a. Early metachronous lesions (5-24 months)

b. Late metachronous lesions (>24 months)

MAHONEY CLASSIFICATION:

Childhood / adolescent synchronous metaphyseal lesions

Adult low grade synchronous lesions

Early metachronous lesions

Late metachronous lesions

Lowbeer[5] classified multifocal osteosarcoma into two groups: A-Childhood synchronous and B-adult metachronous.

Among these AMSTUTZ classification appears to be widely followed.

When dominant lesion is present, it presents as a large aggressive lesion with periosteal reaction, cortical disruption and soft tissue extension. Usual sites are the metaphysis of the long bones. Vascular and articular extension suggestive of high biological aggressiveness is also common. The secondary bone foci are smaller, more sclerotic, more well-defined and without cortical disruption or periosteal reaction. Our present case exemplifies this variety. The pulmonary metastases do not precede the appearance of secondary skeletal foci and are usually picked up on CT.

These biologically aggressive tumors have uniformly poor prognosis. Response to chemotherapy is poor and death usually occurs within 2 years[3]. As the osseous lesions of multifocal osteosarcoma are sclerotic, documentation of successful chemotherapy or radiotherapy may be difficult to document on radiographs. Thus radiographically stable disease may represent satisfactory response if there is no change in the size or number of lesions[3].

Multiple synchronous lesions in children and adolescents (AMSTUTZ type 1) have more secondary skeletal foci, commonly symmetrical and greater localization in the metaphysis.

Multiple synchronous lesions in adults (AMSTUTZ type 2) are less in number and are not symmetrical. These are low grade tumors and have a better prognosis.

Multiple metachronous lesions (AMSTUTZ type 3) usually affect adolescent and young adults. Usually presents as one or more tumors after treatment of the primary osteosarcoma. They are symmetrical, vary in size and may not necessarily be sclerotic. They represent new primary tumor or metastasis. Osseous metastases occur in 10-20% of all osteogenic sarcomas. These secondary lesions predominate in the spine and the pelvic bones. Multiple osteogenic sarcomas can also occur in paget's disease and following irradiation.

References

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