Case Report

Synchronous multifocal osteogenic sarcoma on multimodality imaging including bone scintigraphy

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ABSTRACT

Multifocal osteosarcoma is diagnosed when there are two or more lesions in the skeleton without presence of pulmonary metastases. It is further classified as synchronous type when the patient is demonstrated to have more than one lesion simultaneously at presentation and is known as Synchronous Multifocal Osteogenicsarcoma (MOGS). We report a case of synchronous MOGS showing its multimodality imaging findings including nuclear scan findings with pathological correlation.

Keywords: Metachronous, multifocal/multicentric, osteosarcoma, synchronous, MOGS

INTRODUTION

Osteosarcoma is the most common primary bone sarcoma representing about 25-35% of all cases but constituting only about 0.1% of all neoplasms. It is an aggressive tumor with tendency for local invasion and early metastases. A small subset of patients with osteosarcoma may have involvement of multiple skeletal sites without evidence of visceral metastases. This condition is known as multifocal osteogenicsarcoma (MOGS).[2,7] The lesions may present simultaneously (synchronous) or sequentially (metachronous). MOGS is a rare entity representing only 1-2% of all osteosarcomas. Less than 100 cases of synchronous type of MOGS have been reported in the literature till date.[6,8,9]

CASE REPORT

A 15-year-old male patient presented with 2-month history of gradually progressive pain and swelling around right shoulder and knee. In addition, there was complaint of anorexia and weight loss. No history of recent trauma was present. On palpation ill-defined swelling with focal tenderness was noted at both sites. Biochemical investigations revealed significantly elevated serum alkaline phosphatase and lactate dehydrogenase.

Patient was referred to our department for further imaging evaluation. Anteroposterior (AP) radiograph of the right shoulder showed a diffuse homogenous radiodense lesion with wide zone of transition involving nearly entire upper half of shaft of humerus including the epiphysis and diaphysis. Solid periosteal reaction was noted on either side of proximal part of the lesion [Figure 1a,b and c]. AP radiograph of the bilateral knee revealed multiple well-defined osteosclerotic lesions of varying sizes involving the metaphysis of distal femur and proximal tibia [Figure 1a and b]. An osteosclerotic lesion is also noted in proximal part of right femur in AP radiograph of pelvis with both hips [Figure 1d]. Chest X-ray was unremarkable. A provisional diagnosis of osteosarcoma with systemic metastases was made.

To establish the presence of suspected pulmonary lesions, nonenhanced computed tomography (NECT) scan of thorax was performed on a 16-slice computed tomography (CT) scanner (General Electric, Brightspeed, Milwaukee, Wisconsin, USA) from the thoracic inlet to lowermost part of diaphragm. No evidence of any suspicious lesion was noted in the pulmonary parenchyma [Figure 2c and d]. NECT bone window images demonstrated the shoulder lesion extending upto lower half of humerus and punctuate character of lesions involving right knee [Figure 2a and b].

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For better determination of the skeletal involvement, nonenhanced and contrast-enhanced magnetic resonance (MR) imaging of the right shoulder, right knee, bilateral hip, and lumbar spine was performed on 1.5 Tesla scanner (Siemens, Magnetom Essenza, Erlangen, Germany). T1-weighted images of right shoulder demonstrate replacement of normal bright marrow signal by a diffuse ill-defined hypointense lesion in the upper part of the humerus and associated extramedullary component. Post-contrast T1 fat-suppressed images revealed patchy intramedullary enhancement with marginal enhancement of extramedullary component. No evidence of intra-articular extension is noted [Figure 3b]. T1- and T2-weighted images of right knee showed multiple hypointense lesions (few of which appear confluent) involving either side of joint. Post-contrast proton density fat-saturated images demonstrate multiple peripheral enhancing lesions and patchy marrow enhancement. No soft tissue or intra-articular extension is demonstrated [Figure 3a].

T1-weighted images of bilateral hip and T2-weighted images of lumbar spine revealed hypointense lesion involving head of femur, posterior wall of acetabulum [Figure 4a and b], and L5 vertebral body, respectively [Figure 4c]. No soft tissue or intra-articular extension is noted.

Whole body bone scintigraphy using 99 m technetium-methylene diphosphonate (MDP) revealed intense tracer uptake involving right humeral head, bilateral femur (lower end) and tibia (upper end), and right femoral head. Few additional small areas of uptake

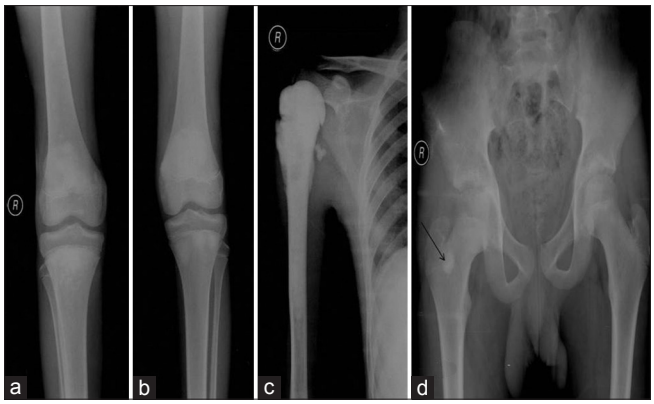

Figure 1: A 15-year-old child with synchronous MOGS, (a and b) AP radiograph of the bilateral knee showed multiple osteosclerotic lesions involving the metaphysis of distal femur and proximal tibia (arrows). (c) AP radiograph of right shoulder showed extensive osteosclerosis involving upper half of humerus, including the epiphysis with solid periosteal reaction on either side of the lesion. (d) AP view of pelvis with both hip showed an osteosclerotic lesion in proximal part of right femur (black arrow). MOGS = Multifocal osteogenicsarcoma, AP = Anteroposterior

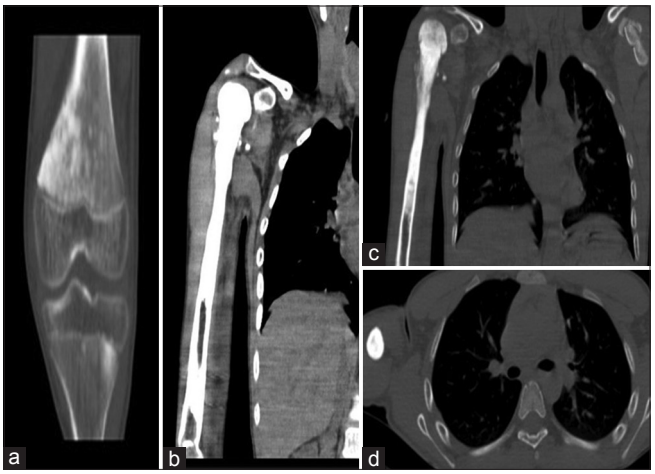

Figure 2: A 15-year-old child with synchronous MOS, (a) NECT right knee coronal image in bone window revealed the punctuate character of lesions involving bilateral knee. (b) NECT bone window image of right shoulder demonstrated the diffuse osteosclerosis involving humerus extending up to its lower half and associated periosteal reaction. (c) NECT thorax coronal and axial images revealed no calcified metastatic lesion in the thorax. MOGS = Multifocal osteogenicsarcoma, NECT = Nonenhanced computed tomography

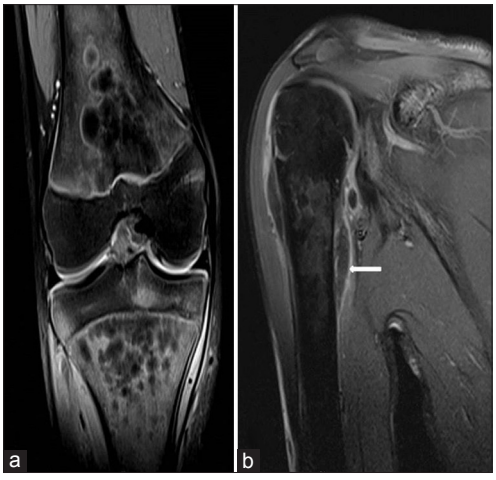

Figure 3: A 15-year-old child with synchronous MOGS, (a) post-contrast proton density-weighted fat-saturated coronal MR image of right knee demonstrated multiple peripheral enhancing lesions and patchy intramedullary enhancement involving lower end of femur and upper end of tibia. (b) T1-weighted fat-saturated post-contrast coronal MR image of right shoulder showed heterogenous intramedullary enhancement with marginal enhancement of extramedullary component (arrow). No evidence of intra-articular extension is noted. MOGS = Multifocal osteogenicsarcoma, MR = Magnetic resonance

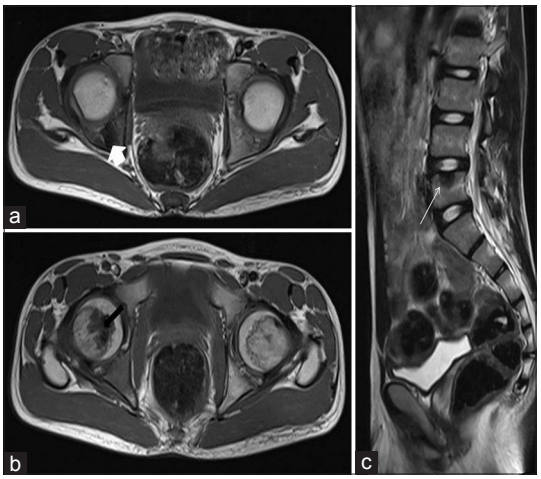

Figure 4: A 15-year-old child with synchronous MOGS-non-contrast T1-weighted axial MR images of bilateral hip (a and b) and T2-weighted sagittal images of lumbar spine (c) revealed hypointense lesion (black arrow) involving head of femur, posterior wall of acetabulum (white arrow), and L5 vertebral body (white line arrow), respectively. MOGS = Multifocal osteogenicsarcoma, MR = Magnetic resonance

are noted in the skeleton, including ribs. The sites of uptake in ribs may be confused with pulmonary involvement, however, the same can be ruled out on careful evaluation of images, especially oblique views [Figure 5].

Biopsy from the shoulder lesion (hematoxylin and eosin stain [H and E] stain ×10 and ×40) showed areas of amorphous chondroid matrix with chondrocytes and chondroblasts. There is evidence of lacy and disorganized basophilic osteoid matrix with cells showing high nucleocytoplasmic ratio and hyperchromasia. Above findings suggested chondroblastic type of osteosarcoma [Figure 6].

Patient was started on combination chemotherapy of methotrexate, pirarubicin, and ifosfamide. The serum calcium levels returned to baseline, but the clinical symptoms were persistent till the last follow up.

DISCUSSION

MOGS is exclusively diagnosed in patients with multiskeletal involvement and no pulmonary involvement because most of osteosarcomas harbor pulmonary micrometastases at the time of diagnosis. MOGS is divided into two types on the basis of pattern of presentation-synchronous and metachronous.[1,6]

The majority of lesions in patients with synchronous MOGS mimic skeletal metastases on radiology. There is at least one lesion with features suggestive of a primary osteosarcoma, with remaining lesions more suggestive of metastases. The metastatic lesions appear as purely sclerotic or heavily mineralized metaphyseal lesions with a narrow transition zone, no evidence of cortical destruction or soft tissue mass, or malignant periosteal new bone formation.[4,7]

In contrast, most of lesions in patients with metachronous MOGS demonstrate imaging features typical of a primary malignant sarcoma of bone including an aggressive mixed lytic and sclerotic pattern of bone destruction, cortical breech, soft tissue extension, wide zone of transition, and malignant periosteal new bone formation.[7]

There are two hypotheses for the pathogenesis of MOGS according to synchronous and metachronous types, respectively: (a) multisite lesions arising simultaneously, presumably all representing multiple independent synchronous primary lesions, and (b) single-site origin, with one dominant site and then early and rapidly progressive metastatic disease.[2,3,7]

Overall, osteosarcomas with multifocal skeletal lesions, especially the synchronous variety, have poorer prognosis compared with unifocal osteosarcoma with skeletal metastases. Magnetic resonance (MR) and CT imaging are superior for assessment of anatomic extent of primary skeletal lesions. However, an accurate assessment of the metastatic disease and metabolic activity of the primary lesions requires a radionuclide study. Whole body bone scintigraphy using 99m technetium-MDP is an ideal technique for this purpose, as it has a high sensitivity in staging metastatic disease. Absence of the lesions in the lung fields in the presence of multiple bony lesions confirms the diagnosis of MOGS differentiating it from a primary osteosarcoma with multiple metastases. In addition, baseline pretreatment MDP scan helps to assess the effectiveness of the therapy when compared with post-therapy scan, even if the lesions appear similar on cross-sectional imaging.[3,11]

The most commonly used classification of MOGS is by Amstutz; however, there appears to be no universal agreement on it.[5,9] From a pure pathological point of view, conventional osteosarcoma has three subdivisions-osteoblastic, chondroblastic, and fibroblastic. In practice, however, most osteosarcomas show varying amounts of three cell types and matrix. The division means to signify greater than 50% predominance of any histologic type. From management point of view, this separation is artificial because treatment for all types is same

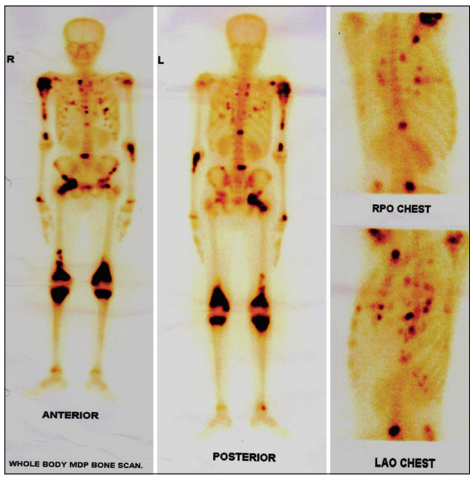

Figure 5: A 15-year-old child with synchronous MOGS-whole body bone 99m technetium-MDP scintigraphy revealed intense tracer uptake involving multiple skeletal sites. Few sites of uptake were also noted in ribs, which mimic pulmonary metastasis, however, the same was ruled out on oblique views. MOGS = Multifocal osteogenicsarcoma, MDP = Methylene diphosphonate

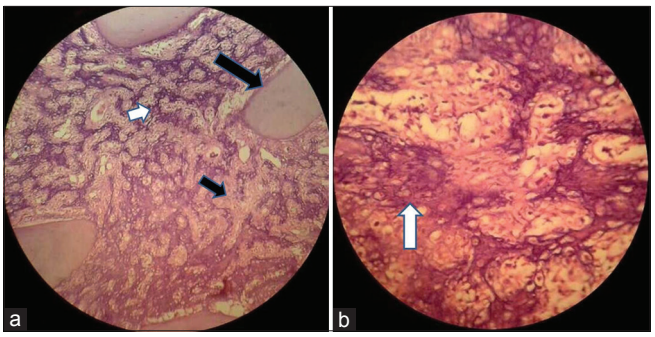

Figure 6: A 15-year-old child with synchronous MOGS-histopathological examination of the right shoulder lesion (H and E stain ×10) (a) and (H and E stain ×40) (b) showed lacy disorganized basophilic malignant osteoid matrix [small white arrow in (a) and large white arrow in (b)] with osteoblastic cells showing high nucleocytoplasmic ratio and hyperchromasia, areas of amorphous chondroid matrix with chondrocytes and chondroblasts (large black arrow in (a)) in a background of eosinophilic stroma (small black arrow). Above findings suggested chondroblastic type of osteosarcoma. MOGS = Multifocal osteogenicsarcoma, H and E = hematoxylin and eosin stain

and there is no statistical difference in survival of patients with high-grade tumors of three histologic types. All synchronous and metachronous MOGS are high-grade (grade 3 and 4) medullary osteosarcomas. Most of them are of osteoblastic subtype with chondroblastic variety being the least common. Grade 4 histology is far more common than grade 3. MOGS are almost never low-grade tumors (grade 1 and 2). Synchronous MOGS typically shows the same histopathological subtype in all multiple tumor locations, in contrast to presence of atleast one lesion characterized by a different histopathological subtype compared with the primary osteosarcoma in metachronous group.[7,10]

CONCLUSION

Synchronous M-GOS can be confused with primary osteosarcoma with diffuse metastases. The distinction between the two entities is possible on the basis of evidence of complete absence of pulmonary lesions in synchronous M-OGS. Initial screening with NECT and confirmatory MDP bone scan is essential to demonstrate this. The clinical distinction has significant prognostic importance, as synchronous M-GOS has an extremely poor prognosis.

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