



Diaphyseal sclerotic osteosarcoma presenting as a pathological fracture

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A diaphyseal, intramedullary, highly sclerotic lesion presenting as a pathological fracture, without a periosteal reaction or an appreciable soft tissue component on radiographs was investigated. A discrepancy between the MRI and histopathological findings led to marginal excision of the lesion only to reveal later that it was a sclerotic variety of osteosarcoma. Such a presentation has not been reported in literature as per our knowledge. We forfeited the opportunity of limb salvage by doing initial marginal excision and fixation. In such circumstances, a representative biopsy is critical and repeat biopsy is warranted before going for definitive management.

Keywords : atypical osteosarcoma ; pathological fracture ; representative biopsy.

INTRODUCTION

Atypical presentation of osteosarcoma is not rare (3,2,6). The literature over the years has tried to encompass all the variants, and several authors have attempted to describe all these various presentations (3,2,6). The current case is another addition to this ever growing list. The purpose of this case report is to reiterate the variability of presentation of osteosarcoma leading to diagnostic delays and therapeutic blunders precluding simpler management which may be possible only in the first instance.

Diaphyseal osteosarcomas are rare (5). Highly sclerotic osteosarcomas are rarer still (6) and it is

very unlikely that a sclerotic variety of osteosarcoma presents with a pathological fracture (1). A diaphyseal, non expansile, sclerotic osteosarcoma with no periosteal reaction and presenting with a pathological fracture has not been described in the past as per our knowledge.

CASE REPORT

A 17-year-old female student sustained a trivial trauma (simple fall from the standing position) and was unable to get up or bear weight on her left lower limb. There was severe pain in the left thigh. There was no history of any symptom in the left thigh or hip region in the past. She was immediately taken to a local hospital where a closed fracture of the middle third of the shaft of the femur was

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Fig. 1. — Anteroposterior and lateral radiograph of the diaphyseal lesion on the 3rd day after sustaining trivial trauma. Note the intramedullary sclerotic lesion on either side of the fracture site without obvious periosteal reaction or a soft tissue component.

diagnosed, and closed reduction with internal fixation using an interlocking nail was planned. However on the operating table, the reamer could not be negotiated through the proximal fragment. The procedure was abandoned and the patient was referred to our centre, 3 days after sustaining injury. On arrival, she had an upper tibial skeletal traction on a Böhler Braun splint applied outside. The anteroposterior and lateral radiograph (fig 1) revealed a transverse fracture of the middle third of the shaft of femur. There was a uniformly sclerotic lesion occupying the medullary canal of both the proximal and distal fragments and extending 4 centimeters on either side of the fracture site. The sclerotic lesion was equally radiopaque as the surrounding cortex and continuous with it. There was no periosteal reaction. There were no lytic lesions either within the confines of the sclerotic lesion or outside it. Tc^{99m} bone scan revealed a moderately

increased uptake limited to the fracture site. There was no sign of increased activity anywhere else in the skeletal system. MRI (fig 2) showed a hypointense signal of the marrow at the fracture site on T₁W and T₂W images indicating osteosclerosis. There was some soft tissue component of the lesion appreciable on the sagittal and axial T₁W sections within the medullary canal and just outside the bone. The postcontrast T₁W images through the same level showed moderate enhancement of the intra and extraosseous soft tissue indicating the possibility of a growth. There was oedema in the surrounding tissue consistent with a fracture.

The blood test failed to reveal any abnormality, with normal erythrocyte sedimentation rate and alkaline phosphatase levels. Computer Enhanced Computed Tomography (CECT) of chest and abdomen revealed no evidence of any metastasis or a primary lesion giving rise to bony secondaries.

A biopsy was done from the lesion. The histological examination (fig 3) of all the submitted material as multiple bony fragments showed exuberant fracture callus formation in the form of osteoid or woven bone lined predominantly by plump osteoblasts seen as regular seams. There was also some chondroid differentiation. There was in addition evidence of organized haemorrhage, dead bone fragments and soft tissue injury. The material represented fracture callus and no bone tumour could be identified. The slides were reviewed again for any evidence of malignancy in view of equivocal radiological findings, but it again failed to show any features suggestive of malignancy. Therefore an excisional biopsy along with definitive fracture fixation was planned.

The patient was taken up for surgery 14 days after injury. Approaching from the lateral side, the biopsy scar was excised. There was abundant callus formation at the fracture site. Marginal excision of the sclerotic lesion was done (leaving only a thin rim of the medial cortex) as there was a doubt about the fracture union in a sclerotic lesion. The lesion appeared uniformly sclerotic on gross appearance and cut sections. There was no separate soft tissue element of the lesion apparent, because of exuberant callus formation which was excised with the lesion. The fracture was fixed with a 12-hole low

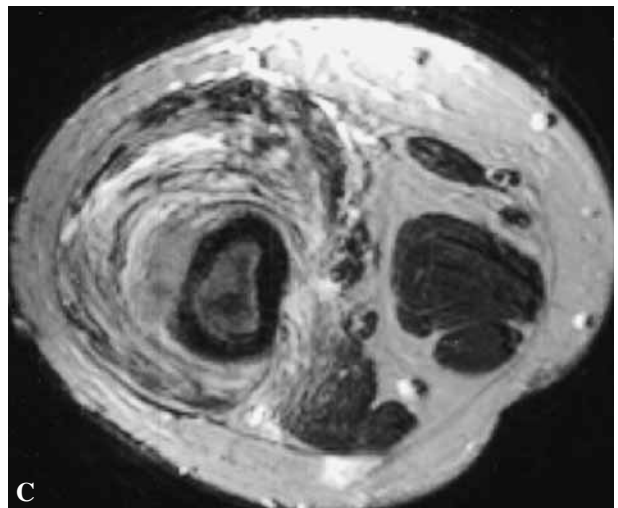
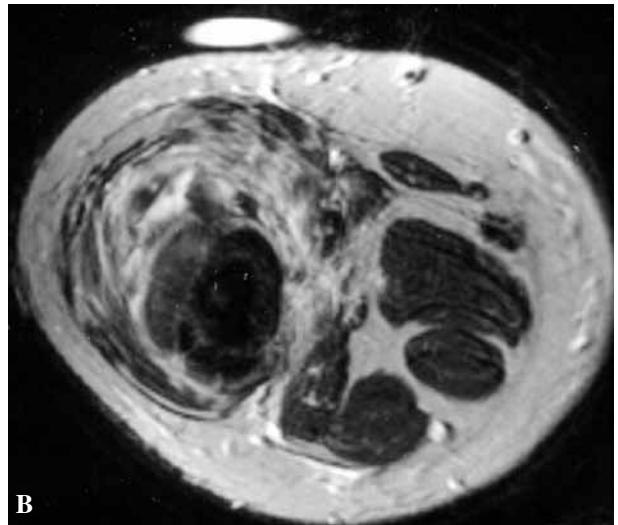
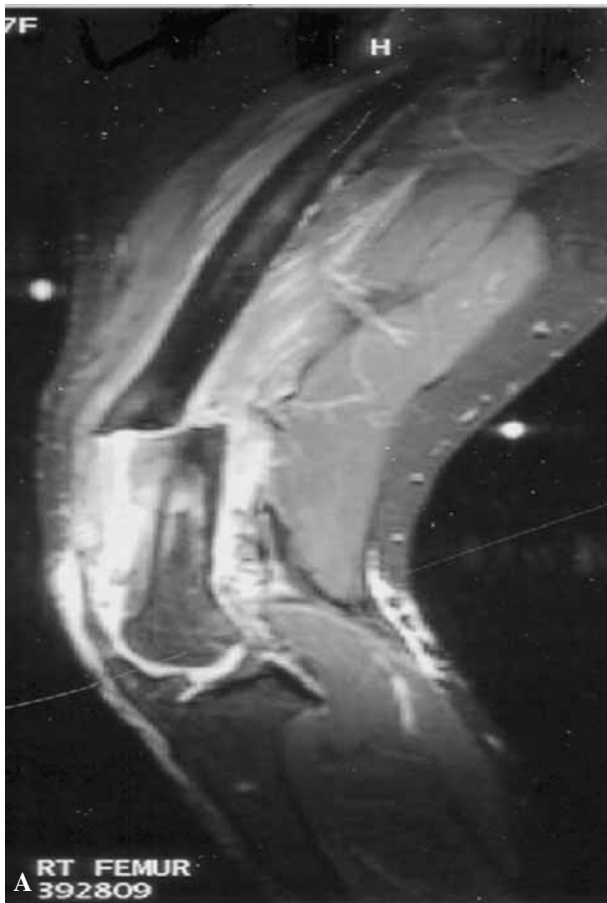


Fig. 2A, 2B & 2C. — Sagittal and axial sections of the femur on MRI showing the sclerotic nature of the lesion. The soft tissue component of the lesion can be better appreciated on the axial sections.

contact dynamic compression plate (LCDCP). The resulting gap was adequately filled with long strut grafts from the iliac crest and the graft was held by a circlage wire. The entire specimen excised was sent for histopathological examination. The sections showed again more mature fracture callus.

On telephonic conversation with the patient 15 days after surgery, she complained of persistence of pain in the left thigh. The pain was present even at rest. So, after a discussion with the pathologist (R.K.V) and the radiologist (V.G) a decision to analyse the remaining portions of the specimen was made. Embedding of all the left over fragments revealed two histological sections that showed infiltration of trabecular bone by masses and irregular sheets of plump, anaplastic, large, occasionally epitheloid-like cells with 3 to 4 nucleoli and few mitotic figures. However there was no necrosis. A large amount of osteoid and woven bone was also

seen. Therefore a diagnosis of osteosclerotic type of osteogenic sarcoma was made (fig 4).

The patient was immediately called for follow-up evaluation. The plain radiograph done 3 weeks after excision and fixation showed a large soft tissue component at the site of the previous fracture, measuring 8 cm × 8 cm (fig 5). The lesion was predominantly sclerotic. CECT of the chest and abdomen along with whole body bone scan was repeated and it revealed no secondaries. In this scenario of an extracompartmental lesion with

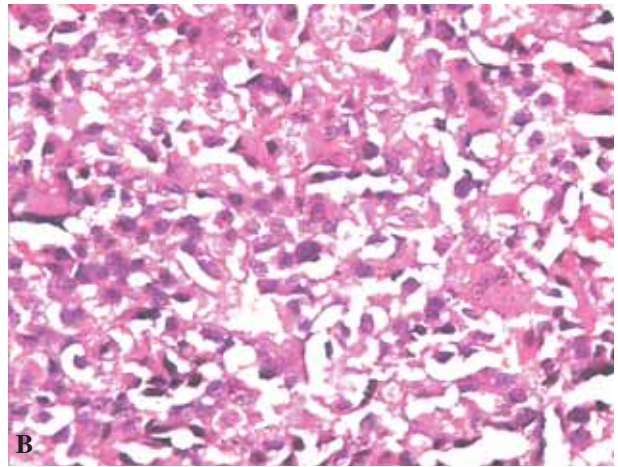
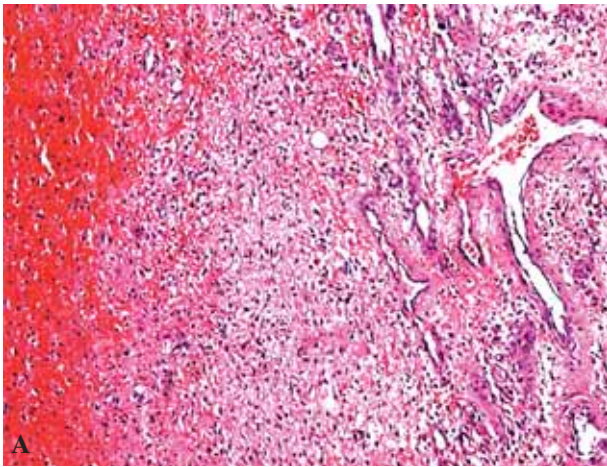


Fig. 3A & 3B. — Low power and high power field view of the first histopathological slide showing exuberant callus formation with osteoid seams. There are no features suggestive of malignancy.

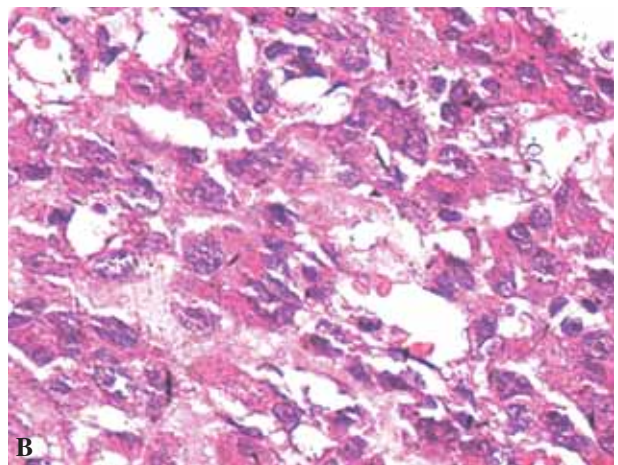
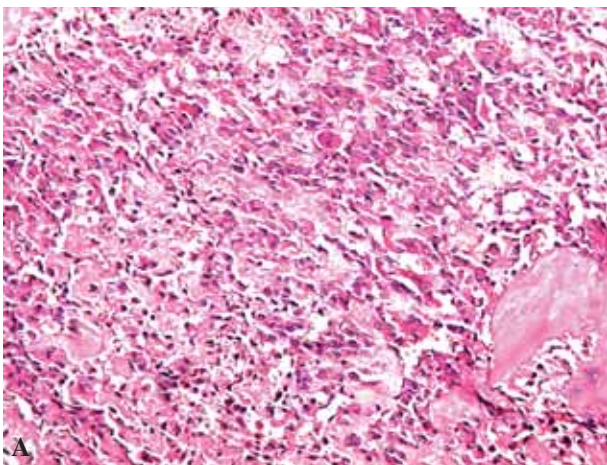


Fig. 4A & 4B. — Low power and high power field view of the final histopathological slide showing features suggestive of osteosarcoma. Note the mitotic figures and predominant osteoid.

marginal excision being done previously, wide margin excision was planned. Before surgery, the patient was given 3 cycles of adjuvant chemotherapy. In each cycle of 3 weeks duration, Cisplatin 100 mg/m² body surface area was given on the first day and Adriamycin 25 mg/m² body surface area was given on the first three days. The hip was disarticulated for wide resection after 3 cycles of chemotherapy. Postoperatively another 3 cycles of chemotherapy was repeated with the same drugs. The patient is under our regular follow-up for 18 months now and is currently asymptomatic.

DISCUSSION

Diaphyseal osteosarcomas constitute 9% of all osteosarcomas (5). Haworth *et al* (5) have analysed 15 cases of diaphyseal osteosarcomas presenting over a 26 year period and classified the radiological features into 4 distinct patterns. Our case does not fit the description of any of the 4 patterns described. The closest comparison would be the group II (Atypical sclerotic) pattern but unlike their cases our case does not have cortical expansion and has presented with a pathological fracture. However



Fig. 5. — Anteroposterior and lateral radiograph of the lesion 3 weeks after excision biopsy and fixation. Note the exuberant growth with typical ‘fluffy’ and ‘cumulus cloud’ like densities in the soft tissue measuring 8×8 cm.

similar to their case, there is no periosteal reaction, the lesion is sclerotic and medullary in location without any lytic component. The biopsies were similarly inconclusive initially in their case, as in our case.

Mirra (6) describing the radiology of the pure sclerosing variant of osteosarcoma opines that the lesions are homogenous masses of ‘fluffy’ or ‘cumulus cloud like’ densities. He also describes fine radiation along the periphery of these lesions which is not present in our case. He hints the clues to the diagnosis as (i) focal nature of the lesion in a single bone (unlike osteopetrosis) (ii) usual metaphyseal location and (iii) focal periosteal reaction of some kind. Our case had no periosteal reaction and was not metaphyseal.

If this lesion were to be detected before the occurrence of a pathological fracture, one would have observed it rather than prophylactically fixing it, as per ‘Mirel’s criteria for the risk of pathological fracture (7). This observation is to emphasize that painless purely blastic lesions are at a much lesser risk of pathological fracture unlike the presented case.

The possibility of an intramedullary bone island or enostosis was kept as the differential diagnosis based on the radiographic picture (4). These are usually metaphyseal and very rarely found in the diaphysis. However the soft tissue component on the MRI negates the diagnosis of a bone island.

The decision of marginal excision of the lesion needs debate. Marginal excision was done because histopathology clearly excluded any possibility of a malignancy in the first instance. There was an element of doubt about the quality of apposing bone surfaces at the fracture site in giving us union and hence excision of the sclerotic area was done.

The possibility of sampling error cannot be ruled out. The fracture with organized haematoma / soft callus in this young female made the sampling of the underlying real lesion difficult, resulting in a delay in the diagnosis. Also exuberant callus can notoriously mask a malignant lesion. Only few sections of the excised lesion in the second surgery showed unequivocal evidence of a highly differentiated sclerotic type of osteogenic sarcoma. In such atypical cases it is better to plan larger areas of excision to get representative biopsy.

Osteosarcoma can evade diagnosis by presenting atypically. MRI is known to be very helpful in such instances in diagnosing malignant lesions (1). In the present case the discrepancy between the radiological and histopathological findings in the first instance, delayed the definitive treatment. As clinicians, when there is an element of doubt in such cases, with non-correlating radiological and histopathological findings, one should not rush to treat till a definitive diagnosis is reached. We shall otherwise forfeit the opportunity to use appropriate treatment modalities, and hence limb salvage. On the other hand undue delay is also detrimental, in that the lesion will continue to grow, breach barriers and warrant wider excision. In such circumstances,

a larger area of excision for a representative biopsy gives a more definite answer to the problem.

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