The many faces of intraosseous haemangiomata: a diagnostic headache

ABSTRACT
Intraosseous haemangiomata constitutes less than ten percent of all primary bone neoplasms. Approximately 75 percent occur in the calvarium or vertebral, with long bones, short tubular bones and ribs constituting the rest. We describe a 52-year-old woman who presented with left knee pain for 4–5 years and loss of weight over one week. An initial radiograph of the knee showed several well circumscribed isodense lesions with sclerotic rims in the medullary cavity of the distal femur and diaphysis of the left tibia. There were also lucent lesions with a slightly sclerotic rim in the diaphysis of the left tibia and proximal left fibula. In view of the clinical presentation and radiological findings, extensive investigations were made to rule out metastases and multiple myeloma. An open biopsy with segmental osteotomy of the left mid fibular lesion revealed an intraosseous haemangiomata.

Keywords: bone haemangioma, intraosseous haemangiomata, skeletal angiomatosis, vascular malformation, vascular anomaly

INTRODUCTION
Haemangiomata of the bone may occur in any age group, with approximately 25% of cases presenting in the fifth decade of life. It is commonly found in the vertebral, followed by the calvarium, and is uncommonly seen in the long bones, tubular bones or ribs. The classic corduroy and sunburst appearance of vertebral and skull haemangiomata are uncommon in extremity sites. The extremity site lesions may have a classic coarse trabecular bone pattern or soap bubble appearance. A permissive pattern of irregular bone destruction is less commonly seen. As there is a wide range of radiological patterns, accurate preoperative diagnosis of non-classical skeletal haemangiomata is difficult to make. Bone haemangiomata are usually asymptomatic, and either discovered incidentally or at autopsy. In our case, the patient presented with worsening knee pain and weight loss over one week duration with multiple lucent lesions with sclerotic margins in the tibia and fibula. Differentials of metastases and multiple myeloma were raised. After extensive investigation, open surgical biopsy with osteotomy of the fibula provided the final histological confirmation of an intraosseous haemangiomata.

CASE REPORT
A 52-year-old woman was referred to an orthopaedic surgeon for left lower limb pain for several years. She
had a left knee pain for 4–5 years which progressively worsened over the years, and resulted in impaired mobility. There was no other site of bone pain. She also complained of a loss of weight of over one week duration. Her past medical history included stage one membranous nephropathy and hypertension. Initial radiograph of the left knee showed two different types of lesions. The first type was isodense to the bone located within the medullary cavity, with a sclerotic margin (Fig. 1), at the epiphysis of the proximal tibia, within the distal femur and proximal tibia. The second type had central lucency with a sclerotic margin (Fig. 1) within the diaphysis of the left femur and proximal left fibula. In view of the clinical presentation of the patient, metastases were raised as a differential diagnosis.

Computed tomography (CT) of the brain, thorax, abdomen and pelvis was performed to search for a primary tumour. Non-specific lucencies were seen in the calvarium, and multiple ring-shaped lucencies with sclerotic borders were seen in the left distal femur and upper tibia, left femoral head extending to the juxta-articular region, left side of the ilium and L5 vertebral body (Fig. 2). A differential diagnosis of multiple myeloma and cystic angiomatosis was made. There were no abnormalities identified on CT to suggest a primary tumour. Screening mammography did not reveal any evidence of breast cancer. Serum carcinoembryonic antigen, alpha-foetoprotein, Ca 19.9 and Ca 125 were normal. Serum beta-2 microglobulin was normal. Myeloma studies, bone marrow aspirate and biopsy examination, flow cytometry of the marrow and myeloma immunophenotyping were all negative for multiple myeloma. A radiographical skeletal survey also showed slightly expansile intracortical lucency (Fig. 3) at the proximal left femur and within the left femoral head. Technetium (Tc)-99m methylene diphosphonate (MDP) bone scan showed foci of increased tracer uptake corresponding to the bony lesions. As the Tc-99m MDP bone scan is rather non-specific, and given the clinical suspicion, metastases remained a differential possibility as suggested in the report.

The decision was made to perform an open surgical biopsy with osteotomy of the left mid fibula. Microscopy revealed a conglomerate of thin-walled blood vessels with adjacent bony trabeculae. The vascular channels comprised dilated venous-calibre type and capillary-sized vessels that were lined by a single layer of cytologically bland endothelial cells, with flat to occasionally plump nuclei. There were no lymphoid aggregates in the stroma and vascular channels to suggest lymphangiomatosis. Intercellular tissue was composed of loose connective tissue with myxoid change, containing foci of lymphocytes, plasma cells and eosinophils. This benign vascular lesion replaced the intertrabecular marrow space. No granuloma, histiocytosis, or malignant cells were present. The features were consistent with haemangioma of the bone (Fig. 4). The patient was treated symptomatically for her knee pain and has been pain-free for one year.

**DISCUSSION**

Primary intraosseous haemangioma is an uncommon bone tumour accounting for less than 1% of bone tumours, with 75% of cases found in the vertebral and calvarium. Other sites like the long bones, short tubular bones and ribs constitute the rest. It occurs more commonly in females than males, with a ratio of 3:2. Bone haemangioma are usually solitary but up to one-third of cases can have multiple lesions. Bone haemangioma usually occur in the medullary cavity and, less commonly, in the cortex, periosteum and subperiosteal regions. In our case, there were both intramedullary and intracortical lesions. In the long bones, the lesions are usually located in the metaphysis or diaphysis, while in our case there were lesions in the epiphysis, metaphysis and diaphysis. There has been only one reported case of the epiphysseal location of haemangioma in the femoral head in the literature, and this would be considered as a second known reported case in the literature. There is a propensity to extend
from the bone into the surrounding tissue or vice versa. There have been various radiological patterns that have been described, including a sunburst appearance for skull haemangioma and a honeycomb appearance for rib haemangioma.\(^7\)

The characteristic radiographical appearance of sclerotic or ivory vertebra with coarse thickened vertical trabeculae resulting in the corduroy-like appearance of the vertebra haemangioma has been described, but these features were not present in our case. Bulging of the posterior cortex or expansion of the vertebral body, which are sometimes seen, was also not present in our case. Radiographically, a radiolucent, slightly expansile and well-defined intraosseous lesion with a radiating pattern is highly suggestive of haemangioma for extraspinal sites.\(^8\) In our case, there were both lucent and isodense lesions with sclerotic rims. Reactive sclerosis could be seen at the margins of the surface-based haemangioma, mimicking osteoid osteoma. This diagnosis would have been considered if it was an isolated intracortical lesion. On axial CT, vertebral body lesions would have a typical polka dot pattern as the thickened vertical trabeculae are seen in cross-section as small punctuate areas of sclerosis; however, this was not present in our case. There were only ring-shaped lucencies with sclerotic rims on CT imaging of the vertebral body lesions. Haemangioma in the metaphysis or epiphysis of the long bones have been described as lytic lesions that exhibit a spiculated pattern known as Irish lace on CT scan, but these were also not present in our case.

On magnetic resonance (MR) imaging, typical signal characteristics for osseous haemangiomas are hypo- to hyperintense on T1-weighted sequence depending on the amount of adipose tissue, and hyperintense on T2-weighted sequence due to vascularity. Vertebral haemangioma demonstrate typical hypointensity on MR imaging for all sequences for the thickened vertical trabeculae portion, and hyperintensity on T1- and T2-weighted sequences due to the presence of intratumoural fat. Extrasosseous components usually do not show a high signal intensity on T1-weighted sequence as there is a paucity of adipose tissue, but there is avid enhancement on administration of intravenous gadolinium due to the vascularity of the lesions. MR imaging was not performed in this case as the patient had already agreed to an open surgical biopsy for definitive diagnosis. Osseous haemangioma usually show normal uptake on Tc-99m MDP bone scans, though in some cases they may demonstrate photopenia and mild to moderate increased activity. In this case, a bone scan showed the lesions to have non-specific mild to moderate increased tracer uptake.

What has been a diagnostic headache in this case was that none of the typically-described radiological appearances, like sunburst or corduroy appearance, was present. The sclerotic or ivory vertebra described on radiographs of vertebral haemangioma was not present. Ring-shaped lucencies that are atypical on CT were seen instead of the typical polka dot pattern. The thoracic spine is the most common location for vertebral haemangioma, but in our case, they were mainly located in the lumbosacral spine. Neither did the presence of lucent and isodense lesions with sclerotic rims in the epiphysis, metaphysis and diaphysis and occurring in both intramedullary and intracortical locations help in the diagnosis of the lesions. The diagnosis of multiple myeloma was excluded based on the various serum, marrow aspirate and biopsy results. CT of the brain, thorax, abdomen and pelvis did not reveal a primary organ lesion or malignancy. Metastatic disease was initially considered as a differential diagnosis, but given the relatively long duration of pain over several
years and lack of destructive features on radiograph and CT, this was unlikely.

Cystic angiomatosis was listed as a differential diagnosis in the initial workup of this patient. Vigorita summarised Devaney et al’s array of syndromes and termed “angiomatosis” to include lesions restricted to the skeleton as cystic angiomatosis of the bone while multifocal or multifocal involvement by a benign vascular lesion involving both bone and extrasosseus tissues as skeletal-extraskeletal angiomatosis. However, other authors have defined cystic angiomatosis as disseminated multifocal haemangioma and/or lymphangiomatous lesions of the skeleton with possible visceral organ involvement. It has also been quoted that in cystic angiomatosis, 60%–70% of cases show visceral organ involvement. Cystic angiomatosis is also often found in patients 10–30 years of age and more frequently in men than in women. At the more severe end of the spectrum of angiomatosis is Gorham’s syndrome, which was previously termed “massive osteolysis” or “vanishing bone disease”. In our case, it was shown that bone haemangioma can occur in various forms throughout the vertebra and long bones. They may not possess any of the known descriptors and it has to be considered as a possible differential diagnosis in a patient with multiple lucent or isodense lesions with sclerotic rim. They can be epiphyseal, metaphyseal or diaphyseal and intramedullary or intracortical in location.

REFERENCES