The many faces of intraosseous haemangioma: a diagnostic headache

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ABSTRACT

Intraosseous haemangioma constitutes less than ten percent of all primary bone neoplasms. Approximately 75 percent occur in the calvarium or vertebrae, 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 haemangioma.

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

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INTRODUCTION

Haemangioma of the bone may occur in any age group, with approximately 25% of cases presenting in the fifth decade of life.[1] It is commonly found in the vertebrae, 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 haemangioma are uncommon in extremity sites. The extremity site lesions may have a classic coarse trabecular bone pattern or soap bubble appearance. A permeative 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 haemangioma is difficult to make. Bone haemangiomas 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 haemangioma.

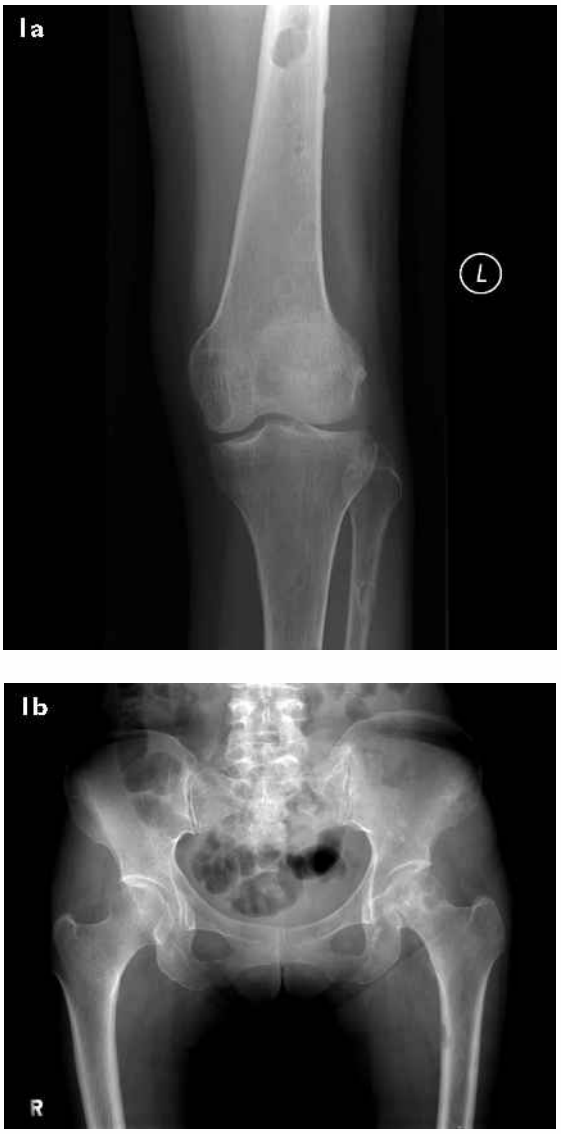

Fig. 1 (a) Anteroposterior radiograph of the left knee shows two different types of intramedullary lesions. There are isodense lesions with sclerotic rim at the epiphysis of the proximal tibia and hypodense lesion within the diaphysis of femur. (b) Anteroposterior radiograph of the pelvis shows intracortical lucencies with sclerotic rim in the proximal left femur and within the epiphysis of the left femur.

CASE REPORT

A 52-year-old woman was referred to an orthopaedic surgeon for left lower limb pain for several years. She

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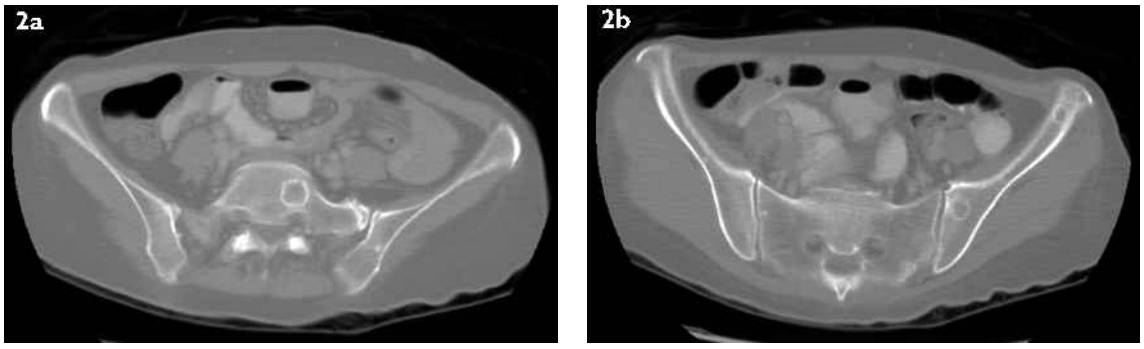

Fig. 2 (a) Axial CT image of the pelvis (bone setting) shows ring-shaped lucency with sclerotic rim within the L5 vertebral body. (b) Axial CT image of the pelvis (bone setting) shows two ring-shaped lucencies with sclerotic rim within the left iliac bone.

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,[2] with 75% of cases found in the vertebra[3] and calvarium.[4] 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.[5] 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 epiphyseal location of haemangioma in the femoral head in the literature,[6] and this would be considered as a second known reported case in the literature. There is a propensity to extend

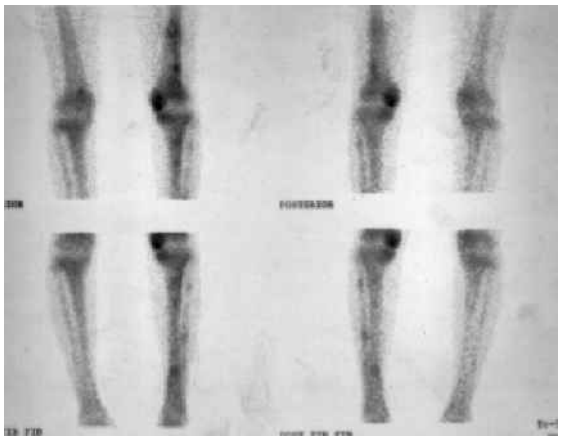

Fig. 3 Tc-99m MDP bone scan shows multiple areas of increased tracer uptake corresponding to the bony lesions. These were initially reported as possible metastases given the clinical presentation.

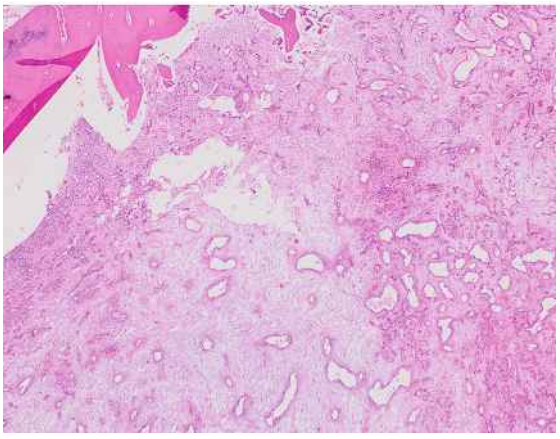

Fig. 4 Photomicrograph shows a conglomerate of thin-walled blood vessels with adjacent bony trabeculae. Vascular channels comprise dilated venous-calibre type and capillary-sized vessels. (Haematoxylin & eosin, × 2).

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. Extraosseous 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 unifocal or multifocal involvement by a benign vascular lesion involving both bone and extraosseous tissues as skeletal-extraskeletal angiomatosis.[9] However, other authors have defined cystic angiomatosis as disseminated multifocal haemangioma and/or lymphangiomatous lesions of the skeleton with possible visceral organ involvement.[4,10] It has also been quoted that in cystic angiomatosis, 60%–70% of cases show visceral organ involvement.[11] Cystic angiomatosis is also often found in patients 10–30 years of age and more frequently in men than in women.[10] 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 this non-familial syndrome, the proliferation of vascular tissue causes massive osteolysis of all or part of the bone and tends to involve one bone only. The aggressive bone destruction is not apparent in this case, and histologically, there is no permeation of cortical bone or soft tissue. Osteoclast-like cells resorbing bone at the margin of the vascular proliferation are likewise not present.

Lymphangiomatosis is usually found in association with polyostotic angiomatosis and must be differentiated on the basis of histological confirmation of thin-walled, widely-dilated irregular spaces lined by flattened endothelium, lumina containing proteinaceous fluid and lymphoid aggregates in the stroma. This was not present in our case. At histological analysis, cystic angiomatosis is indistinguishable from cavernous haemangioma, capillary haemangioma or lymphangioma.[12] The difference that the term "angiomatosis" connotes is that of a multicentric distribution of the vascular lesions.[6] In our case, there was multicentric distribution of the haemangioma of the bones with no extraskeletal involvement. Our patient was a 52-year-old woman with no visceral organ involvement or lymphangiomatous lesions, thus making cystic angiomatosis a debatable clinical diagnosis in this case, depending on which author's definition of the condition was used.

The decision was made for open diagnostic biopsy with an en-bloc specimen. It is known that histopathological diagnosis of a haemangioma from biopsy or tissue curettage can be challenging for the pathologist due to the destructive nature of these procedures. There will be disruption of the thin-walled blood vessels resulting in histological sections showing non-diagnostic empty spaces with scattered bone trabeculae. Four histological variants have been described, according to the predominant type of vascular channel: cavernous, capillary, arteriovenous and venous.[13] There are also mixed types as in our case: the bone haemangioma was a capillary and venous mixed type. Haemangioma is slow-growing and there is no known report of malignant degeneration. There have been cases of locally aggressive patterns mimicking malignant lesions.[14]

For most cases, masterly inactivity would be the most prudent choice, but in symptomatic cases, preoperative embolisation, surgery, percutaneous vertebroplasty or direct ethanol injection have been used. Calvarial cases tend to be the most clinically significant,[15] presenting as palpable lumps, while those with an aggressive growth pattern can present with pain. Haemangioma in the long bones may present with pain, limb hypertrophy or swelling. 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

1. Unni KK. Dahlin's Bone Tumors: General Aspects and Data on 11087 Cases. 5th ed. Philadelphia: Lippincott-Raven, 1996: 7.
2. Dorfman HD, Steiner GC, Jaffe HL. Vascular tumors of bone. Hum Pathol 1971; 2:349-76.
3. Osborn AJ. Tumours, cysts, and tumour-like lesions of the spine and spinal cord. In: Diagnostic Radiology. 3rd ed. St Louis: Mosby, 1994.
4. Murphey MD, Fairbairn KJ, Parman LM, et al. From the archives of the AFIP. Musculoskeletal angiomatous lesions: radiologic-pathologic correlation. Radiographics 1995; 15:893-917.
5. Devaney K, Vinh TN, Sweet DE. Surface-based hemangiomas of bone. A review of 11 cases. Clin Orthop Relat Res 1994; (300):233-40.
6. Chawla A, Singrakhia M, Maheshwari M, Modi N, Parmar H. Intraosseous haemangioma of the proximal femur: imaging findings. Br J Radiol 2006; 79:e64-6.
7. Dorfman HD, Czerniak B. Vascular lesions. In: Bone Tumors. St Loius: Mosby, 1997: 729-814.
8. Resnick D, Kyriakos M, Greenway GD. Tumors and tumor-like lesions of bone. In: Resnick D, ed. Diagnosis of Bone and Joint Disorders. 4th ed. Philadelphia: Saunders, 2002: 3979-85.
9. Vigorita VJ. Vascular and other mesenchymal tumours and lesions. In: Vigorita VJ. Orthopaedic Pathology. Philadelphia: Lippincott Williams & Wilkins, 1999: 389-98.
10. Lateur L, Simeons CJ, Gryspeerdt S, et al. Skeletal cystic angiomatosis. Skeletal Radiol 1996; 25:92-5.
11. Ballina-García FJ, Queiro-Silva MR, Molina-Suaréz R, et al. Multiple painful bone cysts in a young man. Ann Rheum Dis 1996; 55:346-9.
12. Boyse TD, Jacobson JA. Case 45: cystic angiomatosis. Radiology 2002; 223:164-7.
13. Weiss SW, Goldblum JR. Benign tumors and tumor-like lesions of blood vessels. In: Weiss SW, Goldblum JR, eds. Enzinger and Weiss's Soft Tissue Tumors. 4th ed. St Louis: Mosby, 2001: 837-90.
14. Okumura T, Asamura H, Kondo H, Matsuno Y, Tsuchiya R. Hemangioma of the aib: a case report. Jpn J Clin Oncol 2000; 30:354-7.
15. Khanam H, Lipper MH, Wolff CL, Lopes MB. Calvarial hemangiomas: report of two cases and review of the literature. Surg Neurol 2001; 55:63-7.