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Case report 671

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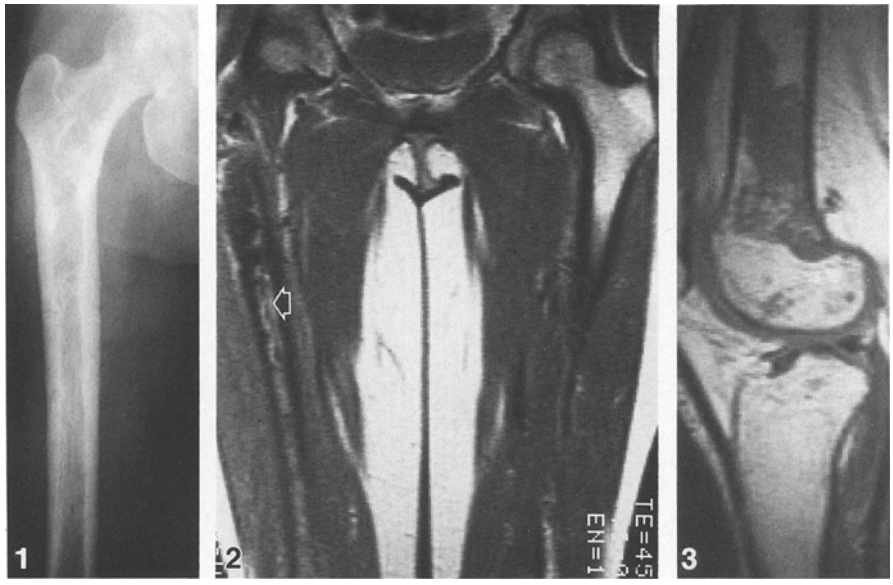

Fig. 1. Frontal view of right femur demonstrates extensive, ill-defined areas of lucency with prominent nutrient foramina involving a long segment of bone

Fig. 2. Coronal magnetic resonance (MR) image of the right femur (TR 2000/TE 45 ms) demonstrates serpiginous areas of signal void (*arrow*) throughout the marrow. No associated soft-tissue mass is noted

Fig. 3. Sagittal MR image of right knee (TR 341/TE 21 ms) shows an irregular geographic area of abnormal signal intensity in the diametaphyseal region of the distal end of the femur with associated joint effusion

Clinical information

A 35-year-old woman presented with intermittent pain and swelling of the right knee and ankle, present since the age of 6 years. Physical examination demonstrated a diffusely warm and swollen knee and calf. No birthmarks or leg length discrepancies were present. Arthrocentesis yielded 35 cc of yellowish, turbid fluid with numerous white and red blood cells. Routine laboratory studies, including antinuclear antibodies (ANA) and rheumatoid factor, were negative. Significant past medical history included "phlebitis" of the right leg as a child, with reportedly normal roentgenograms.

Plain radiographs of the right femur (Fig. 1) showed a long segment of abnormality with alternating ill-defined areas of lucency and sclerosis, endosteal irregularity, prominent nutrient foramina, and asymmetric cortical thickening. No identifiable soft-tissue mass or periosteal reaction was noted. Heterogeneous uptake of tracer, technetium 99m methylene disphosphonate (^{99m}Tc-MDP), was present throughout the right lower extremity. Selected multislice spin echo T_1- and T_2-weighted magnetic resonance (MR) images demonstrated serpiginous areas of signal void running through the marrow cavity (Fig. 2, arrow). In addition, asymmetric prominence of the ipsilateral femoral vessels was noted. Parasagittal surface coil images of the knee demonstrated foci of abnormally decreased signal in the supracondylar region of the femur (Fig. 3); these foci of decreased signal intensity showed enhancement on the T_2-weighted sequences. Similar areas were noted in both the tibial and femoral condyles. The corresponding plain films demonstrated only very subtle sclerosis in the regions noted on the MR images.

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Diagnosis: Arteriovenous malformation of the right lower extremity with associated intraosseous hemangiomatosis

Selective right common femoral arteriograms confirmed hypertrophy of the profunda femoris, with an abnormal regional network of tortuous vessels and arteriovenous shunting, consistent with an arteriovenous malformation (Fig. 4).

Histological examination from both cortical bone specimens and bone marrow curettings from the femur revealed fibrovascular tissue with large numbers of engorged vascular spaces (Fig. 5), consistent with benign intraosseous hemangiomatosis. The endothelial cells lining the spaces were flattened and inconspicuous.

MR and angiographic images directed the eventual percutaneous transcatheter embolization with isobutyl cyanoacrylate.

Discussion

The term "arteriovenous malformation" includes a spectrum of abnormalities of which hemangiomatosis is just one facet. We present this case because this woman had findings including intraosseous microscopic hemangiomatosis and macroscopic (angiographic) evidence of a concomitant arteriovenous malformation.

Intraosseous vascular neoplasms have variable appearances, ranging from discrete hemangiomas to their permeative, more aggressive counterparts. Although the diagnosis may be suspected on plain film radiography, the advent of MRI has enabled the radiologist to discern further both the tissue characteristics and the extraosseous extent of the lesion. This may then aid in both the preoperative management and the planning for possible embolization.

Hemangiomas in the appendicular skeleton are not uncommon and have been well-described [10]. When present in the lower extremities, they have an epiphyseal or metaphyseal predominance [8].

Since intraosseous hemangioma is not uncommon, their importance lies in the exclusion of primary malignant skeletal neoplasms. The plain film differential diagnosis of our case includes malignant hemangioendothelioma (angiosarcoma), hemangiopericytoma, and fibrous dysplasia. In addition, the T_1-weighted MR image of the distal end of the femur may resemble hyerplasia of the red marrow; however, the marked signal enhancement on T_2-weighted images noted herein is extremely unusual for this entity.

The largely geographic, osteolytic appearance lacking periosteal response may also be seen in malignant hemangioendothelioma. In the primary epithelioid variety, described by F.J. Martinez-Tello et al., marked osteolytic destruction, lacking a

Pathological features and femoral arteriogram

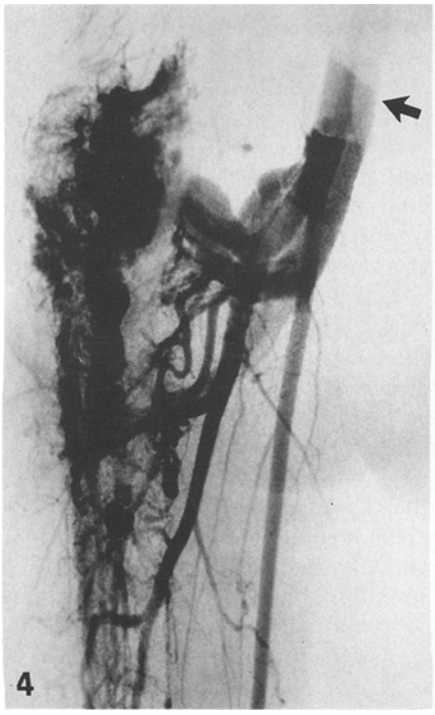

Fig. 4. Right common femoral arteriogram shows extensive arteriovenous malformations and early opacification of the iliac and femoral veins. (Courtesy of R. Rosen, M.D., NYU Medical Center)

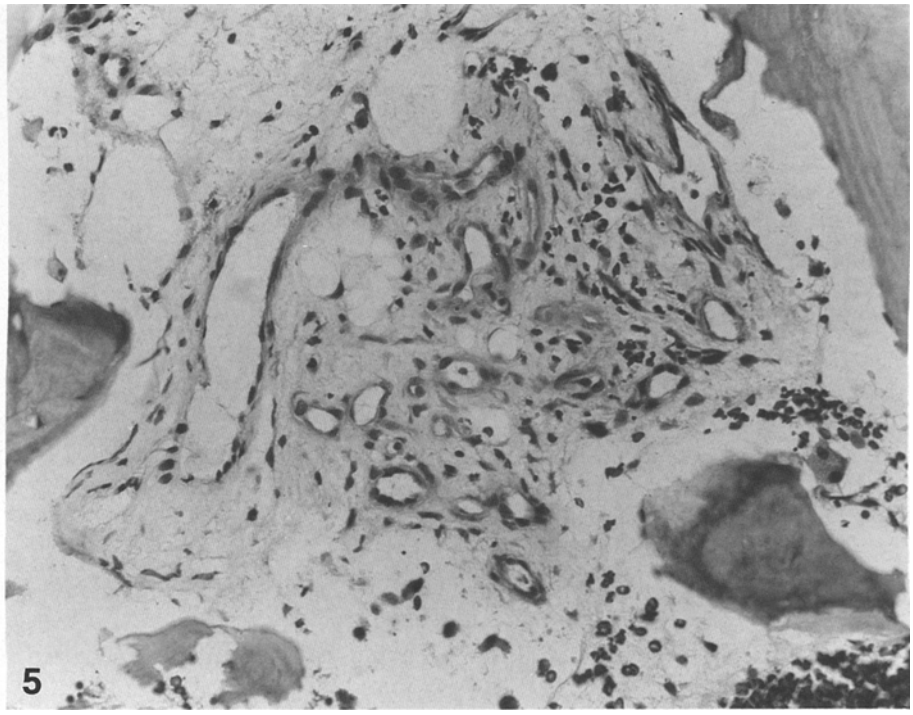

Fig. 5. Histological specimen showing well-formed vascular channels, lined by endothelial cells, present in the medullary bony spaces. This appearance is typical for a localized hemangioma or for a component of diffuse osseous hemangiomatosis (H&E, ×100)

sclerotic rim, with soft tissue invasion [6] may be present. The plain film appearance of a benign vascular tumor may often mimic a malignant process, with an irregular, lytic, or expanding pattern [9]. MR imaging may, therefore, be invaluable in discerning characteristics that speak for a more malignant process (e.g., soft-tissue mass, cortical destruction). The age of our patient and the lack of an associated soft-tissue mass made malignant vascular tumor unlikely. In addition, hemangioma is more common in women [8], as opposed to hemangioendothelioma, which has a male predominance [5].

The presence of well-formed vascular channels lined by flattened, typical endothelial cells, combined with a lack of cellular atypia and pleomorphism, speaks for a benign intraoesseous hemangioma. The plump, eosinophilic cells noted in the "histiocytoid" variety, described by Rosai and later noted by Jaffe et al. [3], were not seen in our case.

Given the characteristic nature of this process, fibrous dysplasia was quickly excluded from the list of differential possibilities.

Multicentricity, as observed in our case, is not an uncommon finding in tumors of vascular origin [6, 8]. Indeed, it has been noted in tumors such as benign hemangioma, histiocytoid hemangioma, and hemangioendothelioma [2].

The angiographic findings of hemangioma typically include engorged vascular spaces which persist late into the venous phase of an arterial injection. The presence of arteriovenous fistulae and an early draining vein is rare in hemangioma; this feature, in conjunction with an enlarged feeding artery, depicts an angiographic pattern typical for a macrofistulous communication or arteriovenous fistula [4]. Given the lack of history of either penetrating trauma or a frank pathological fracture, the malformation was most likely a congenitally formed failure of angiographic differentiation [1].

In *summary*, since extensive involvement of the lower extremity with arteriovenous malformations occurs often combined with the potential risk of hemorrhage, transcatheter embolization has become an attractive alternative to surgical resection [7, 9]. We describe an unusual case of extensive intraosseous hemangiomatosis, with a concomitant arteriovenous malformation that presented in a 35-year-old woman as synovitis in a knee. MR images demonstrated that the lesion was far more extensive than originally suggested and directed the subsequent biopsy and embolization.

The differential diagnosis was discussed and the subject of intraosseous hemangiomatosis with associated AVM was considered in some detail.

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