

Intraosseous myelolipoma

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Abstract Two patients with lesions in the acetabulum and femur, evident on imaging, were found to have normal marrow elements without a histopathological lesion on curettage of the acetabulum and resection of the femur, respectively. Radiographs on the first patient showed a sclerotic benign appearing acetabular lesion reminiscent of a fibro-osseous lesion while MRI showed the entire lesion to contain fat. The histological features were neither of a fibro-osseous lesion nor a lipoma, but only normal to mildly hypercellular marrow elements. The second patient had a long and expansive femoral osteolytic lesion which on biopsy showed cellular marrow. A subsequent stress fracture led to resection and prosthetic replacement. Microscopic findings revealed only cellular hematopoietic marrow with mature red and white cell lines, megakaryocytes, and a few mast cells devoid of trabecular bones. The microscopic features in the

first patient raised the possibility of hematopoietic hyperplasia. Neither the radiographic nor MRI features were consistent with that diagnosis. The spectrum of imaging findings and microscopic appearances in both patients best fit the diagnosis of intraosseous myelolipoma.

Keywords Bone · Myelolipoma · MRI · Radiographs · Histology

Introduction

The presence of a tumor-like lesion in bone which on histologic examination reveals only hypercellular marrow elements without a histopathologic lesion has been termed focal hematopoietic hyperplasia (FHH) [1–3]. Myelolipoma is a tumor-like lesion composed of a variable mixture of a mature fat and hematopoietic elements most commonly encountered in the adrenal gland constituting less than 4% of adrenal tumors [4]. Extra-adrenal myelolipomas are known to occur infrequently. In an Armed Forces Institute of Pathology series, 10 of 86 myelolipomas were extra-adrenal; eight of which were in the retroperitoneum and pelvis [5]. Myelolipomas have been described in other rare sites such as the liver, mediastinum, stomach, and renal sinuses. We are aware of one report, in Italian, of a mandibular myelolipoma without any pre-operative imaging [6]. We are not aware of any reports of intraosseous myelolipoma in the English literature. We report on two patients with intraosseous lesions in which the curetted and resected specimens did not reveal a histopathological entity but only normal marrow elements. In our first patient, the normal to mildly cellular marrow raised the possibility of FHH, but the imaging findings, especially MRI, was

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Fig. 1 **a, b** Radiographs of the right hip show a 4×4 cm benign appearing fibro-osseous lesion in the roof of the right acetabulum with a predominance of sclerosis

inconsistent with that diagnosis (Figs. 1 and 2). The constellation of imaging and microscopic findings in the two patients reported here, we believe, best fit the diagnosis of intraosseous myelolipoma.

Case histories

Patient 1 A 35-year old otherwise healthy female presented elsewhere with gradual onset of right hip pain. Radiographs (Fig. 1a,b) and MRI of the right hip (Fig. 2a,b) were obtained. The lesion curetted and then she was referred to

Fig. 2 **a** T₁ and **b** T₂ weighted coronal MR images show the lesion to be filled with fat

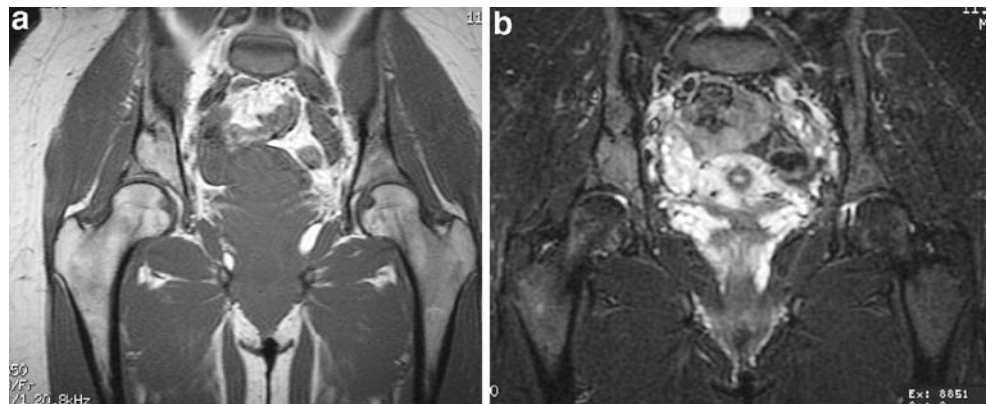
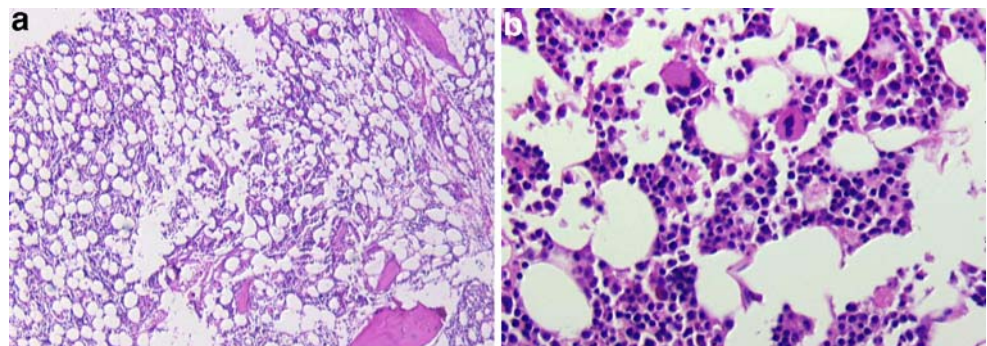


Fig. 3 **a** Low magnification of showing hematopoietic bone marrow with interspersed adipocytes and rare bony trabeculae. **b** Higher magnification showing only normal hematopoietic bone marrow



our institution because she did not get better and the diagnosis was unclear. The radiographs demonstrated a 4 cm × 4 cm well-marginated mixed sclerotic/osteolytic lesion in the roof of the right acetabulum (Fig. 1). The favored diagnosis was fibrous dysplasia. The MR images showed the lesion to be filled with fat (Fig. 2a,b) displaying signal intensity similar to the patient's subcutaneous fat. This prompted a broadening of the diagnosis to include intra-osseous lipoma or fibrous dysplasia with an unusually large amount of fat. The histology confirmed neither fibrous dysplasia nor lipoma but contained mildly hypercellular/normocellular marrow with normal hematopoietic elements (Fig. 3a,b). Because of persisting symptoms, an MR hip arthrogram was performed to exclude communication between the joint and roof of the acetabulum. A mini surgical exploration of the previously operated on site demonstrated a cavity caused by the previous curettage with normal surrounding marrow. The combination of a non-osteolytic mass, marrow signal of fat on MRI and histological features of marrow with normal hematopoietic elements was consistent with myelolipoma. At last follow-up, the patient's symptoms had decreased.

Patient 2 A 51-year-old diabetic was seen in February 1988 for hip pain attributed to degenerative arthritis. A radiograph (Fig. 4) showed a non-aggressive osteolytic lesion in the



Fig. 4 Patient 2. AP pelvic radiograph shows a long benign appearing osteolytic lesion of the femoral diaphysis

proximal femur that was described as being consistent with fibrous dysplasia. Technetium bone scan, computed tomography (CT) and MRI all performed at that time are no longer available. The bone scan was reported as showing no radioactivity in the lesion or elsewhere. The CT scan confirmed the intracompartmental confines of the lesion. The MRI signal features are not known. Biopsy of the lesion showed hematopoietic bone marrow with occasional fat cells (Fig. 5a,b).

Subsequently the patient had a stress fracture of the lesion which led to a resection and prosthetic replacement. The resection specimen (Fig. 6) demonstrated a gelatinous mass that could be shelled out. Microscopically, the lesion showed hematopoietic marrow with mature red and white cell lines (Fig. 7). There were mast cells but no granulomas, fibrous tissue or any other microscopic features of a tumor. Because of the histological make up of red bone marrow the lesion was signed out as “an extra-adrenal myelolipoma in a previously undescribed location.”

Discussion

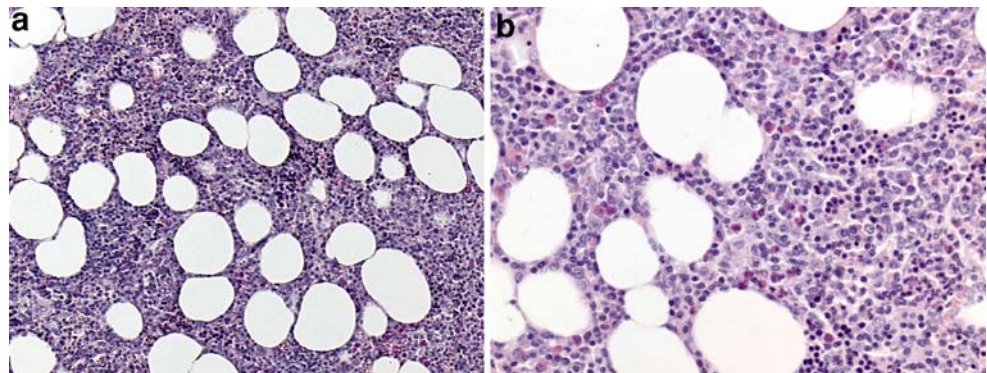
Myelolipomas are relatively rare, benign, nonfunctioning tumors composed of adipose tissue and hematopoietic

elements seen outside the bone marrow. The adrenal gland is the most common location for myelolipomas with fewer than 40 cases reported in extra-adrenal locations [4, 5]. The most common extra-adrenal location is the retroperitoneum [5]. The differential diagnosis of an extra-adrenal myelolipoma would be influenced by the anatomic location of the mass. Extra-adrenal myelolipomas unlike bone marrow do not contain reticular sinusoids or bone spicules but rather the presence of a combination of hematopoietic elements and mature adipose tissue which distinguishes them from retroperitoneal fatty tumors. In the mediastinum, retroperitoneum and pelvis, the major diagnostic microscopic challenge is distinguishing extra-adrenal myelolipomas from extra-medullary hematopoietic tumors [4, 7]. Microscopically, they may be indistinguishable; however, the clinical history aids in their distinction. Extra-medullary hematopoiesis is encountered in patients with chronic underlying hematologic disorders while extra-adrenal myelolipomas are not associated with an underlying systemic disease. There has been a single case report, in Italian, of a lesion in the mandible that was felt to conform to an extra-adrenal myelolipoma [6]. There was no pre-operative imaging and the lesion discovered at surgery. An English translation of this paper did not offer insights as to why the authors believed the lesion was a myelolipoma.

Both patients in our report had lesions confined to bone which on microscopic examination did not show a histological lesion but only cellular marrow. The cellularity was felt to be within the normal range given the ages of the patients and otherwise normal hematopoietic marrow. The histological examinations unequivocally excluded the pathologic entities considered on the imaging findings.

In patient 1, FHH was the histological entity that we most strongly considered but the constellation of imaging findings did not fit what has been described in that entity. FHH was first described by Edelstein and Kyriakos in two patients with rib lesions. Subsequently, two further patients with this diagnosis, again in ribs were reported [2, 3]. All of the rib lesions were predominantly osteolytic, two were expansive, and one had punctate calcification [1–3].

Fig. 5 a, b Low and higher magnification showing hematopoietic bone marrow with occasional fat cells



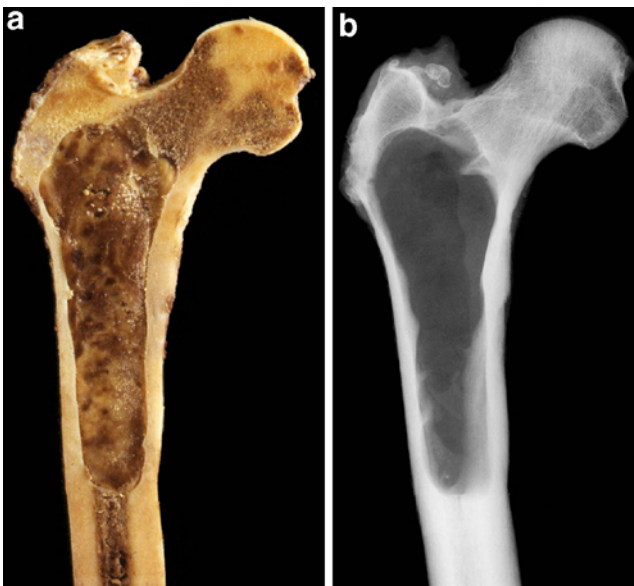


Fig. 6 Resected specimen of femur shows an expansive osteolytic mass with sharply defined pushing margins consisting of marrow-like tissue **a** Photograph and **b** radiograph)

Edelstein and Kyriakos in the discussion of their two cases mention that the supposition of hypercellularity was based on what would be considered “normal” cellularity of rib marrow at the ages at which the lesions were discovered. Our patient’s lesion was sclerotic and histologically the cellularity of marrow was felt to be within the normal range for the patient’s age. A case report on the MRI findings of FHH that was thought to represent a malignant process demonstrated long T_1 , and long T_2 and was falsely positive on a PET scan [8]. Biopsy of this lesion excluded a histopathological entity and revealed only hypercellular marrow. The MRI signal characteristics of the lesion in this case are concordant with what is encountered in patients

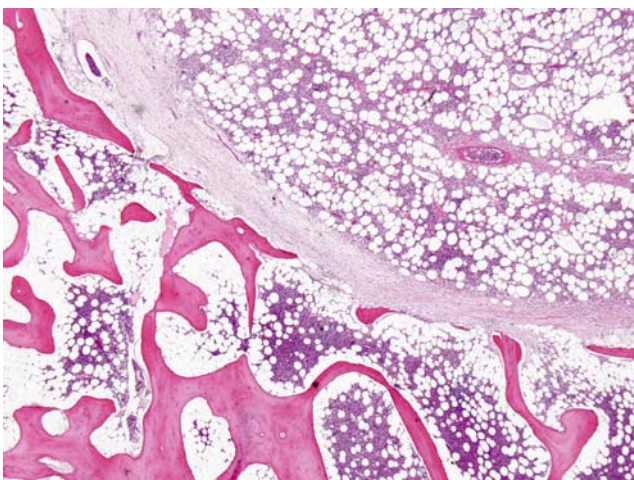


Fig. 7 Low magnification showing myelolipoma (upper right) with a thin fibrous capsule forming a pushing border (in the middle of picture) between receding bone and the space occupying mass consisting of fatty and hematopoietic tissue that lack bony spicules

who receive granulocyte colony stimulating factors (G-CSF) with their chemotherapy [9]. The objective in G-CSF administration is to stimulate the marrow. A hypercellular marrow is a desired response. It is known that the desired G-CSF response can be mistaken for metastases and may be falsely positive on a PET scan [9]. The long T_1 and increased T_2 signal is what would be expected with hypercellular marrow. In our patient, the MRI signal of a short T_1 corresponding to the signal of subcutaneous fat is what is encountered in fat containing lesions and not red marrow hypercellularity.

It is noteworthy in patient 2 that the long osteolytic lesion was cold on a radionuclide bone scan. Two of the four rib lesions showed some accumulation of radioactive tracer [1, 3]. A negative bone scan may be encountered in benign lesions such as cystic fibrous dysplasia or an unusual cyst; both entities could have an appearance similar to the lesion in the femur of patient 2. Biopsy and the resected specimen (Figs. 5 and 7), however, revealed only hematopoietic marrow in patient 2. The lesion had behaved like a tumor in expanding and weakening the femoral shaft. Efforts in recovering the MRI study from 20 years ago have unfortunately been unsuccessful. Based on our analyses of these two patients, intraosseous myelolipoma would appear to be a real entity and is a diagnosis that could plausibly be made when an unequivocal lesion on imaging has no histopathological counterpart and is found to contain only normal cellular hematopoietic marrow.

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