

Research papers

Extra-adrenal myelolipoma and extramedullary hematopoiesis: Imaging features of two similar benign fat-containing presacral masses that may mimic liposarcoma[☆]



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ARTICLE INFO

Keywords:

Extramedullary hematopoiesis

Extra-adrenal myelolipoma

Presacral mass

ABSTRACT

Objective: To describe the imaging characteristics of extra-adrenal myelolipoma and mass-forming extramedullary hematopoiesis (EMH) occurring in the presacral region.

Materials and Methods: Retrospective review between 1980 and 2015 revealed 11 patients with biopsy-proven presacral extra-adrenal myelolipoma and 11 patients with presacral EMH and radiologic imaging studies.

Results: All cases of both myelolipoma and EMH directly abutted the anterior sacrum and were centered on the midline.

Myelolipomas were round or bilobed (8/11), or were multilobulated in appearance (3/11). On CT, margins could be smooth or ill defined. On MRI, all masses were well-marginated with evidence of a “capsule”. All myelolipomas showed evidence of macroscopic fat on both CT and MRI with scattered hazy and nodular soft tissue components. On MRI, all cases showed areas of high T1 signal intermixed with areas of intermediate T1 signal. On T2, all cases showed areas of low and high T2 signal. Enhancement was usually minimal or mild.

Most EMH cases had a well-defined round, bilobed or macro-lobulated presacral mass (9/11); 2/11 cases were ill-defined. On CT, when a dominant fatty component was present, the soft tissue components had hazy and nodular features, similar to myelolipoma. On MRI, the masses demonstrated intermediate T1 signal, isointense to skeletal muscle with varying amounts of hyperintense T1 signal. On T2-weighted images, 3 of 4 cases showed areas of mildly hyperintense T2 signal with varying amounts of dark signal. All patients with clinically or biopsy-proven EMH had a hematologic or myeloid disorder.

Conclusion: Presacral myelolipoma and presacral EMH are two benign fat-containing masses that can present as discrete, often encapsulated, lipomatous masses with varying amounts of non-lipomatous soft tissue components and should be considered in the differential diagnosis of a well-defined fat-containing presacral mass.

1. Introduction

Although well-differentiated liposarcoma (WDLS) is the most common fat-containing retroperitoneal sarcoma, presacral myelolipoma and presacral extramedullary hematopoiesis (EMH) are two benign fat-containing masses that can be mistaken for WDLS on imaging studies. The presacral region is the most common extra-adrenal site for myelolipoma and the 2nd most common site for mass-forming EMH [1]. Both of these masses can present as discrete predominantly fat-containing masses with varying amounts of non-lipomatous soft tissue components [1–9].

Myelolipomas occur primarily in the adrenal gland, but have been well-described at extra-adrenal sites (Fig. 1). Histologically, they are composed of variable amounts of mature adipose tissue admixed with hematopoietic marrow elements undergoing normal trilineage hematopoiesis. They can, however, have increased numbers of megakaryocytes. In contrast to extramedullary hematopoiesis, myelolipomas are almost invariably associated with normal bone marrow. Cytogenetic evidence of clonality and the identification of a balanced chromosomal translocation in a myelolipoma suggest that they are best classified as (benign) tumor [1–4,8,10–12].

Mass-forming extramedullary hematopoiesis (EMH) is another fat-

Abbreviations: EMH, extramedullary hematopoiesis; WDLS, well-differentiated liposarcoma; Gad, gadolinium

[☆] Presented at: RSNA Annual Meeting, 2015, Educational Exhibit.

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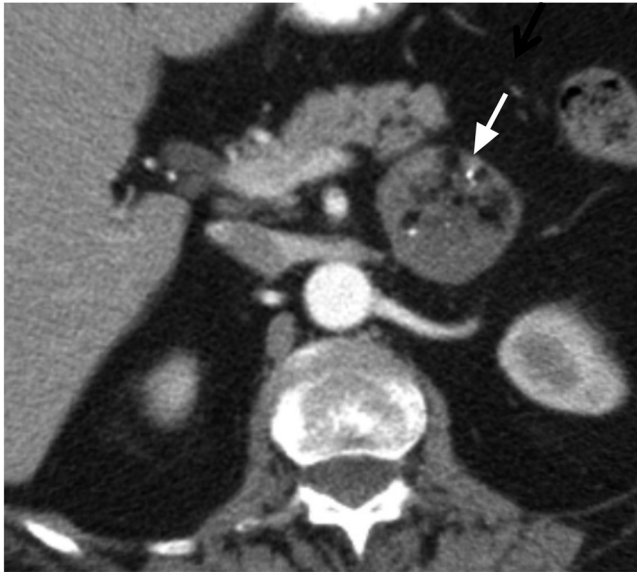


Fig. 1. Adrenal myelolipoma. Axial contrast-enhanced CT image through the upper abdomen demonstrating a round soft tissue mass containing foci of macroscopic fat in the left adrenal gland (arrow). There are a few scattered calcifications. Findings are typical of benign adrenal myelolipoma.

containing, mass-forming lesion with very similar histologic features to those of myelolipoma. EMH is also characterized histologically by varying amounts of adipose tissue admixed with hematopoietic marrow elements showing trilineage hematopoiesis. However, in contrast to myelolipoma, EMH typically occurs in patients with hematologic, myeloid or lymphoid disorders, or other bone marrow-infiltrating processes, and reflects inadequate or ineffective medullary hematopoiesis.

The purpose of our study was to characterize the imaging findings of presacral extra-adrenal myelolipoma and EMH and delineate any radiologic features or clinical factors that may aid in making the diagnosis.

2. Materials and methods

Following IRB approval, our institution's radiology and pathology databases were searched for cases of presacral extra-adrenal myelolipoma and presacral mass-forming EMH from 1980 through 2015, using key words "fat-containing presacral mass"; "presacral extra-adrenal myelolipoma" and "presacral extramedullary hematopoiesis". These cases were reviewed for imaging features and patient demographics. Among cases with image-guided biopsy; histologic findings were correlated with clinicoradiologic findings to distinguish between EMH and myelolipoma.

A thorough review of the medical record was performed, including the patients' clinical diagnoses, pathology reports, and clinical laboratory data. Patient age, gender and relevant clinical diagnoses were recorded. Additional imaging studies were also reviewed for other pertinent findings such as the presence or absence of the following: additional soft tissue masses (including thoracic paraspinal masses typical of EMH), adrenal masses, splenomegaly, and bone marrow abnormalities including multiple bone lesions or signs of marrow expansion or infiltration.

CT images were assessed for shape of the mass (round/oval, bilobed or multilobulated, irregular or infiltrative), margination (well-defined smooth, well-defined irregular or infiltrative), the presence or absence of a discrete "capsule", the amount of identifiable fat present (if any), the pattern of non-lipomatous components within the mass, and the enhancement pattern. MRI images were assessed for similar features including shape, margination, presence of a capsule, signal

characteristics, amount of identifiable fat, and enhancement characteristics. In addition to the imaging findings, the radiologist's initial impression and most likely diagnosis or diagnoses were also recorded.

FDG PET-CT images were assessed for the presence of FDG activity within the presacral masses. SUV max was determined in the standard fashion by creating a three-dimensional volume of interest incorporating the gross neoplasm volume. Planar whole body Tc-99 m sulfur colloid images were assessed for the presence of radiotracer uptake in the presacral region, best viewed on the posterior planar images. The presence of a marrow expansion pattern and any additional sites of abnormal extraskelatal radiotracer uptake were noted.

3. Results

3.1. Patient demographics

There were 11 cases of biopsy-proven presacral extra-adrenal myelolipoma. All 11 patients were females in their 7th–9th decade of life (average 71 years of age). All 11 had at least one CT and 10 of 11 had MRI exams available for review. Two patients had FDG PET-CT scans available for review.

There were 11 cases of presacral mass-forming EMH. Five cases were biopsy proven. Six cases were clinically proven through either a known hematologic or myeloid disorder with additional sites of paraspinal extramedullary hematopoiesis and/or uptake on Tc-99 m sulfur colloid bone marrow scan. All 11 cases had a hematologic or myeloid disorder: Beta thalassemia (3), iron deficiency anemia (2), combined iron and B12 deficiency anemia (2), congenital hemolytic anemia (1), myelofibrosis (1), myelodysplastic syndrome (1), and polycythemia vera (1). Although 3 patients had a lifelong history of beta thalassemia, all presacral soft tissue masses were diagnosed between the ages of 42 and 83 years. The average age of diagnosis for presacral EMH was 61 years. Of the 11 patients with EMH, all 11 had CT scans, 4 had MRI exams, 2 had FDG PET-CT scans, and 5 had Tc-99 m sulfur colloid bone marrow scans available for review.

3.2. Imaging of presacral extra-adrenal myelolipoma

Shape and margins: On CT and MRI, all presented as a discrete well-defined soft tissue mass or masses that were confined to the presacral region. In all cases, on both CT and MRI, the tumor directly abutted and contacted the anterior sacral cortex with epicenter on the midline and without evidence of bone destruction or remodeling (Fig. 2). The majority of the masses (8 of 11) were round, oval or bilobed; 2 cases were multilobulated with an appearance more suggestive of a cluster of contiguous round midline masses in the craniocaudal plane, which was best characterized in the sagittal plane (Fig. 2). On CT, although all were discrete focal masses, 5 cases had somewhat irregular margins and 6 cases had smooth well-defined margins with a thin peripheral "capsule" (Fig. 3). In all cases of myelolipoma, even when margins appeared ill-defined on CT, MRI demonstrated well-defined smooth encapsulated margins ("capsule sign"). This "capsule sign" was typically best demonstrated in the sagittal plane (Fig. 3).

Presence of lipomatous components: All masses were heterogeneous and showed imaging evidence of fat on both CT and MRI (Figs. 4 and 5). Masses with 50% or greater fat composition were found in 8 of 11 CT exams and 8 of 10 MRI exams. Less than 50% fat was found in 3 of 11 CT and 2 of 10 MRI exams. However, foci of fat were readily identifiable on MRI and CT in all cases, including the minority of cases where the quantity of non-lipomatous tissue exceeded the lipomatous component.

CT Characteristics: On CT, the masses typically presented as a focal round or lobulated presacral mass comprised predominantly of fat with diffuse hazy and scattered nodular non-lipomatous components (Figs. 2 and 3). As noted previously, the margins of the mass were sometimes difficult to define on CT due to the presence of fat at the periphery of

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