An extremely rare case of synchronous intrathoracic and intra-abdominal tumors

Preeti Goyal, Binit Sureka1, Devendra Laddha2
Departments of Radiology and2Pathology, Getwell Polyclinic and Hospital, Jaipur, Rajasthan 1Department of Radiology, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi, India

Address for correspondence: Dr. Binit Sureka, Department of Radiodiagnosis and Imaging, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi - 110029, India. E-mail: binitsurekapgi@gmail.com

BACKGROUND

A 42-year-old man was evaluated for general malaise and abdominal fullness in our center. On examination the patient was afebrile, and his physical examination shows flank fullness with mild abdominal tenderness. Laboratory findings were unremarkable; white blood cell count (12300/mm³), increased proportion of neutrophils on differential analysis (75% neutrophils) and hemoglobin were 14 g/dl. Conventional chest radiograph was done [Figure 1]. Given the patient’s history, physical examination, and laboratory findings, abdominal sonography (USG) was also done [Figure 2]. Patient also underwent computed tomography (CT) scan of chest and abdomen and MRI of upper abdomen [Figures 3 and 4].

QUESTIONS

• What is the radiological finding demonstrated in plain chest radiograph, ultrasound scans, CT and MRI?
• What is the diagnosis in this patient?
• What are the other important differential diagnoses for the given radiological findings on CT scan and MRI?

Figure 1: Conventional chest radiograph shows bilateral lobulated mediastinal masses

Figure 2: (a) Ultrasound abdomen shows right perirenal echogenic masses completely surrounding the kidney (b) left perirenal echogenic masses (c) splenomegaly with multiple variable sized well-defined intrasplenic echogenic mass

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Quick Response Code:  
Website: www.lungindia.com  
DOI: 10.4103/0970-2113.159607
ANSWERS

Answer 1
The given chest radiograph shows lobulated bilateral paraspinal masses which does not silhouette the cardiac borders and a pleural-based opacity [Figure 1]. Abdominal sonography (USG) shows perinephric, intrasplenic and presacral echogenic masses [Figure 2]. Contrast-enhanced computed tomography (CT) scan showed multiple fat containing intrathoracic paravertebral and pleural-based masses, bilateral bulky perirenal and presacral pelvic masses. All of them revealed fat attenuating areas (−20 to −94 HU) with interspersed non-fatty soft-tissue densities [Figure 3]. Spleen was also enlarged with multiple intrasplenic hypoattenuating lesions. No lymphadenopathy was noted. Both kidneys were entirely embedded in the fatty masses. Multiple MRI sequences were done to further characterize the masses. Typical fat characteristics were displayed as hyperintense signal on $T_1$-weighted images and intermediate signal on $T_2$-weighted images. Fat suppression images demonstrated loss of signal in the fatty components of the lesion. Myeloid elements had low signal intensity on $T_1$-weighted images and intermediate intensity on $T_2$-weighted images. Intrasplenic lesions were better demonstrated [Figure 4].

Answer 2
USG-guided fine-needle aspiration and core biopsy of both intrathoracic and perirenal masses were performed. The core biopsy of masses showed plenty of adipose tissue with interspersed hematopoietic precursors including myeloid, erythroid, and megakaryocyte elements. Several mature plasma cells and occasional hemosiderin-containing macrophages were found. Increased aggregates of lymphoid cells were also found as well. Results were consistent with the diagnosis of myelolipoma. Bone marrow biopsy revealed normal cellularity with no evidence of fibrosis, granulomatous inflammation, or cells extrinsic to the marrow.

Answer 3
Appearing as a fat containing soft tissue tumor, the radiological differential diagnoses are liposarcomas, extramedullary hematopoiesis (EHT), lipoma, angiomyolipoma and myolipomas.

DISCUSSION
Myelolipomas, first described in 1905 by Gierke, are rare benign nonfunctional tumors composed of a variable mixture of mature adipose tissue and hematopoietic elements resembling bone marrow.[1] These lesions most commonly involve the adrenal glands, constituting <4% of adrenal tumors. With improved non-invasive diagnostic imaging in current scenario, incidental discovery of myelolipomas has become increasingly common.[2] Possibilities include development from embryonic mesenchymal rests in the adrenal gland, hematogenous bone marrow emboli, stress-induced reticuloendothelial cell metaplasia and proliferation of embryonic rests of hematopoietic stem cells.[3]

Myelolipomas are found in wide age range with no sex predilection. Their sizes are also highly variable. Malignant degeneration has not been reported yet.[3] Most of the patients...
are asymptomatic, local tumor extension or rare occurrence of bulky multifocal masses may lead to non-specific complaints. Imaging findings are quite characteristic. CT shows predominantly low-attenuation areas of adipose tissue with high-attenuation areas of hematopoietic elements. These lesions are well defined with preserved adjoining fat planes. MRI also accurately depicts macroscopic fat using fat saturation technique. Myelolipomas may be discovered incidentally on USG as echogenic masses at various sites but with less accuracy.

Only about 50 cases of extra-adrenal myelolipomas (EAML) are reported in the literature. Their incidence is 0.08-0.4% at autopsy. Thetypical lesion is a solitary, well-defined mass within the abdomen, commonly in the retroperitoneal presacral area (comprising approximately 1/2 of reported cases). Other infrequent sites of involvement include pelvic, perirenal and hepatic regions, with sporadic cases in thoracic, splenic and gastric locations. In thorax these are usually found as posterior mediastinal masses with very rare occurrence of pleural-based mass. The sizes of EAML have been reported to range from 4 to 15 cm, with a mean diameter of 8.2 cm. EAML occurs more commonly in women, with a male: female ratio of 1:2. The median age at diagnosis is 66.5 yr. A strong association of EAML with underlying inflammatory disorders, diabetes, and cardiovascular disease has been described, similar to that seen with adrenal myelolipomas. Since EAML are histologically identical to their adrenal counterpart, the CT and MR findings appear similar.

The radiological differential diagnoses are mentioned above. Specific tissue diagnosis plays the major role and often needed in reaching a final diagnosis. The radiological appearance of liposarcomas depends on its histological type and is usually not well circumscribed. Histologically, well-differentiated liposarcomas are non-hemorrhagic and have lipoblast zones of atypical cellular features. Extramedullary hematopoiesis is a compensatory event in patients with underlying hematological disorder showing marked bone marrow hyperplasia, splenomegaly or other organomegaly and multifocal masses. Unlike extra-adrenal myelolipomas, masses of extramedullary hematopoiesis are usually ill-defined and lack fat. EHT typically demonstrates highcellularity, with universal presence of erythroid hyperplasia, scanty adipose tissue and lack of lymphoid aggregates. Angiomyolipomas are well-defined fat-containing vascular masses that typically arises from the renal cortex, therefore producing a defect in the renal parenchyma. Myelolipomas, on the other hand, have a smooth interface between the cortex and parenchyma that might help in differentiating these lesions. Angiomyolipomas can be differentiated by the lack of megakaryocytes, the presence of thick-walled blood vessels, and scattered spindle cells, which are immunoreactive to HMB-45. Lipomas contain only adipose tissue. No soft tissue seen within them. Myolipomas are very rare and donot contain marrow elements.

Specific therapy of these benign myelolipomas is often not required, as most patients are asymptomatic. Few of the patients which are symptomatic due to local compression require surgical resection. In asymptomatic patients, follow-up with serial CT scans is appropriate.

REFERENCES

1. Gierke E. Uber Knochenmarkesgewebe in der Nebenniere. Beitr Pathol Anat 1905;7:311-25.
2. Kenney PJ, Wagner BJ, Rao P, Heffess CS. Myelolipoma: CT and pathologic features. Radiology 1998;208:87-95.
3. Kammen BF, Elder DE, Fraker DL, Siegelman ES. Extraadrenal myelolipoma: MR imaging findings. AJR Am J Roentgenol 1998;171:721-3.
4. Talwalkar SS, Shaheen SP 2nd. Extra-adrenal myelolipoma in the renal hilum: A case report and review of the literature. Arch Pathol Lab Med 2006;130:1049-52.
5. Hunter SB, Schemankewitz EH, Patterson C, Varma VA. Extra-adrenal myelolipoma: A report of two cases. Am J Clin Pathol 1992;97:402-4.
6. Kumar M, Duerinckx AJ. Bilateral extraadrenal perirenal myelolipomas: An imaging challenge. AJR Am J Roentgenol 2004;183:833-6.
7. Temizoz O, Gencelac H, Demir MK, Unlu E, Ozdemir H. Bilateral extra-adrenal perirenal myelolipomas: CT features. Br J Radiol 2010;83:e198-9.
8. Sabate CJ, Shahian DM. Pulmonary myelolipoma. Ann Thorac Surg 2002;74:573-5.
9. Fowler MR, Williams BB, Alba JM, Byrd CR. Extra-adrenal myelolipomas compared with extramedullary hematopoietic tumors: A case of presacral myelolipoma. Am J Surg Pathol 1982;6:363-74.
10. Zieker D, Kunigtrainer I, Miller S, Vogel U, Sotlar K, Steurer W, et al. Simultaneous adrenal and extra-adrenal myelolipoma-an uncommon incident: Case report and review of the literature. World J Surg Oncol 2008;6:72.
11. Sawhney R, McRae B, Lazarchick J. A rare case of a multifocal extra-adrenal myelolipoma with markedly hypocellular bone marrow. Ann Clin Lab Sci 2006;36:208-11.