Multifocal extra-adrenal myelolipoma arising in the greater omentum

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ABSTRACT

Myelolipomas are rare benign tumors composed of mature fat and hematopoietic elements. They are most often discovered incidentally within the adrenal glands, with extra-adrenal myelolipomas being extremely rare tumors. We report a case of multifocal omental extra-adrenal myelolipoma in a patient who had undergone bilateral adrenalectomy. To our knowledge, this is the first reported case of an intraperitoneal extra-adrenal myelolipoma.

CASE REPORT

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A 32-year-old man presented to the emergency department of our institution following acute onset of fever in the setting of a one-week history of left lower quadrant abdominal pain. The patient was known to have a personal and family history of multiple endocrine neoplasia type IIA syndrome, and had undergone total thyroidectomy at the age of five for medullary carcinoma, and bilateral adrenalectomy at the age of ten for bilateral pheochromocytomas. On examination the patient was febrile, and his physical exam was significant for left lower quadrant abdominal tenderness, otherwise unremarkable. Laboratory testing revealed elevated white blood cell count (12300/mm³) and increased proportion of neutrophils on differential analysis (75% neutrophils). Results of other routine hematologic tests, electrolyte levels and renal function were normal.

Given the patient's history, physical examination, and laboratory findings, CT scan of the abdomen and pelvis with intravenous and oral contrast was ordered for evaluation of possible diverticulitis and to rule out other gastrointestinal, renal, and urologic disease. CT scan demonstrated the presence of acute sigmoid diverticulitis; incidentally, two large mass lesions in the omentum containing both soft tissue and fat components were identified (Figure 1. A, B). One lesion measured 11.4 x 10 x 11 cm (in the transverse, anteroposterior, and craniocaudal dimensions respectively) and the other measured 12 x 9 x 13.6 cm (in the transverse, antero-posterior, and cranio-caudal dimensions respectively).

The patient was subsequently treated with intravenous antibiotics and diet modification over the course of 3 days with resolution of symptoms.

The significance of the omental masses was unclear and it was considered unlikely that the masses are related to the resected adrenal pheochromocytomas, twenty two years ago. Prior to discharge the patient underwent percutaneous CT-guided biopsy and the pathological differential diagnosis included extra-adrenal myelolipoma, extramedullary hematopoiesis, sclerosing extramedullary hematopoietic tumor, and angiomyolipomas.

The decision was then made by the patient and his care team to follow the masses for progression, regression, and disease potential with hematologic laboratory evaluations, physical exams, and serial CT imaging at regular intervals. A follow-up CT examination after a 5-month interval demonstrated stability of the omental masses. Throughout this time the patient remained asymptomatic without any sign of myeloproliferative disease. Three months later the masses were laparoscopically resected for definitive pathologic diagnosis. One mass measured 14 x 12 x 8 cm, weighed 650 grams, and was attached to a segment of omental fat measuring 14 x 5 x 2 cm (Figure 2). Another mass measured 14 x 10 x 7.5 cm, and was attached to a 41 x 18 x 3 cm segment of omentum. The masses were encapsulated, yellow-tan to red in color, nodular in appearance, and solid on cut sections without evidence of necrosis. The sections revealed mature adipose tissue with intermingled hematopoietic cells representing all three lineages of hematopoiesis (Figure 3). These morphological features are most consistent with a myelolipoma. Flow cytometry showed no evidence of leukemia or lymphoma. There were no additional masses or lymph nodes found in the omentum. The final pathologic diagnosis of extra-adrenal myelolipoma was made. In the interim, two years have elapsed and the patient is in good health without any sequela attributed to the extra-adrenal myelolipomas.

DISCUSSION

Myelolipomas, first described in 1905 by Gierke, are benign tumors comprised of fat and hematopoietic elements (1). They typically are nonfunctional incidental tumors discovered on crossectional imaging within the adrenal glands. Malignant degeneration has not been reported to date (2). The exact aetiology of myelolipomas is still unknown. Derivation from hematopoietic stem cells that transmigrate into the adrenal glands during development is one valid theory; another theory is that they are derived from bone marrow rests that are trapped in the adrenal cortex and a third theory states that myelolipomas are found predominantly in the adrenals because previously uncommitted adrenal cortical mesenchymal cells undergo metaplasia to become myelolipomas (2).

On imaging, the diagnosis of a myelolipoma can be made with confidence when macroscopic fat is identified within an adrenal mass (3). The preferred imaging modality is CT, which shows focal fatty density within the mass. MRI also accurately depicts macroscopic fat using fat saturation technique. Myelolipomas may be discovered incidentally on ultrasound (US), which otherwise is not used routinely to characterize adrenal neoplasms.

Only about 50 cases of extra-adrenal myelolipomas are reported in the literature (2), and the diagnosis of such lesion is a challenge, predominantly to the radiologist when this tumor is first encountered. The radiological and pathological features of extra-adrenal myelolipomas are the same as the adrenal counterparts except that they are not related to the adrenal glands. Appearing as a fat containing soft tissue tumor, the radiological differential diagnosis will include predominantly liposarcoma, and subsequently tissue diagnosis will play the major role in reaching a final diagnosis.

The pathogenesis of extra-adrenal occurrence is still unclear; however, it has been postulated that they may be the result of misplaced myeloid cells during embryogenesis, followed by hyperplasia (4). Myelolipomas are found in a wide age range. There is no sex predilection, and their sizes are highly variable; most measuring between 5 and 10 cm. There have been reported cases in which they have weighed more than 5000 grams and have measured more than 40 cm in diameter (4). Clonality in myelolipomas has been demonstrated (5, 6).

In this case, the pathological differential diagnosis included extramedullary myeloid tumors and angiomyolipomas. Extramedullary myeloid tumors are usually accompanied by marked bone marrow hyperplasia, splenomegaly or other organomegaly, and diffuse extramedullary hematopoiesis. Unlike extra-adrenal myelolipomas, foci of extramedullary hematopoiesis usually are ill-defined and lack fat (2). Angiomyolipomas can be differentiated by the lack of megakaryocytes, the presence of thick-walled blood vessels, and scattered spindled cells, which are immunoreactive to HMB-45 (7). Microscopic examination, in our case, revealed encapsulated tumors composed of mature adipose tissue with sheets or intermingled hematopoietic cells, and the diagnosis of extra-adrenal myelolipomas was made (Figure 3).

What is unique about the case we have presented is that the patient had prior bilateral adrenalectomy, and developed a multifocal extra-adrenal myelolipoma in a previously unreported intraperitoneal location, within the greater omentum. Excluding lesions within the liver and the stomach, all reported cases were located within the extra-peritoneal spaces of the abdomen: the presacral space, pararenal and perirenal spaces, and renal sinus (7). Extra-abdominal myelolipomas were reported within the spinal canal, the lungs and the mediastinum (8-10). Zieker D, et al. (1) reported a case of synchronous, adrenal and extra-adrenal myelolipomas.

TEACHING POINT

Extra-adrenal myelolipomas are extremely rare tumors, arising predominantly in the retroperitoneum. We report the first case arising in the peritoneal cavity in a patient who had undergone bilateral adrenalectomy.

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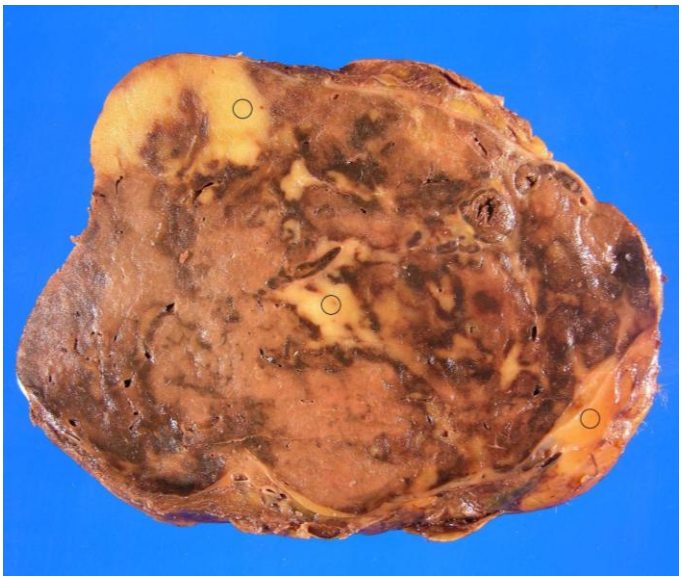

Figure 2 (top): 32-year-old male patient with multifocal extra-adrenal myelolipoma. Gross appearance of the surgically resected extra-adrenal myelolipoma with yellow areas (black cursors) containing adipose tissue admixed with hemorrhagic foci consistent with bone marrow tissue.

FIGURES

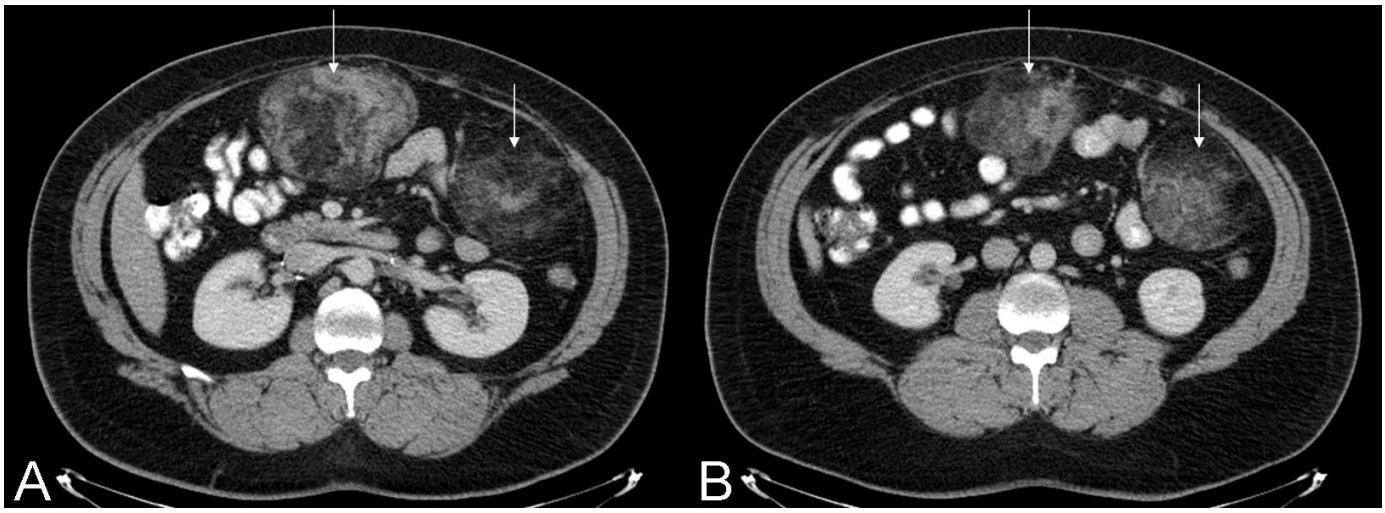

Figure 1: 32-year-old male patient with multifocal extra-adrenal myelolipoma. A, B: Axial contrast enhanced (IV and PO) CT images through the mid abdomen obtained in the equilibrium phase show two large well circumscribed soft tissue masses containing significant amount of fat (arrows), located anterior to the small bowels, within the greater omentum. (Technique: KVp = 120; mA = 389; Slice Thickness = 5.00 mm; Dose of intravenous contrast: Isovue-300 (Iopamidol), 100 ml)

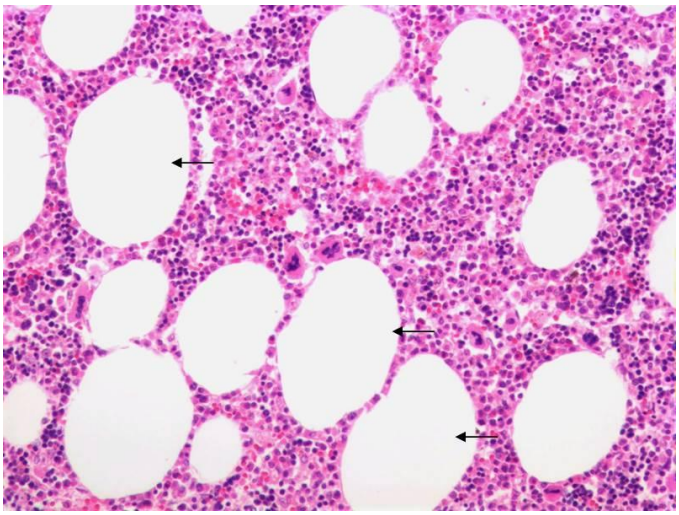

Figure 3: 32-year-old male patient with multifocal extra-adrenal myelolipoma. Extra-adrenal myelolipoma showing normal adipocytes (arrows) with normal maturing myeloid, erythroid, and megakaryocytic lineages (hematoxylin-eosin, original magnification x 20).

ABBREVIATIONS

CT = Computed tomography
MRI = Magnetic resonance imaging
IV = intravenous
PO = per os

KEYWORDS

Myelolipoma, extra-adrenal, omentum, peritoneum