

Symptomatic giant adrenal myelolipoma associated with cholelithiasis: Two case reports

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Abstract

In this article, we have discussed about two cases of adrenal myelolipoma and aim to discuss the role of imaging in their diagnosis and their management. Different imaging techniques such as ultrasound, computed tomography and magnetic resonance imaging were used to aid in diagnosis in each of the cases. The findings have been highlighted here. In each of the cases, the diagnosis could be confirmed by imaging, and there was cholelithiasis seen associated with unilateral adrenal myelolipoma. Adrenal myelolipomas are rare, benign, non-functional tumors of adrenal gland. Most tumors are unilateral and small; bilateral, giant myelolipomas are extremely rare. The association of adrenal myelolipoma with gallstones is uncommon. To our knowledge only two cases of such an association have been reported in the literature. However, the possibility does exist and steps should be taken to ensure a complete diagnosis. Also, it is important to understand the key points which help us in diagnosing adrenal myelolipomas by imaging.

Key Words: Adrenal myelolipoma, computed tomography, magnetic resonance imaging, magnetic resonance cholangiopancreatography, ultrasound

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INTRODUCTION

Adrenal myelolipomas are benign, biochemically inactive neoplasms composed of mature adipose and hematopoietic tissues in varying proportion. They were first described by Gierke in 1905, and were named as myelolipoma in 1929 by Oberling.^[1] Since they are usually asymptomatic and diagnosed incidentally at autopsy or on imaging studies, they are often classified as 'incidentalomas'. Although myelolipomas are commonly found in adrenal glands, rare extra-adrenal myelolipomas are also

known to occur at various sites, including lung, mediastinum, liver, spleen, mesentery, pelvis, presacral region, retroperitoneum and thoracic spine.^[2] The association of myelolipomas with obesity, hypertension, and diabetes mellitus is well known;^[3-5] however, their association with gallstones is uncommon. To our knowledge only two cases of such an association are present in literature.^[6,7] We herein report two cases of giant symptomatic adrenal myelolipoma associated with cholelithiasis. We further discuss the role of imaging in the diagnosis of myelolipomas, various important but rare complications associated with them, and their management alternatives.

CASE REPORTS

Case 1

A 65-year-old obese female with remote past history of cholecystectomy (for cholelithiasis), presented with complaint of nausea, mild jaundice and persistent dull aching pain in whole upper abdomen of few weeks duration. There was

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no significant past or present medical history. Per abdomen examination revealed a well defined, smooth mass in left hypochondrium. On laboratory analysis, liver function test revealed increase in total bilirubin (3.24 mg/dl), conjugated bilirubin (2.17 mg/dl), and alkaline phosphatase (224 U/L). Renal function test was unremarkable. Routine blood examination revealed mildly decreased hemoglobin (8 g/dL). Endocrine evaluation of the adrenal mass revealed normal 24 hours urinary metanephrine (0.9 mg/24 hours) and plasma cortisol (<5 µg/dl). Furthermore, the levels of two tumor markers, carcinoembryonic antigen (1.25 ng/ml) and alpha-fetoprotein (1.00 ng/ml) were also within normal range. The patient was sent to the radiology department for evaluation of obstructive jaundice and left hypochondrial lump.

Ultrasound examination revealed a large well encapsulated, heterogeneous left suprarenal mass lesion [Figure 1a]. In addition, non-visualization of gall bladder (status post-cholecystectomy for cholelithiasis), with dilatation of common bile duct (CBD 20.8 mm diameter), increased width and prominence of central intrahepatic biliary radicals (IHBR) in absence of any intraluminal filling defect were also noted. The plain and contrast computed tomography image [Figure 1b] of the abdomen revealed a well circumscribed, heterogeneous enhancing mass of 91.4 (AP) × 67.3 (TR) × 101.3 (SI) mm in the left supra-renal region causing inferior displacement of the left kidney. The lesion was predominantly of fat density (-30 to -90 HU) with few enhancing soft tissue components and irregular septations. Few tiny flecks of calcifications were also visible in the lateral wall of the mass. The coronal and sagittal CT images [Figure 1c] confirmed the adrenal origin of the mass. The function of both kidneys appeared normal. Abdominal magnetic resonance imaging revealed a heterogeneous, high intensity mass on T1 and T2-weighted images [Figure 1d], with hypointense septae. Linear hyperintense areas posteriorly on T2WI represented intratumoral bleed. Fat-saturated T2-weighted gradient echo sequences [Figure 1e] showed loss of signal intensity in the fatty part of the mass, while the hematopoietic tissue and hemorrhage demonstrated persistent areas of increased signal intensities. Magnetic resonance cholangiopancreatography (MRCP) confirmed the ultrasonographic finding of dilated CBD and central IHBR; T2-haste coronal sequence demonstrated partial benign stricture at ampullary end of CBD as a cause of biliary dilation [Figure 1f].

The imaging findings were highly suggestive of left adrenal myelolipoma, with CBD and central IHBR dilatation due to benign stricture at ampullary end of CBD. Right adrenal gland was normal. The patient underwent surgical resection of large adrenal mass and endoscopic stenting of the biliary stricture. Histopathology of resected left adrenal mass showed mature fat cells admixed with hematopoietic tissues without cellular

atypia. The diagnosis of adrenal myelolipoma was confirmed. The postoperative recovery was uneventful.

Case 2

A 33-year-old obese female presented with vague right abdominal pain for last three months. She had no other significant medical history. On laboratory analysis, hemogram, biochemical and endocrine function tests were within normal limits. Liver and renal function tests were also normal. Levels of tumor markers such as carcinoembryonic-antigen and alpha-fetoprotein were unremarkable. Physical examination revealed a right hypochondrial lump.

Ultrasound examination of abdomen showed a huge, well encapsulated, solid echogenic mass lesion between the liver and right kidney [Figure 2a]. Normally distended gallbladder with multiple intraluminal calculi was also noted. CBD and IHBR were normal. Plain and contrast CT abdomen [Figure 2b] revealed a well circumscribed 83.9 (AP) × 72.2 (TR) × 104.4 (SI) mm sized right suprarenal mass predominantly of fat density (-50 to -90 HU) with peripheral small enhancing soft tissue component on the postero-medial aspect of the mass. A punctate focal calcified area was also seen adjacent to the soft tissue component. Furthermore, multiple small focal calcific lesions (calcified granulomas) randomly distributed within the splenic parenchyma were seen. Suprarenal nature of the mass was confirmed by coronal and sagittal CT images [Figure 2c], which also showed postero-inferior displacement of right kidney. Left adrenal gland and both kidneys appeared normal. MR abdomen revealed predominantly hyperintense mass on T1- and T2-weighted image [Figure 2d]. T2W-fat-suppression image [Figure 2e] showed loss of signal from large fatty portion of the mass, but persistence of increased signal intensity in areas occupied by hematopoietic tissue and intratumoral hemorrhage. Double echo chemical shift gradient echo imaging [Figure 2f] demonstrated few focal areas of signal loss in out-of-phase image as compared to in-phase image, consistent with the presence of microscopic (intracellular) fat, while the major portion of the mass demonstrated high signal intensity consistent with the presence of macroscopic (extracellular) fat.

The imaging finding and hormonal evaluation highly suggested the diagnosis of a nonfunctioning right adrenal myelolipoma. Right adrenalectomy and cholecystectomy was performed in the same setting to prevent future risk of spontaneous hemorrhage and gall stone associated complications. Histopathology confirmed the diagnosis. Post-operative period was uneventful.

DISCUSSION

Adrenal myelolipomas are slow growing, uncommon benign

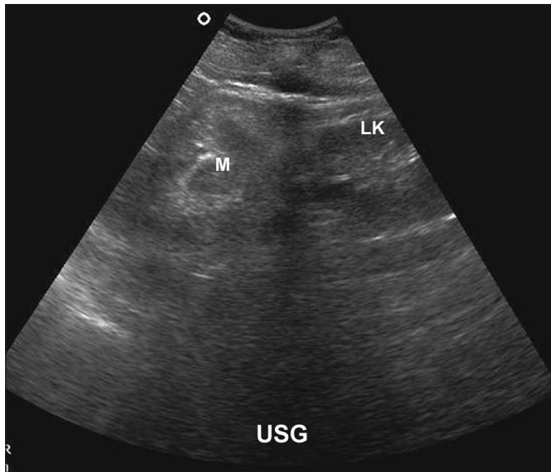


Figure 1a: Ultrasound abdomen reveals a large, well encapsulated, heterogeneous left suprarenal mass

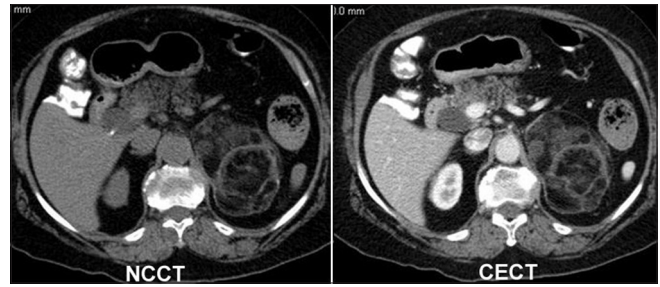


Figure 1b: On axial plain and contrast-enhanced computed tomography images through upper abdomen, the mass appears to be well circumscribed and predominantly of fat density (-30 to -90 HU). Heterogeneous enhancement pattern is due to presence of enhancing soft tissue component (myeloid tissue) and septations. Few tiny flecks of calcifications are also visible in the lateral wall of the mass



Figure 1c: Coronal and sagittal computed tomography images confirm the adrenal origin of the mass. Left kidney is displaced inferiorly by the mass

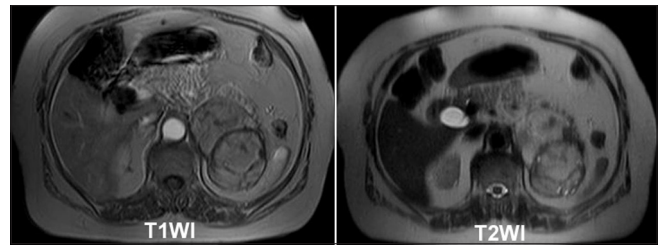


Figure 1d: On T1- and T2-weighted magnetic resonance imaging, the mass is predominantly of high signal intensity with interspersed low to intermediate signal intensity hematopoietic elements and hypointense septae. Small linear hyperintense foci posteriorly represent intratumoral bleed



Figure 1e: Fat-saturated T2-weighted gradient echo sequences demonstrate loss of signal intensity in the fatty part of the mass, while the hematopoietic tissue and hemorrhage represent persistent areas of increased signal intensity

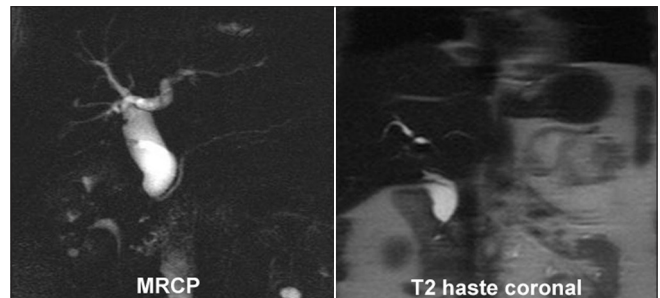


Figure 1f: Magnetic resonance cholangiopancreatography (MRCP) shows moderately dilated CBD and central intrahepatic biliary radicals. T2-haste coronal image demonstrates benign stricture at the ampullary end of common bile duct

tumors of adrenal cortex. According to a recent study these tumors account for 2.6% of the primary adrenal masses.^[8] The tumor has similar sex predilection. Although most frequently encountered in individuals between fifth and seventh decades,

they are also reported in children.^[9] They are usually small (<5 cm), but giant myelolipomas reaching up to 34 cm are also known to occur.^[1] Unilateral myelolipomas are more common; only 10 cases of bilateral myelolipomas have been documented worldwide till date.^[10,11] Myelolipomas are non functional tumors but have been known to coexist with other endocrine disorders of adrenal gland such as Conn's syndrome, Cushing's syndrome, congenital adrenal hyperplasia, adenoma and pheochromocytoma.^[12]

The etiopathogenesis of adrenal myelolipoma is controversial. Although, many theories have been proposed, the most accepted hypothesis is metaplasia of reticuloendothelial cells of blood

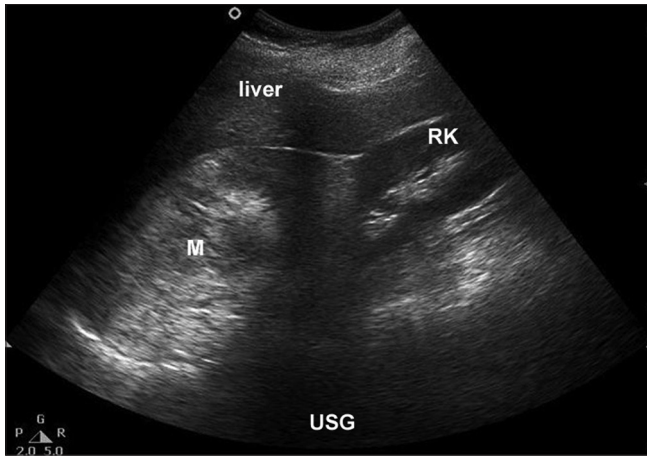


Figure 2a: Ultrasound abdomen shows a large, well encapsulated, solid echogenic mass between the liver and right kidney (RK)

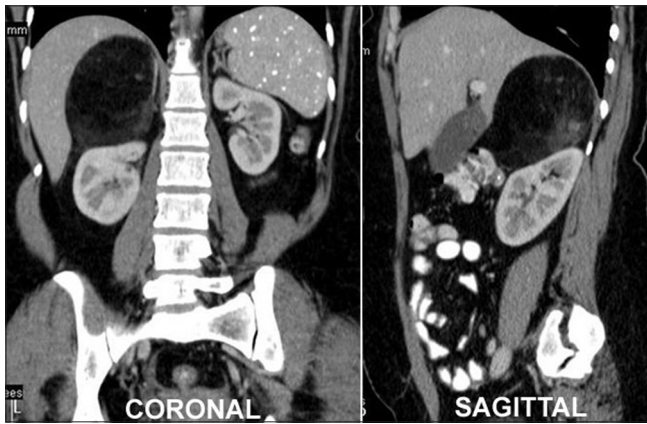


Figure 2c: Suprarenal nature of the mass is confirmed by coronal and sagittal computed tomography images, which also demonstrate inferior displacement of right kidney. Left adrenal gland and bilateral kidneys appears normal



Figure 2e: T2W-fat-suppression image shows loss of signal from large fatty portion of the mass, and persistence of increased signal intensity in areas occupied by hematopoietic tissue and intratumoral bleed

capillaries in the adrenal gland in response to stimuli such as chronic infection, stress or necrosis. Since histologically myelolipomas are composed of fat and hematopoietic tissues,

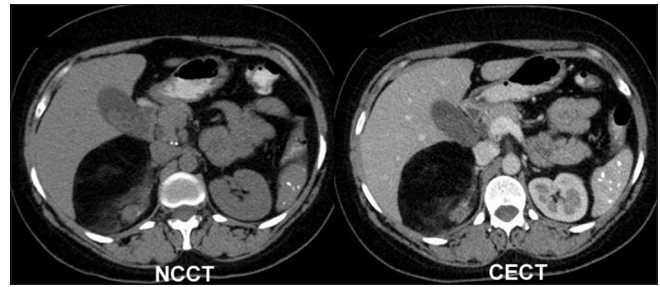


Figure 2b: Plain and contrast computed tomography abdomen reveals a well circumscribed right suprarenal mass predominantly of fat density (-50 to -90 HU) with small peripheral enhancing soft tissue component on the postero-medial aspect of the mass. A punctate focal calcified area is also seen adjacent to the soft tissue component. Incidental note is made of multiple small, calcified granulomatous lesions randomly distributed within the splenic parenchyma

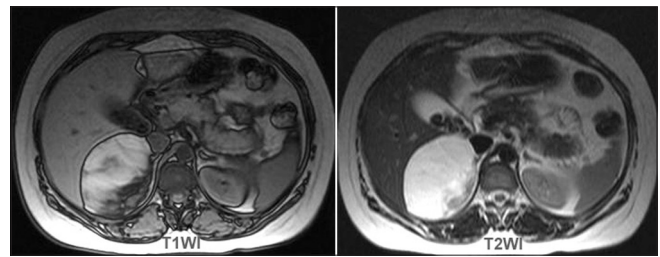


Figure 2d: Magnetic resonance imaging abdomen reveals predominantly hyperintense mass on T1-and T2- weighted images. Multiple gall bladder calculi are also evident

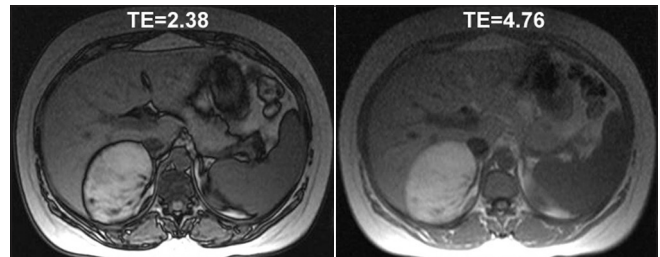


Figure 2f: Double echo chemical shift gradient echo imaging demonstrates few small focal areas of signal loss in out-of-phase (echo time=2.38) image as compared to in-phase image (echo time=4.76), consistent with the presence of microscopic (intracellular) fat, while the major portion of the mass shows high signal intensity consistent with the presence of macroscopic (extracellular) fat

the theoretical possibility of pathogenesis due to bone marrow embolization also exists.^[2]

Although most adrenal myelolipomas are usually asymptomatic, abdominal/flank pain and an abdominal mass are the most common presenting symptoms. The compression of neighboring structures by large tumors (mass effect), infection, tumor necrosis and intratumoral hemorrhage are the main complications associated with it. Intestinal obstruction and spontaneous retroperitoneal hemorrhage are rare complications usually encountered with tumors >10 cm in diameter.^[1,13,14] A remote possibility of malignant change has also been reported with these rare neoplasms.^[11] In our cases, large tumor size and

intratumoral hemorrhages were probably responsible for the patient's symptoms.

Most myelolipomas are asymptomatic and discovered incidentally on abdominal imaging for some other indications; however, with the advent of cross sectional imaging, the diagnosis of this neoplasm has improved remarkably, both in asymptomatic and symptomatic patients. The imaging features of myelolipoma depend on the varying proportion of fat, hematopoietic tissues, hemorrhage or calcification present within it. Calcification may be seen in these masses in around 27% of cases.^[8]

Although a predominantly hyperechoic adrenal mass strongly suggests the diagnosis of myelolipoma, ultrasonography is not specific and computed tomography (CT) or magnetic resonance imaging (MRI) should be obtained for further evaluation. Smaller lesions less than 2 cm are usually masked by adjacent echogenic retroperitoneal fat and are difficult to be diagnosed by ultrasound (USG). The presence of hemorrhage or calcification further alters the sonographic appearances.^[15-17]

CT is the imaging modality of choice for these tumors because of its sensitivity, and easy availability. A well defined fatty adrenal mass with negative attenuation value (-30 to -100 HU) is almost diagnostic of myelolipoma. The presence of hematopoietic elements, intratumoral bleed, calcification and adrenal tissue accounts for non-fat density and heterogeneity. If the mass contains more hematopoietic tissue and little fat, it has soft tissue attenuation and mimics adrenal adenoma or retroperitoneal liposarcoma. After contrast administration, the myeloid component of the mass enhances. If peritumoral hemorrhage has occurred, high or low-attenuation fluid may be present, depending on the age of the blood.^[15-17]

MRI is complimentary to CT in the workup of patients with adrenal masses. Its multiplanar capability is helpful in confirming the adrenal origin of the mass. Furthermore, MRI also accurately depicts both microscopic and macroscopic fat using chemical shift imaging and explicit (chemically selective) fat saturation technique, respectively. On explicit fat-saturated sequences, the macroscopic fat shows loss of signal intensity, whereas the myeloid tissue or hemorrhage results in persistent areas of high signal intensity. Double echo chemical shift phase selective gradient echo imaging detects microscopic (intracellular) fat that is present in the same voxel as the water proton. Two scans are obtained through the mass using parameters that are identical in all respect, except for echo time (TE), which is selected in such a manner that the fat and water protons are either in-phase (TE=4.76) or out-of-phase (TE=2.38) in 1.5 Tesla machine. In out-of-phase sequences, the microscopic fat shows signal loss, since fat and water protons in the same voxel cancel out each

other. Thus in an adrenal myelolipoma, explicit fat saturation sequence shows more signal loss than chemical shift imaging, since the presence of macroscopic fat is typical of a myelolipoma. In contrast, a lipid- rich adrenal adenoma will show a greater loss of signal intensity on chemical shift imaging as it contains microscopic fat.^[18,19]

Since myelolipomas contain fat and myeloid tissues in different proportion, a definitive diagnosis using cross sectional imaging may be difficult, although rarely so, if only a small amount of fat is present. Under such circumstances USG or CT guided percutaneous fine needle aspiration cytology (FNAC) confirms the diagnosis. The presence of mature adipose cells admixed with hematopoietic tissues without cellular atypia is diagnostic of myelolipoma.^[8,13] In our cases, we did not obtain FNAC because imaging findings were highly suggestive of myelolipoma.

The differential diagnosis includes retroperitoneal lipoma or liposarcoma, upper pole renal angiomyolipoma, and retroperitoneal teratoma.

The management of myelolipomas is usually individualized. Asymptomatic, small myelolipomas (<5 cm) are treated conservatively with 6-12 months interval follow-up with ultrasound or CT; whereas symptomatic and large tumors (>10 cm) should be extirpated because of risk of malignant change and spontaneous rupture with retroperitoneal bleed.^[11] With the advent of minimally invasive surgery, laparoscopic adrenalectomy has become a standard treatment for both functioning and non-functioning adrenal tumors.

This procedure has shown a considerable decrease in the perioperative morbidity, hospital stay and convalescence after the procedure when compared with open surgery.^[20]

Both of our patients were obese and had adrenal myelolipoma as well as gall stones. The association of obesity with adrenal myelolipoma and cholelithiasis is well documented; however, the occurrence of adrenal myelolipoma and cholelithiasis is extremely rare. The co-occurrence of adrenal myelolipoma and cholelithiasis may be an incidental finding or might have common pathophysiologic basis. The accumulation of additional cases is needed to throw further light on this correlation. Further, awareness of the imaging findings of myelolipoma helps to exclude the unwanted surgical exploration or extensive surgery.

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