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Case Report

A rare case of co-existing adrenal and pelvic myelolipomas

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ABSTRACT

We are reporting a case of co-existing left sided pelvic and right sided adrenal myelolipomas in a 68-year-old male patient. Both lesions were incidentally discovered on CT whilst undergoing a staging scan for suspected urinary bladder cancer. The patient had a background of hypertension and type 2 diabetes. Contrast enhanced CT scan showed both lesions to be of mixed fat and soft tissue density. Given the size, location, and attenuation characteristics of the pelvic mass, retroperitoneal liposarcoma was thought to be a differential diagnosis, prompting the decision for an elective CT-guided biopsy. Both masses were targeted successfully using core biopsy needles. Subsequently, histopathology results for both the right adrenal and the left pelvic masses showed features compatible with myelolipomas. The right retroperitoneal mass was compatible with an adrenal myelolipoma and left pelvic mass was deemed as an extra-adrenal myelolipoma (EAML).

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Introduction

Myelolipomas are rare lesions composed of mature adipose tissue combined with myeloid and erythroid cells. When contained within the adrenal gland, the presence of macroscopic fat on imaging is pathognomonic of an adrenal myelolipoma, which can be confidently diagnosed on cross sectional imaging alone. On the other hand, extra adrenal myelolipomas (EAMLs) are very rare tumors and can be difficult to differenti-

ate from malignant retroperitoneal lesions such as liposarcomas, hence, requiring histopathological confirmation. We are reporting a rare case where an adrenal myelolipoma and an EAML co-existed in a 68-year-old male with a background of hypertension and type 2 diabetes.

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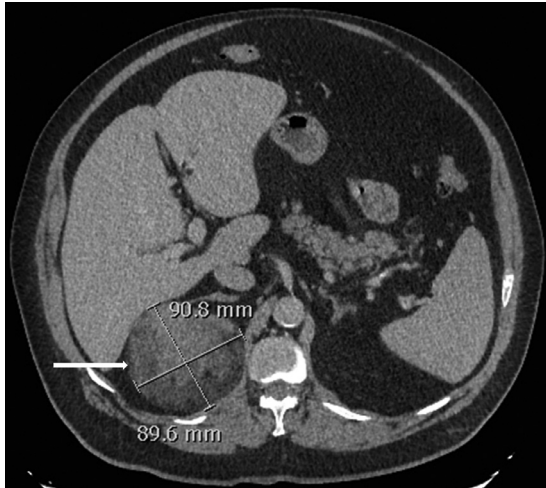


Fig. 1 – Axial section of contrast enhanced CT scan of abdomen showing a 9.1 x 9.0 cm mixed attenuation soft tissue mass with fatty components, arising from right adrenal (white arrow).

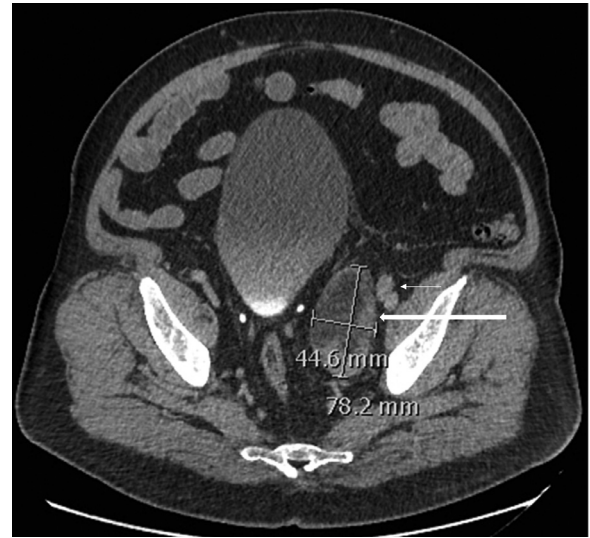


Fig. 2 – Axial section of contrast enhanced CT scan of pelvis showing a 7.8 x 4.5 cm mixed attenuation soft tissue mass with fatty components (thick white arrow), medial to the left external iliac vessels (thin white arrow).

Discussion

Adrenal lesions greater than 1 cm in size on cross sectional imaging are referred to as ‘incidentalomas’ [1]. With the advancements in modern day imaging they are now detected more frequently. Benign adrenal lesions are found in 3%-10% of the general population, of which non-functioning adrenal adenomas are the majority [2]. Other primary adrenal lesions which are encountered include adrenocortical carcinomas, cysts, myelolipomas, and pheochromocytomas.

Edgar von Gierke in 1905 was the first person to describe myelolipomas [3]. The underlying aetiology has still not completely been established with several theories existing including the fact that they could be remnants of fetal bone marrow, embolism of bone marrow cells, and hyperplasia of heterotopic reticulum cells [4,5]. Although, the diagnosis of an adrenal myelolipoma can be confidently made based on computed tomography (CT) or magnetic resonance imaging (MRI) features alone, because it is the only known entity composed of macroscopic fat in this location [6]. In the case of this patient, cross sectional imaging was accurately used to make a diagnosis of an adrenal myelolipoma (Fig. 1) but due to the size of the mass, a decision was made to conduct a biopsy to provide further clarity on the lesion. The presence of intralesional macroscopic fat is a very characteristic feature for myelolipomas. When using CT, a negative attenuation (between -30 and -90 Hounsfield units) is generated by fatty elements and myeloid components appear higher in attenuation. An encapsulated mass, with macroscopic fatty tissue intermixed with areas of soft tissue that does not invade adjacent structures, strongly suggests the diagnosis of a myelolipoma. As adrenal myelolipomas are asymptomatic lesions of the adrenal cortex and have no malignant potential, surgical management is usually not required. Surgery is generally only recommended for symptomatic patients or when the growth of the mass is

progressive. Small asymptomatic tumors (less than 4 cm) have been suggested to be monitored routinely due to the minimal risk of spontaneous rupture or bleeding [7]. There is no clear consensus about lesion size for which surgery should be considered, with some reporting sizes greater than 4 cm or 7 cm as measurements for which surgery should be done due to higher risks of complications [8,9].

Moreover, approximately 15% of myelolipomas arise as extra adrenal lesions, most commonly in the presacral region [10]. A summary of 37 cases of EAMLs in one report identified the following sites: the presacral space ($n=15$, 40.5%), the retroperitoneum ($n=8$, 21.6%), the thoracic cavity ($n=5$, 13.5%), the pelvic cavity ($n=3$, 8.1%), the kidneys ($n=2$, 5.4%), the stomach ($n=1$, 2.7%), the liver ($n=1$, 2.7%), the bladder ($n=1$, 2.7%), and multiple lesions ($n=1$, 2.7%) [11]. EAMLs are very rare, regardless of the location they are identified at. Since their histological composition is similar to adrenal myelolipomas, they share similar appearances on imaging (Fig. 2).

EAMLs are described as well circumscribed round masses with a size recorded up to 26 cm [9]. A review of the literature has shown that they are more likely to be discovered in middle aged females, with a female to male predominance of 2:1 [12,13]. Akin to myelolipomas, they are often asymptomatic with a good prognosis, hence they may be followed up by CT or MRI imaging. EAMLs have the potential to grow causing symptoms, such as distension, anorexia, and pain. The tumoral mass effect can influence surrounding organs, for example, renal failure from perirenal lesions, sciatic pain or urinary retention in presacral lesions or numbness, and gait disturbance in the case of intraspinal lesions [14,15].

Nevertheless, EAMLs being rare tumors, with limited number of clinical cases reported, can be difficult to establish from imaging alone. Calcification can be used to identify 20% of adrenal myelolipomas and in only a few cases of EAMLs [17,18]. Calcification is hyperdense, often punctate, and it may

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