

A Rare Case of Ruptured Giant Adrenal Myelolipoma Presenting with Spontaneous Intra-Abdominal Haemorrhage: A Case Report

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Abstract

Adrenal myelolipoma is a rare benign neoplasm of mesenchymal origin composed of mature fatty tissue and bone marrow elements. It is a non-functioning benign tumor, usually asymptomatic and commonly detected incidentally during evaluation for unrelated symptoms; hence, it is usually referred to as an “incidentaloma”. Large myelolipomas can cause mass effects and vague abdominal pain, spontaneous tumor rupture with massive hemorrhage is a more dramatic manifestation, though, it is an uncommon entity. We report the case of a 41-year-old anemic male patient who underwent an abdominal CT study and a huge hemorrhagic adrenal myelolipoma was detected to reiterate the importance of recognizing the characteristic radiological features of myelolipoma to accurately diagnose these tumors and emphasize the inclusion of myelolipoma in the lists of differential diagnosis of causes of spontaneous intra-abdominal hemorrhages.

Keywords: Intra-abdominal haemorrhage, Myelolipoma, Ruptured

Introduction

Adrenal myelolipoma is a rare benign neoplasm of mesenchymal origin composed of mature fatty tissue and bone marrow elements (Lam, 2017). This disease entity was first described by the German pathologist Edgar von Gierke in 1905, (Kaifer *et al.*, 2020) but the name myelolipoma was coined by the French pathologist Charle Obenling in 1929 (Pillay, 2018; Haguenu, 2003). In reported autopsy studies the incidence of myelolipoma is 0.2% in the general population (Lui *et al.*, 2017). However, in recent times, adrenal myelolipomas are increasingly reported due to the widespread use of abdominal imaging, such as ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI) (Lam and Lo, 2001; Khater and Khauli, 2011).

Adrenal myelolipoma is a non-functioning benign tumor, usually asymptomatic and commonly detected incidentally during evaluation for unrelated symptoms; hence, it is usually referred to as an "incidentaloma" (Grumbach *et al.*, 2003). Nonetheless, these tumours may be associated with an endocrine disorder such as Cushing's disease, Addison's disease, hyperaldosteronism, and obesity of unknown aetiology. They are usually unilateral and occur more often on the right (Kenney *et al.*, 1998; Murakami *et al.*, 1988). Albeit huge myelolipomas have been reported in the literature, they uncommonly measure more than 5cm (Yildiz *et al.*, 2000).

Large myelolipomas can cause mass effect and vague abdominal pain, spontaneous tumour rupture with massive haemorrhage is a more dramatic manifestation; though, it is an uncommon entity (Lui *et al.*, 2017; Patel *et al.*, 2006). A Radiologic diagnosis of myelolipoma can be made with ultrasound, computed tomography and magnetic resonance imaging because of their characteristic imaging appearances (Khater and Khauli, 2011; Kraimps *et al.*, 1992). We report the case of a 41-year old anaemic male patient who underwent abdominal CT study and huge haemorrhagic adrenal myelolipoma was detected to reiterate the importance of recognizing the characteristic radiological features of myelolipoma to accurately diagnose these tumours and emphasize the inclusion of myelolipoma in the list of differential diagnosis of causes of spontaneous intra-abdominal haemorrhages.

Case report

A 41-year old male patient was referred from emergency department to the department of radiology for an abdominal computed tomography (CT) scan following a history of sudden onset dull right abdominal pain and syncope. There was no history of trauma. Patient was said to be healthy until he developed the above symptoms. On physical examination he was pale and serial haemoglobin estimation revealed continuous reduction indicating persistent bleeding. These findings raised the suspicion of possible internal

haemorrhage and consequently he was referred for an abdominal CT scan with contrast. The scan revealed a large well-defined rounded retroperitoneal right suprarenal mass with heterogeneous density. The mass measures 11.4 x 11.6cm and shows areas of gross fat content with low density mixed with otherwise soft tissue density components. Post contrast images show mild enhancement of the soft tissue elements in the lesion.

The mass displaces the ipsilateral kidney downwards. Perinephric hyperdense collection (relative to background mesenteric fat) tracking down the paracolic gutter into the pelvis is evident. (Figures 1- 4)

The left kidney and adrenal are unremarkable.

A diagnosis of right adrenal haemorrhagic myelolipoma was made and consequently patient was taken over by the urology team for expert/definitive management.



Figure 1. Non-contrast axial computed tomography of the abdomen showing a huge well-defined rounded retroperitoneal right suprarenal mass with fat and soft tissue components (arrow)

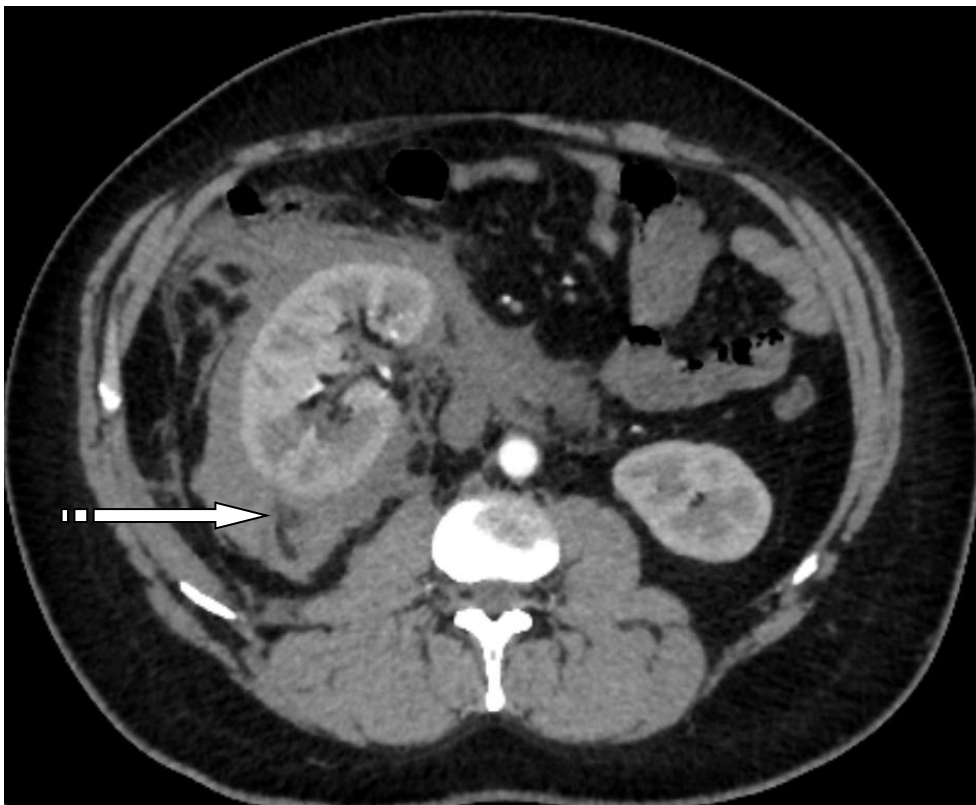


Figure 2. Contrast enhanced axial computed tomography of the abdomen showing haemorrhage around the kidney (striped arrow)

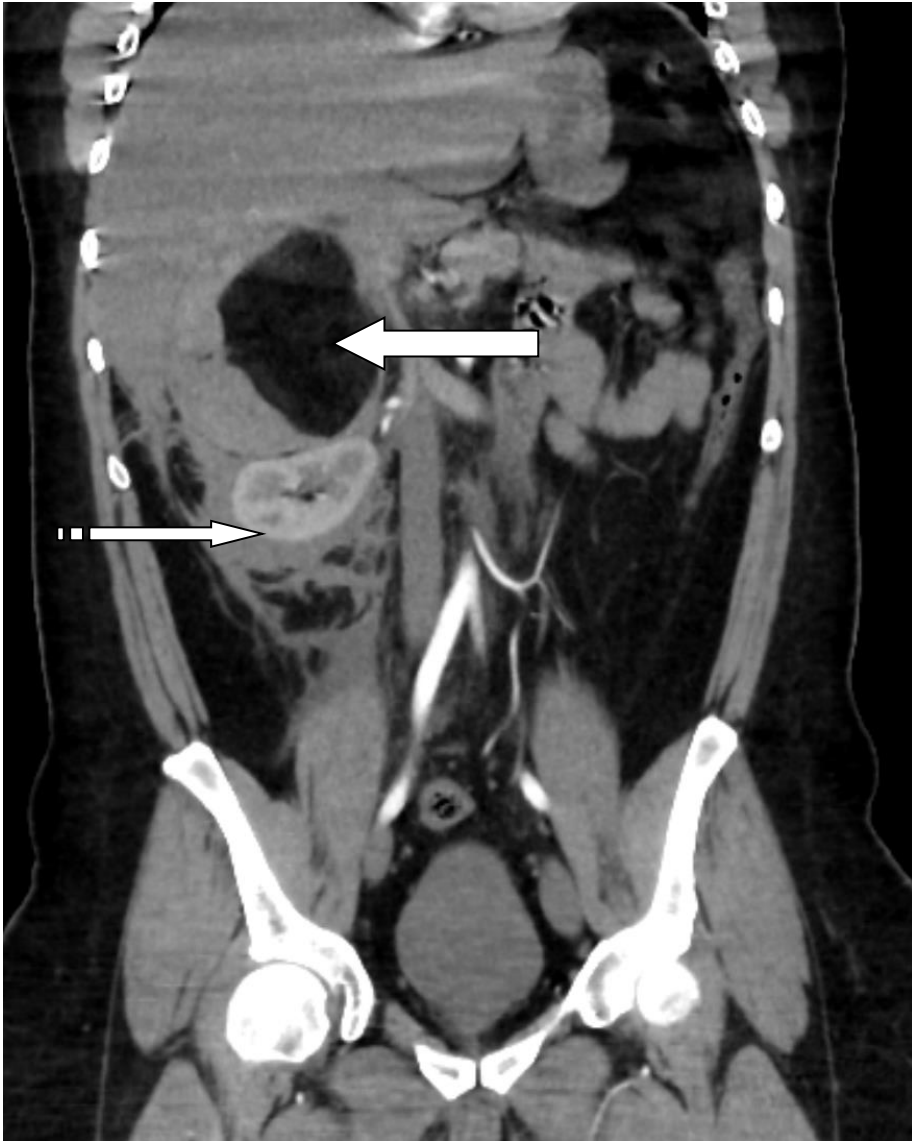


Figure 3. Contrast enhanced coronal reformatted computed tomography of the abdomen showing a huge well-defined rounded retroperitoneal right suprarenal mass with minimal enhancement of the soft tissue component (arrow) and haemorrhage around kidney (striped arrow)



Figure 4. Contrast enhanced sagittal reformatted computed tomography of the abdomen showing a huge well-defined rounded retroperitoneal right suprarenal mass with minimal enhancement of the soft tissue component (arrow) and haemorrhage around kidney (striped arrow)

Discussion

Myelolipoma is a tumour with nebulous pathogenesis and the second most common tumour of the adrenal gland (Decmann *et al.*, 2018). Most myelolipomas are incidentally discovered on cross-sectional imaging. Large myelolipomas are more likely to cause mass effect symptoms, have haemorrhagic changes and undergo resection. Spontaneous tumour rupture

can occur more commonly in large myelolipomas measuring more than 10cm (Liu *et al.*, 2017; Decmann *et al.*, 2018).

Cases of spontaneous rupture of myelolipoma have been reported. The case under review is a typical case of spontaneous rupture, occurring in large tumours. Our patient's tumour measures 11.4 x 11.6cm and ruptured spontaneously resulting in massive intra-abdominal haemorrhage with syncopal attack. Similarly, Liu *et al* (2017), reported a case of spontaneous rupture of myelolipoma with shock in a male patient and subsequent en bloc surgical excision.

Retroperitoneal haemorrhages resulting from spontaneous rupture of myelolipoma are extremely uncommon and occur commonly with large tumours. The CT imaging appearance of the lesion is usually characteristic but with haemorrhage findings may be confusing. CT typically shows a well-margined adrenal lesion with fat-containing components, though lesions with mostly fat may be difficult to separate from surrounding retroperitoneal fat. The quantity of fat in myelolipoma varies and may vary from a few small regions in an otherwise mostly soft tissue density mass (10%) to masses made up of roughly equal components of fat and soft tissue (50%) or almost completely composed of fatty tissue (40%)(Joseph, 2006). Punctate calcification may be seen in about 25-30% of cases (Joseph, 2006; Craig *et al.*, 2009). Haemorrhage in the lesion is seen as areas of high CT attenuation and is commonly seen in large lesions (Routhier *et al.*, 2009).

Radiology plays a significant role in differentiation of adrenal tumours and cross-sectional imaging is the mainstay of imaging for identifying and assessing these tumours. Ultrasonography, CT and MRI are all effective in diagnosing more than 90% of adrenal myelolipoma based on identification of fat, with CT scan being the most sensitive(Kenney *et al.*, 1998; Daneshmand and Quek,2006). Myelolipoma is easy to recognize on CT or MRI because they contain areas of macroscopic fat. Adrenal myelolipomas are benign lesions with no currently recognized malignant potential (Kenney *et al.*, 1998). Small myelolipoma with characteristic imaging features does not require treatment or follow-up. In contrast, if imaging findings are equivocal, a percutaneous biopsy can be considered. Surgical resection should also be considered in symptomatic patients with large tumours, evidence of haemorrhage or tumour growth.

Conclusion

Adrenal myelolipoma is usually asymptomatic rare tumours and can be diagnosed based on their imaging characteristics. Patients may present with radiographic haemorrhagic imaging changes, though, adverse events such as tumour rupture and acute retroperitoneal haemorrhage are extremely rare. Good knowledge of the imaging signature of myelolipoma is required to

establish the accurate diagnosis without exposing the majority of patients to the burden of clinical workup, interventions and imaging follow-up. The differential diagnoses of spontaneous intra-abdominal haemorrhage are many and myelolipoma, although rare, should be considered as well.

Conflict of interest: None declared among the authors.

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