

Adrenal Myelolipoma Presenting with Spontaneous Rupture and Retroperitoneal Hemorrhage: A Case Report

자발성 파열 및 후복막강 출혈로 발견된 부신 골수지방종: 증례 보고

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Adrenal myelolipoma is a rare, benign, and nonfunctioning tumor, which is generally small and asymptomatic and incidentally discovered at autopsy or on radiological examination. In rare cases, large tumors can produce symptoms due to their mass effect or spontaneous rupture with hemorrhage. We reported a case of a 36-year-old man who presented with retroperitoneal hemorrhage due to spontaneous rupture of a large adrenal myelolipoma discovered using multiple imaging modalities.

Index terms

Myelolipoma
Adrenal Gland
Hemorrhage
Rupture
Multidetector Computed Tomography

Adrenal myelolipoma is a rare, benign tumor that is commonly small and non-functioning. It generally comprises adipocytes and mature hematopoietic elements. It occurs unilaterally and originates from the adrenal cortices in most cases (1). Most adrenal myelolipomas are asymptomatic and detected incidentally by imaging modalities such as ultrasonography (US), computed tomography (CT), and magnetic resonance imaging (MRI) (1, 2). However, in rare cases, they may be symptomatic when the tumor size is large enough to compress adjacent structures or when tumor rupture causes hemorrhage (1, 3). Spontaneous retroperitoneal hemorrhaging from an adrenal myelolipoma is uncommon (1). Herein, we reported a case of adrenal myelolipoma with spontaneous retroperitoneal hemorrhage and briefly reviewed the relevant literature.

CASE REPORT

A 36-year-old male patient visited a local clinic with the complaint of right flank pain for 10 days prior; he was referred to our hospital for further evaluation and treatment. At presentation, his blood pressure was 150/80 mm Hg. Regular checkups for the previous 8 years had indicated high blood pressure for which, he had never taken hypertensive medication. Laboratory tests indicated that hemoglobin, aspartate aminotransferase, and alanine transaminase levels were 12.9 g/dL, 122 IU/L, and 145 IU/L, respectively. Renal function test results were unremarkable.

US and contrast-enhanced CT performed a week earlier at the outside hospital showed a right retroperitoneal mass. On US, a very large, well-circumscribed, heterogeneous, echogenic mass was visible in the right suprarenal area (Fig. 1A). Axial pre-contrast CT showed a heterogeneous, 11.9 × 10 cm diameter, fat-containing mass between the right kidney and liver (Fig. 1B). The

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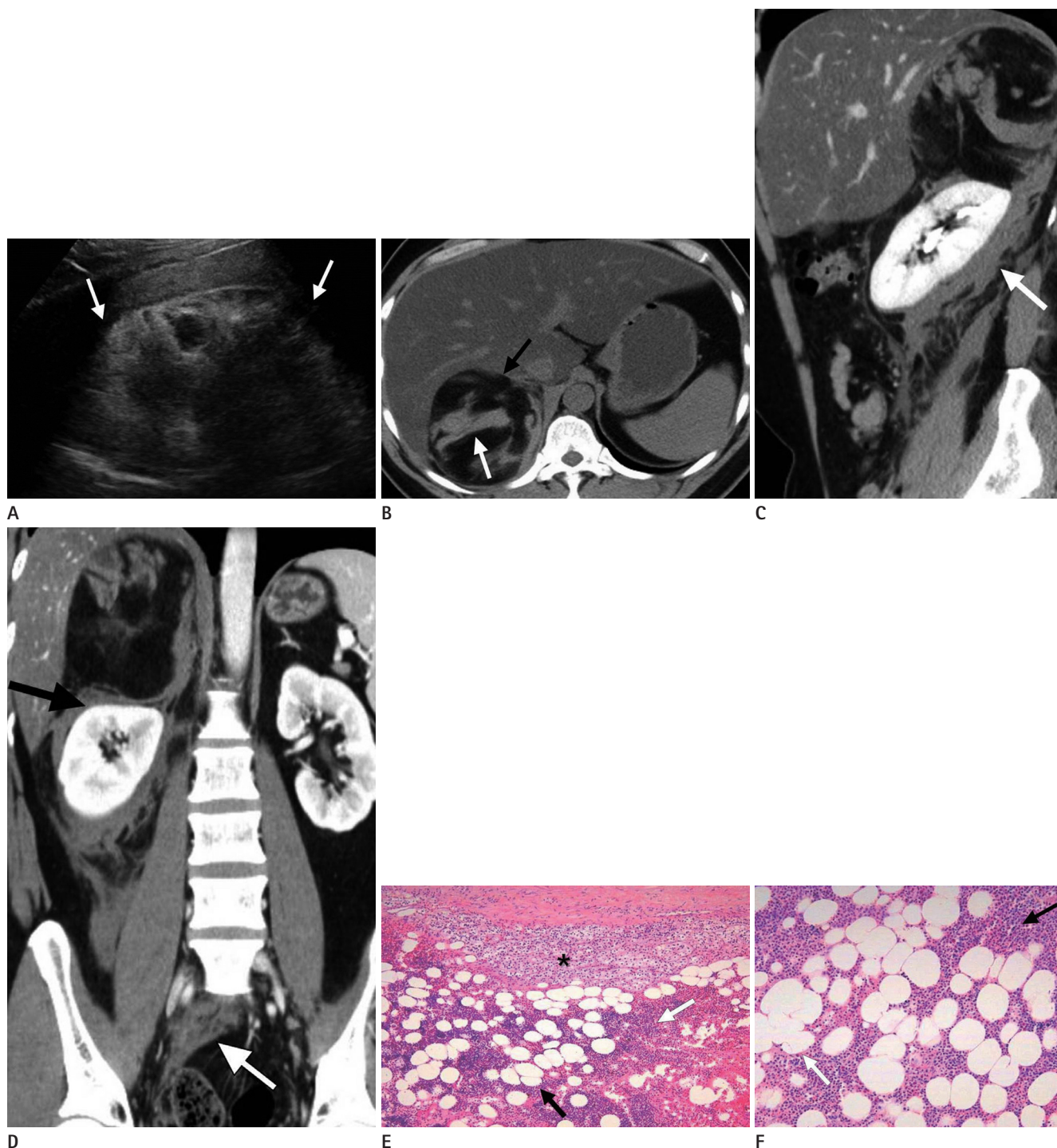


Fig. 1. A 36-year-old man with a ruptured adrenal myelolipoma.

A. Ultrasound image shows a well-circumscribed supranrenal mass with internal heterogeneous echogenicity (arrows).

B. Axial pre-contrast computed tomography (CT) image shows a well-defined mass containing intratumoral macroscopic fat (black arrow) in the right adrenal bed with an internal high attenuated portion (white arrow), suggestive of hemorrhage.

C. Sagittal reformatted contrast-enhanced CT image shows hemorrhage (arrow) along the posterior perinephric space.

D. Coronal reformatted contrast-enhanced CT image shows supranrenal location of the right retroperitoneal mass with the inferiorly displaced right kidney (black arrow). The peritumoral hemorrhage extends into the right pelvic retroperitoneum (white arrow).

E. Photomicrograph of the tumor mass (hematoxylin & eosin staining, $\times 100$) shows a mixture of mature adipose tissues (black arrow) and hematopoietic elements (white arrow), pushing the adrenal cortex (asterisk).

F. Higher magnification of the tumor (hematoxylin & eosin staining, $\times 200$) shows bone marrow cells (black arrow) and adipocytes (white arrow).

right adrenal gland could not be identified. The CT attenuation values of the tumor were -110 to -95 Hounsfield units (HU) and 45–60 HU for areas in the central portion of the mass, suggestive of recent hemorrhage. On sagittal reformatted contrast-enhanced CT, high attenuation areas were observed in the posterior of the mass extending inferiorly and partially around the right kidney, concordant with both intratumoral and peritumoral hemorrhages (Fig. 1C). Coronal and sagittal reformatted CT images showed an inferiorly displaced right kidney but intact upper pole (Fig. 1C, D). Retroperitoneal bleeding due to spontaneous rupture of a large adrenal myelolipoma was suspected on the basis of these findings.

The tumor was symptomatic and required differentiation from other retroperitoneal tumors; hence, an operation was performed a week later. The resected specimen revealed a well-circumscribed tumor with multiple hemorrhagic areas and fat tissue, measuring 12 × 13 × 4 cm. The tumor comprised mature adipose tissue, focal hematopoietic cells, and hemorrhagic and necrotic areas (Fig. 1E, F). These findings were histologically compatible with adrenal myelolipoma. The patient recovered without major complications and was discharged 2 weeks postoperatively.

DISCUSSION

Adrenal myelolipoma is an uncommon and asymptomatic tumor generally found incidentally in < 1% of the general population via autopsy or imaging performed for other purposes (4). It is a benign tumor comprising mature adipocytes interspersed with hematopoietic cells, resembling bone marrow (1, 5). Adrenocortical metaplasia secondary to necrosis, infection, or stress is a possible precipitating factor in the development of adrenal myelolipoma (1, 5, 6).

The incidence of adrenal myelolipoma is similar between the sexes (7). It occurs in patients aged 40–70 years, although cases have been reported in all age groups (8). Although adrenal myelolipomas are usually small, they can range from several mm to > 30 cm in size (4). Of the 64 adrenal myelolipomas identified in a retrospective review of CT images, 9 (14%) were > 4 cm (9). Adrenal myelolipomas are not hormonally functional. However, very rare myelolipomatous foci are associated with adrenal functional disorders such as Cushing syndrome (1, 3). These are usually asymptomatic unless they grow large (> 5 cm) enough

to exert mass effect or cause internal hemorrhaging (1, 3). Therefore, tumor size exceeding 7 cm is an indication for removal owing to mass effect and increased risk of bleeding (3). Hypertension may occur coincidentally or due to direct compression of the adjacent kidney or renal artery (7). Sudden onset pain, nausea, and vomiting are the commonly reported symptoms of acute internal bleeding from the tumor (2). In the present case, the patient complained of sudden onset right flank pain due to retroperitoneal hemorrhage.

Imaging is an integral part of the investigation of patients with symptomatic myelolipomas. On US, the appearance varies depending on the tissue component of the tumor i.e., it appears hyperechoic if it contains more fatty tissue, and hypoechoic if myeloid cells are predominant (4). The typical appearance of an adrenal myelolipoma on non-contrast CT images is an adrenal mass with mixed regions of soft tissue (20–50 HU) and fat (-115 to -30 HU) attenuation. Focal calcifications are commonly detected and may be single, multiple, or peripheral in distribution (7). After intravenous contrast medium administration, soft-tissue enhancement may be absent or significant (up to a 60 HU increase) according to the hematopoietic elements (7). Hemorrhage within the mass may enhance soft-tissue density and rapidly increase the tumor size (6). On MRI, the fatty component of adrenal myelolipomas exhibits increased signal intensity on T1-weighted images and decreased signal intensity on fat-saturated T2-weighted images. Opposed-phase imaging demonstrates low signal intensity in the voxels containing both fat tissue and water (3).

Differential diagnostic considerations for adrenal myelolipoma include other fat-containing retroperitoneal masses such as exophytic renal angiomyolipoma, retroperitoneal liposarcoma, and retroperitoneal teratoma (8). Invasive tissue sampling is rarely required for a definitive diagnosis. However, if imaging results are unclear, percutaneous needle biopsy may be required. A tissue sample exhibiting mature adipocytes and hematopoietic elements is suggestive for diagnosis (4, 5).

Adrenal tumor associated with spontaneous hemorrhage is a very rare condition that includes metastases, pheochromocytomas and adrenocortical cancers (10). In case of benign tumors such as adrenal myelolipoma, spontaneous hemorrhage is even rarer with fewer than 15 cases reported, including the present case (1). In the present case, despite the retroperitoneal hemor-

rhage, there were no remarkable abnormalities on laboratory examination or life-threatening conditions besides right flank pain. However, large myelolipomas with acute hemorrhaging may present with anemia and hemodynamic compromise. Therefore, diagnosis as well as management of ruptured tumors should be performed earlier if a large hemorrhagic suprarenal mass is detected by imaging. Even small myelolipomas that are asymptomatic and < 5 cm in size, should be followed-up regularly with imaging such as CT or MRI in order to prevent an emergent condition (4, 6).

Although adrenal myelolipoma presenting with spontaneous rupture is relatively rare, it should be suspected in patients with a large suprarenal mass containing fat and hemorrhages. Knowledge of imaging findings of ruptured adrenal myelolipomas is crucial for improving diagnostic accuracy and reducing mortality caused by spontaneous bleeding.

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자발성 파열 및 후복막강 출혈로 발견된 부신 골수지방종: 증례 보고

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부신의 골수지방종은 흔하지 않은 비기능성 양성종양이며, 일반적으로 크기가 작고 증상이 없어 부검이나 영상의학 검사에서 우연히 발견된다. 드문 경우에서 크기가 큰 골수지방종이 증상을 유발하기도 하는데, 이는 종양 자체에 의한 종괴효과나 종양의 자발성 파열에 의한 출혈 때문이다. 본 저자들은 36세 남성에서 여러 영상기법에 의해 발견된 자발성 파열에 따른 후복막강 출혈을 동반한 큰 부신 골수지방종 증례를 보고하고자 한다.

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