Adrenal Myelolipoma With Spontaneous Hemorrhage

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Adrenal myelolipoma is a rare disease entity that is usually asymptomatic and found in elderly patients. We report the case of a young female patient with this tumor who presented with spontaneous retroperitoneal hemorrhage. The 32-year-old woman suffered from acute onset of right loin pain and visited our emergency department. She had no specific history of any medical disease. A bulging mass was palpable in the right upper area of the abdomen. Other than anemia, complete blood count was normal. Serum adrenocorticotropic hormone, plasma renin activity, aldosterone, and 24-hour urine vanillylmandelic acid levels were all within normal limits. Abdominal computed tomography showed retroperitoneal hemorrhage of a large tumor with fat density in the right suprarenal region. The tumor was surgically removed. Pathological findings revealed a proliferation of adipocytes and myeloid tissues, which were compatible with adrenal myelolipoma. She had a smooth recovery, and was stable at the 6-month postoperative follow-up.

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1. Introduction

Adrenal myelolipoma is a rare, benign tumor that is composed of mature fat tissue and hematopoietic elements.1 The tumor is most frequently located in the adrenal gland, but it may also occur as an isolated soft tissue mass at various sites, especially in the pelvic region. Myelolipomas are generally asymptomatic due to their small size, and usually found incidentally during radiologic examination or at autopsy. Larger tumors may be associated with abdominal pain, constipation or vomiting. Myelolipomas are usually diagnosed in patients in their fifth to seventh decades of life.2 We report a young female patient with adrenal myelolipoma who presented with spontaneous hemorrhage, which is a rare presentation for this tumor.

2. Case Report

A 32-year-old woman visited our emergency department because of acute onset of right loin pain. She had no history of trauma or other medical diseases. A bulging mass was palpable in the right upper area of the abdomen. Laboratory data showed an elevated white blood cell count of 11,890/mm3 and a hemoglobin concentration of 8.9 g/dL. Blood biochemistry tests were within normal ranges. Urinalysis was normal except for micro-hematuria.

On admission, computed tomography of the abdomen and pelvis revealed a large mass with fat components measuring 10 × 9 × 4 cm in the right-sided suprarenal area (Figure 1). The mass was surrounded by hematoma. The mass extended posteriorly, with anteroinferior displacement of the ipsilateral kidney. The differential diagnoses included hemorrhagic adrenal myelolipoma, retroperitoneal liposarcoma, extramedullary hematopoiesis, and adrenal cortical carcinoma. Serum adrenocorticotropic hormone, plasma renin activity, aldosterone, and 24-hour urine vanillylmandelic acid levels were within normal limits, consistent with a biochemically inactive mass.

The patient responded well to medical treatment, demonstrating stabilization of hematocrit and improvement.
Adrenal myelolipoma with hemorrhage

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Adrenal myelolipoma is a rare, benign tumor composed of adipose tissue and bone marrow. It is most commonly diagnosed in patients in their fifth to seventh decades of life, affecting both sexes at similar rates. The incidence was previously estimated to be between 0.08% and 2% at autopsy, but it has been found to comprise up to 15% of adrenal incidentalomas because of the frequent use of noninvasive techniques. Myelolipomas are usually unilateral and their size may vary from 2 mm to 34 cm.

The pathogenesis remains unclear. The embolization of hematopoietic stem cells and ectopic myeloid hyperplasia has been implicated in the etiology. Some authors consider this tumor to be a variant of multiple endocrine neoplasia because of its association with clinical conditions such as Cushing’s syndrome, Conn’s syndrome, Addison’s disease, and obesity. A range of factors such as tissue necrosis, adenocorticotropic hormone and hypersecretion of androgens may contribute to the development of myelolipoma.

Our case is unusually rare due to the patient’s young age (32 years old) and behavior of the tumor. Furthermore, the presentation was also different from those reported in the literature. Myelolipomas are usually found incidentally and may take many years to become symptomatic. The most frequent symptoms of large myelolipomas are nonspecific abdominal pain, hematuria, renovascular hypertension, and other symptoms secondary to mechanical compression of adjacent organs. Spontaneous tumor rupture has been reported in only a few cases. Our patient presented with spontaneous hemorrhage that caused acute onset of loin pain and abdominal distension.

Computed tomography scans of myelolipoma usually demonstrate bulk fat mixed with areas of soft tissue attenuation. Macroscopic fat within an adrenal mass is diagnostic of an adrenal myelolipoma. In our case, compression by intratumoral hemorrhage made it difficult to distinguish the mass from other adrenal neoplasms.

3. Discussion

Adrenal myelolipoma is a rare, benign tumor composed of adipose tissue and bone marrow. It is most commonly diagnosed in patients in their fifth to seventh decades of life, affecting both sexes at similar rates. The incidence was previously estimated to be between 0.08% and 2% at autopsy, but it has been found to comprise up to 15% of adrenal incidentalomas because of the frequent use of noninvasive techniques. Myelolipomas are usually unilateral and their size may vary from 2 mm to 34 cm.

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Focal or diffuse areas of fat can also be seen in a liposarcoma, which was the next diagnostic consideration. However, the expansive tumor growth observed in this case is more common in a myelolipoma than in a liposarcoma, which tends to extend between adjacent organs. Because of the image of a displaced kidney by a separated suprarenal mass, the diagnosis of renal angiomyolipoma was excluded in this case. Thus, the adrenal origin of the mass was established. Extramedullary hematopoiesis or adrenal cortical carcinoma may present with a retroperitoneal mass, but a fatty component would be uncommon.

Management of adrenal myelolipoma is controversial. For small, non-functioning tumors, conservative treatment is generally recommended; however, for larger tumors with a higher risk of spontaneous rupture, surgical removal should be considered. For symptomatic tumors, such as in our patient, adrenalectomy is indicated. We suggest that the possibility of adrenal myelolipoma be considered when dealing with spontaneous retroperitoneal hemorrhage.

References