Isolated Renal Angiomyolipoma Presented as Retroperitoneal Hemorrhage: A Case Report

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Renal angiomyolipoma (AML) is a benign tumor composed of vascular, smooth muscle and adipose tissue. It is usually asymptomatic until the tumor becomes large or complications arise. Emergency physicians and other primary care physicians should be familiar with this disease and its manifestations at different stages of progression. Adequate care for asymptomatic patients, and timely diagnosis and treatment for patients with complications are then possible.

Key words: hemorrhage, renal angiomyolipoma

Introduction

Renal angiomyolipoma (AML) is a benign tumor composed of vascular, smooth muscle and adipose tissue. It is thought to be derived from perivascular epithelioid cells and is a prevalent complication in patients with tuberous sclerosis\(^1\). There are two types of renal AML: isolated AML, which occurs sporadically, is often solitary and is usually asymptomatic until the tumor becomes large or complications arise\(^{2-3}\), and AML associated with tuberous sclerosis that is often bilateral and multiple\(^4\). Here we report on a 47-year-old woman who visited the Emergency Department (ED) because of sudden onset of left flank pain. Ruptured AML of the left kidney was found with retroperitoneal hemorrhage.

Case Report

A 47-year-old woman visited our ED because of sudden onset of left flank pain without preceding trauma. On arrival, she was conscious and vital signs were: respiration 20 breaths/min, heart rate 66 beats/min, blood pressure 113/65 mmHg and temperature 35.5°C. Significant tenderness of the left flank was apparent, but other physical examinations were unremarkable. X-ray examination showed a blurring of the left kidney outline and obscured ipsilateral psoas muscle (Figure 1). Her blood pressure dropped soon after X-ray examination. Fluid resuscitation was given, followed by emergent computed tomography (CT) scanning of the suspected left retroperitoneal lesion. Left kidney hemorrhage was revealed with extravasation of contrast media (Figure 2). Left nephrectomy was performed during emergent surgery because of active bleeding from the ruptured tumor. Histopathological studies of the excised kidney confirmed it was a ruptured AML (Figure 3). The patient had an uneventful recovery after surgery and was discharged 7 days later.

Discussion

Renal AML, whether it is isolated or...
associated with tuberous sclerosis, is usually an incidental finding during image studies of the abdomen for other purposes. It may be symptomatic when its size is large enough or complications occur. Symptomatic patients may manifest with flank discomfort, a palpable mass, fever, weight loss, hypertension or hematuria. Sudden onset of flank or abdominal pain is due to intratumoral or retroperitoneal hemorrhage. It can occur spontaneously, as a result of trauma or during pregnancy. Devastating results may be caused by hypovolemic shock. It is also potentially fatal if adequate treatment is not provided in a timely fashion.

Whether the patient presents with related signs and symptoms or not, a solitary mass in the kidney can be disclosed by various image study methods. Fat density within a non-calcified renal mass is the most important AML diagnostic finding. If there is calcification detected within a renal mass, a renal cell carcinoma should be first excluded. Although ultrasonography and CT scanning can differentiate between an AML, other benign renal tumors and malignancies, preoperative differentiation can sometimes be difficult.

When a patient presents with flank/back pain and hypovolemia, a systemic approach for intraperitoneal or retroperitoneal emergencies should be undertaken, together with resuscitation and symptomatic relief. Abdominal ultrasonography or CT scanning can usually confirm the diagnosis of AML and its complications. Emergent angiography should be undertaken not only for bleeding localization but also transcatheter arterial embolization, if necessary. With the increasing availability of multidetector CT angiography, this can provide much more useful information for planning interventional therapy to preserve most renal function.

Although renal AML is a benign tumor, it can be complicated by aneurysm rupture. A tumor size of 4 cm or larger has been used as a predictor of impending rupture. Although partial nephrectomy has been utilized for treatment,
Fig. 2 Contrast enhanced CT scanning revealed deformed contour of left kidney. Hematoma in the left renal fossa was impressed. Extravasation of contrast media into surrounding retroperitoneal space can also be found. Ruptured kidney with retroperitoneal hemorrhage was diagnosed.
Fig. 3 Gross and histopathological studies of the excised kidney

A. Gross section of the left kidney. A yellow, firm tumor, measured 9 cm × 6 cm × 5 cm, can be found. There is a ruptured cavity, measured 3.5 cm × 2.0 cm × 2.0 cm, in the middle portion of the kidney with hemorrhage

B. Histopathological examination on sections of the renal tumor showed a picture of a variable mixture of mature fat, thick-wall, poorly organized blood vessels, and smooth muscle. Mitosis was rare and cellular atypism was mild
simple enucleation has also been successful in some elective patients, with optimum results\(^{(19-20)}\). It provided an excellent long-term local control and nephron sparing, and no patient had urinary leakage or fistula complications\(^{(20)}\). During hemorrhagic complications, selective arterial embolization should be used as the treatment of choice\(^{(21-22)}\). Partial nephrectomy is reserved for patients who fail to respond to or are unable to receive embolization therapy.

Although recurrence is rare in sporadic AML, it is high in patients with tuberous sclerosis\(^{(23)}\). Lifelong surveillance is therefore necessary. Sporadic renal AML is usually found incidentally or when complications arise. To diagnose and treat AML in a timely fashion, emergency physicians and other primary care physicians should be familiar with this disease. At the same time, attention should be paid to patients suffering from tuberous sclerosis because renal AML is much more common in such patients and is often multiple.

## References

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以後腹腔出血為表現的單發性腎肌脂肪血管瘤

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腎肌脂肪血管瘤是由血管、平滑肌、與脂肪組織所構成的良性腫瘤。這病早期通常沒有症狀，直到腫瘤大到一定程度或是發生併發症才會引起注意。急診醫師必須對此不常見疾病有所了解，特別是疾病各個階段不同的表現。也唯有如此，才足以對此類患者提供適當的診斷、建議與治療。

關鍵詞：出血，腎肌脂肪血管瘤