Oncology

Spontaneous rupture of large bilateral renal angiomyolipomas in a patient with tuberous sclerosis complex: A case report and literature review

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ABSTRACT

Enormous bilateral renal angiomyolipoma (AML) are extremely rare, their spontaneous bleeding also called Wunderlich’s syndrome (WS) is also an unusual situation. It is considered as a life threatening condition requiring fast and effective care.

We present a case of hypovolemic shock due to spontaneous rupture of bilateral giant angiomyolipomas in a 35-year-old female patient with tuberous sclerosis complex (TSC). The hemodynamic instability of the patient leads to an immediate surgery and unilateral nephrectomy was done for the biggest angiomyolipomas. The review of the literature revealed only few cases of spontaneous rupture of renal angiomyolipomas of comparable size.

Introduction

Wunderlich’s syndrome is a rare disease, defined as spontaneous renal bleeding of non-traumatic origin, due to a wide variety of renal tumors of which angiomyolipomas (AML) are the most common.

It is considered as an emergency that may cause life threatening. Huge bilateral renal angiomyolipoma are extremely rare, only few cases were reported in published papers.

We report a case of hypovolemic shock due to a spontaneous rupture of bilateral giant renal AMLs or Wunderlich’s syndrome in a patient with known tuberous sclerosis complex.

Case presentation

A 35-year-old woman with history of tuberous sclerosis was referred to the emergency room following an acute onset of bilateral flank pain lasting since 5 hours accompanied by gross hematuria.

She had a recent history of abdominal distension and recurrent flank pain which never been explored.

The clinical examination revealed abdominal tenderness along the whole abdomen especially in the right flank and also a palpated mass in the right flank.

She was in a state of hypovolemic shock, pale and sweating with a blood pressure of 90/80 mmHg and tachycardia of 110/min.

She had major symptoms of tuberous sclerosis including brown “coffee-with-milk” colored macules with angiofibromatosis on her abdomen.

Initial investigations showed low haemoglobin at 6 g/dl with a hematocrit of 35% and serum creatinine was normal.

Abdominal computed tomography (CT) was performed and showed enormous and bilateral heterogeneous masses. Those masses had fatty content that replace and displace renal parenchyma and extended into the pelvis.

They had a volume of $28 \times 14 \times 15$ cm in the right and $20 \times 10 \times 9$ cm in the left in accordance with bilateral massive renal angiomyolipomas (empty arrow, Fig. 1A and B).

CT scanning demonstrated also perirenal and retroperitoneal hematomas predominantly in the right side with bleeding micro and macroaneurysms inside these huge masses (black arrow Fig. 2).

The patient’s condition deteriorated, her blood pressure dropped to 80/60 mmHg and the heart rate was 135/min.

Angiographic embolization was not available and direct transfer to the operating theatre was necessary.

The major bleeding seems to be secondary to the rupture of the
The largest AML developed in the right kidney, and this kidney was chosen to be removed.

The patient underwent an emergency right lumbotomy under general anesthesia.

At the opening of the retroperitoneal space, about 4 L of blood and clots were evacuated, the right renal artery and vein were controlled with difficulty and a nephrectomy comprising the fatty tumour was realized. Due to hemodynamic instability, the surgical time had to be limited, and after wound closure, the patient was quickly transferred to the intensive care unit.

Histological examination showed an unencapsulated, circumscribed tumor arising from the kidney, composed of dystrophic blood vessels, interspersed with mature adipose tissue and smooth muscle, typical of an angiomyolipoma as cause of the bleeding (Fig. 3).

The hemorrhagic shock was unfortunately sustained and the patient’s need of vasoactive drugs was crescent despite transfusion of 5 red blood cells units with 10 fresh frozen plasma units, her last hemoglobin rate was 5.2 g/dl with hematocrit of 37%.

The follow up showed real deterioration of hemodynamic condition under adrenalin perfusion with installation of disseminated intravascular coagulation syndrome leading to death at day 6 post-operative.

Discussion

Renal angiomyolipoma is a mesenchymal tumor composed of blood vessels, smooth muscle and fat elements in varying proportions, uncommonly, they may become extremely large.

It is a rare entity, with an incidence of 0.1–0.22% in the general population.\(^1\)

AML is seen in two distinct clinical settings: sporadic or in association with tuberous sclerosis complex, an autosomal dominant phacomatoses, classically characterised by epilepsy, mental retardation, and sebaceous adenoma.\(^2\)

The two forms vary to some degree in their imaging features but are histologically indistinguishable.

AMLs associated with tuberose sclerosis tend to be larger, bilateral and multifocal. They appear at an early age compared with sporadic form.

They also present a higher risk of rupture causing a haemorrhagic shock secondary to retroperitoneal bleeding and hematuria.\(^3\)

AML size is directly correlated with the risk of spontaneous rupture which represents the most significant and feared complication in all forms.

Large AML (>4 cm of diameter) develop micro and macro-aneurysms that can lead to spontaneous rupture.

More recent evidence suggests that aneurysm size is more important than tumour size in determining risk of bleeding.

Yamakado et al. demonstrated that the effect of aneurysm size on rupture was greater than that of tumor size.\(^3\)

The proper treatment choice of the AML is very related to the

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**Fig. 1.** (A) and (B): Contrast-enhanced CT scan showing huge heterogeneous masses with fatty content (empty arrow) in bilateral kidneys in accordance with giant AML.

**Fig. 2.** Contrast-enhanced CT scan showing large false aneurysm with microaneurysm formations (black arrow) within right AML.
patient’s condition, tumor size, single or multiple lesions and the presence of acute haemorrhage.

The current management options include observation, embolization, partial and total nephrectomy. Oesterling et al. recommended that symptomatic tumors less than 4cm should be observed regularly with CT or ultrasound, whereas AML greater than 4cm should undergo arterial embolization or surgery.

In general, symptomatic masses or masses greater than 8 cm would require special intervention. Selective arterial embolization of the lesion should be considered especially in patients with tuberous sclerosis who may have limited nephritic reserve due to replacement of the renal parenchyma by multiple cysts and AMLs. But, patients who are haemodynamically unstable and refractory to resuscitation may require emergency surgery.

Radical nephrectomy should be managed if angiographic embolization is not available. Unfortunately, nephrectomy carries a high incidence of morbidity due to the loss of renal function and complications associated with hemodialysis.

In our case, the huge ruptured AML with unstable hemodynamic markers and the unavailability of angiographic embolization, the surgery was imperative and urgent which was done by a right nephrectomy.

Conclusion

Renal angiomyolipoma is a rare benign tumor, it may be associated or not to tuberose sclerosis disease. Their spontaneous rupture is considered as the most feared complication related not only to the tumor size but also to the presence and size of aneurysms.

Our patient had one of the largest bilateral angiomyolipoma ever reported in literature; with a particularity of spontaneous rupture interesting in the same time both sides.

Declaration of competing interest
COI

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