CASE REPORT

Misleading Presentation of a Ruptured Renal Angiomyolipoma

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ABSTRACT

We report a case of a 37-year-old pregnant lady at 27 weeks of gestation presented with contraction pain mimicking abruptio placenta. She developed hypovolemic shock six hours following hospital admission. Subsequent emergency caesarian section revealed a viable fetus with retroperitoneal hematoma but no evidence of placenta abruption. Post-operative abdominal CT angiography demonstrated a ruptured and bleeding left renal mass with fatty component consistent with renal angiomyolipoma. In view of the patient's clinical condition who was persistently critical, an emergency left nephrectomy was proceeded which confirmed the diagnosis of a ruptured left renal angiomyolipoma intraoperatively and histologically.

Keywords: Angiomyolipoma, Ruptured, Pregnancy, Bleeding

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INTRODUCTION

Renal angiomyolipoma (AML) is benign neoplasm containing adipose tissue, smooth muscle and vessels. It is uncommon, with prevalence about 0.2% to 0.6% of the population and has strong predilection in women (1). Pregnancy is a known risk factor of growing renal AML, which may result in further complications. Spontaneous retroperitoneal hemorrhage is one of the complications that usually lead to symptomatic presentation, even though it is only seen in 15% of cases (2). We present a case of ruptured and bleeding renal angiomyolipoma during pregnancy with a misleading presentation.

CASE REPORT

A 37-year-old lady, Gravida 5 Para 4+1 at 27 weeks of gestation presented to our hospital complaining of one-day history of contraction pain radiating to the back. No associated leaking liquor, per vaginal bleeding or hematuria. Clinically her uterine contraction was noted to be 1:10-20 seconds with a closed cervical os. The patient was then admitted for preterm labor. Her vital signs on admission were normal with blood pressure of 120/86 mmHg and pulse rate of 80 bpm. However approximately six hours in the ward, she complained of worsening pain particularly over the left upper abdomen with pain score of 9/10. She was noted to be pallor, hypotensive and tachycardic with blood pressure of 85/55mmHg and pulse rate of 143 bpm. She hyperventilated at a rate of 28 per minute. There was associated mild hypothermia with body temperature of 33.5 degree Celsius. Her abdomen was soft on palpation with no elicitable woody-hard tenderness. An urgent bed side transabdominal ultrasound performed by the obstetrician showed a viable fetus with absence of retroplacental clot and free fluid. Her condition deteriorated despite fluid resuscitation therapy. Blood investigations revealed reduced hemoglobin level from 10g/dL to 7g/dL, hematocrit level dropped from 30.9% to 21.4%, whereas the platelet count increased from 287 x10^9/L to 353 x 10^9/L. Arterial blood gas (ABG) showed metabolic acidosis as evidenced by pH of 7.329, partial oxygen (PO_2) level of 122.8mmHg with oxygen saturation of 98.2%, partial carbon dioxide (PCO_2) level of 25.8mmHg, and bicarbonate level of 13.3mmol/L. Urea and creatinine levels were normal. Constellation of findings deduced decompensated hemorrhagic shock associated with decompensated metabolic acidosis. With clinical diagnosis of abruptio placenta, the obstetric team proceeded with emergency caesarean section. The anesthetic and pediatric teams were alerted for post-operative intensive care and to anticipate neonatal resuscitation respectively.

Intraoperatively, a viable baby was delivered, and uterus and placenta were found to be normal. However, there was a huge retroperitoneal hematoma in the left abdominal cavity. The surgical team who was called

to attend to the patient intraoperatively, was unable to find the source of retroperitoneal hematoma. Hence, an urgent post-operative abdominal CT Angiogram (CTA) was performed. The CTA demonstrated a heterogeneous small fat-containing mass at the lower pole of the left kidney measuring 3.1cm (AP) x 4.3cm (W) x 4.4cm (CC) (Figure 1). There was discontinuation of the lesion at the lower margin, suggesting tumor rupture. Huge left perinephric hematoma was seen extending to the left peritoneum and pelvic cavity (Figure 2). Associated saccular pseudoaneurysm with evidence of active bleeding into the left perinephric space was noted (Figure 3).

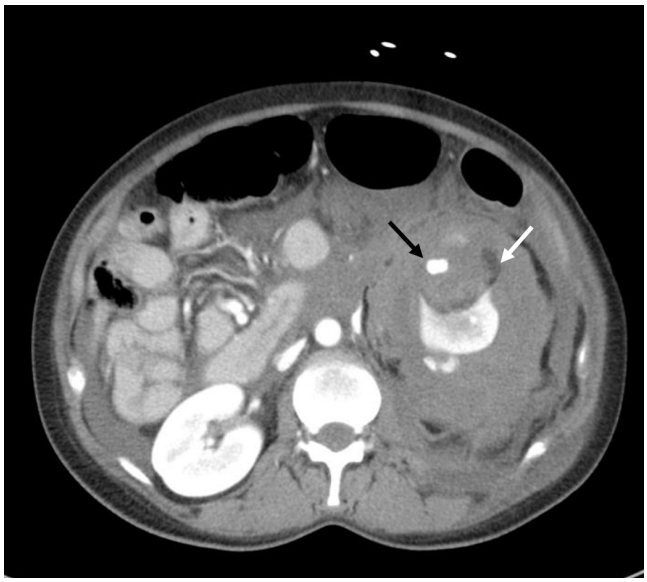

Figure 1: Axial CTA Abdomen in the portovenous phase showing a hypodense mass with fat attenuation component (white arrow) in the lower pole of the left kidney in keeping with an angiomyolipoma. Associated surrounding hematoma. Note the blot of contrast within the mass (black arrow) consistent with pseudoaneurysm

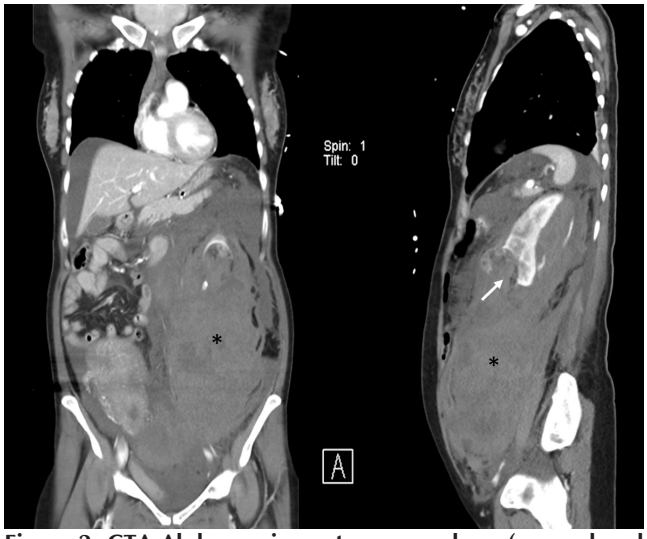

Figure 2: CTA Abdomen in portovenous phase (coronal and sagittal images) showing poor demarcation of the lower part of the lesion suggesting ruptured tumor (white arrow). Note the large left perinephric hematoma (*) displacing the bowels and mesentery to the right. The hematoma extends into the peritoneal cavity, particularly within the pelvic cavity

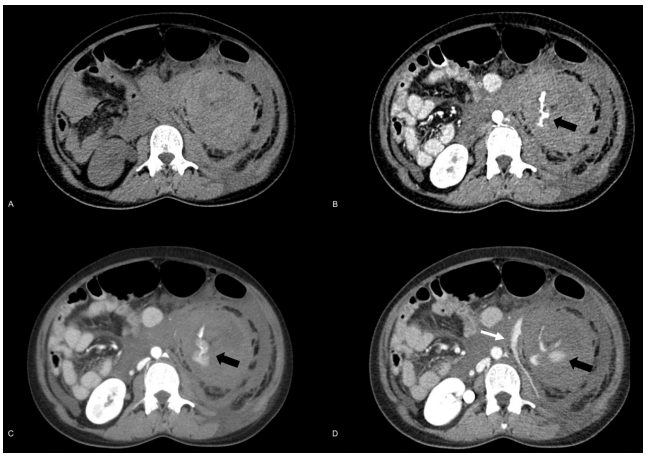

Figure 3: CTA Abdomen in plain (A), arterial (B), portovenous (C) and delayed (D) phases demonstrating progressive contrast pooling and 'blushing' within the left perinephric hematoma (black arrow) indicating active bleeding from the pseudoaneurysm and the ruptured tumour. Contrast tracking is seen in the pararenal space (white arrow)

On further scrutinization of her clinical history, no documentation was found regarding renal nor other abdominal mass previously or recently. The patient remained critically ill with poor hemodynamic status despite intensive care management. Her blood pressure failed to normalize, and she was persistently tachycardic. In view of her persistent critical status and the CT findings of tumor rupture, the urology team opted for emergency re-laparotomy for left nephrectomy rather than angio-embolization. Intraoperative findings confirmed a ruptured 3 cm tumor at the lower pole of the left kidney (Figure 4) associated with large retroperitoneal hematoma causing contralateral shift of the adjacent structures and extension into the pelvic cavity.

Histopathologically the tumor originated from the renal cortical tissue with an area of capsular perforation and mixed with blood clots. The tumor composed of

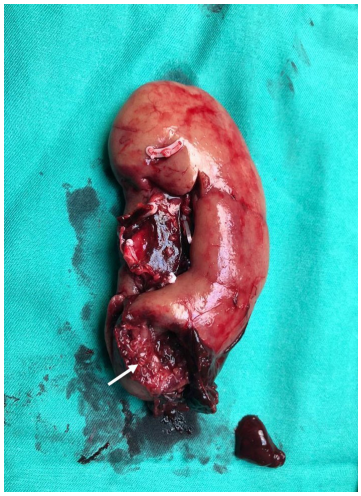

Figure 4: Gross specimen showing the ruptured tumor component with blood clot at the posterior surface of the lower pole of the left kidney measuring 30x15mm (arrow)

spindle cells, epitheliod cells, mature adipose tissue and variable thick-walled vessels, consistent with renal angiomyolipoma. No lymphovascular invasion was observed. The patient recovered uneventfully post-nephrectomy and was discharged well. Her baby was admitted to neonatal intensive care unit for prematurity.

DISCUSSION

Angiomyolipoma is a benign neoplasm consisting of a variable proportion of adipose tissue, smooth muscle and vessels. It is an uncommon benign renal tumor, with prevalence of 0.2 to 0.6% of population, and has a female predilection (1). About 80% of the cases occurs sporadically, while the other 20% have association with tuberous sclerosis complex (TLC) or less commonly, pulmonary lymphangioleiomyomatosis (LAM). Sporadic tumor usually unilateral, solitary and slow growing, while genetic-related tumor commonly bilateral, multiple and grows faster. In our case it was a sporadic angiomyolipoma as the tumor was unilateral and the patient has no syndromic characteristics.

Pregnancy is a known risk to cause enlargement of the tumor, most likely due to increased maternal circulation and tumor response to hormonal changes. These may cause pseudo-aneurysm formation and subsequently extensive retroperitoneal hemorrhages. Rupture of the tumor is another complication which usually associated with the size of the lesion, genetic abnormality, aneurysm formation and pregnancy (3). In our case CTA demonstrated both bleeding pseudoaneurysm and tumor rupture. Bleeding and ruptured renal AML in pregnancy is rare, only reported less than 50 cases in the past 42 years (4,5). The incident requires emergency surgical treatment by the multidisciplinary team as in our case (5).

Most of the patients have no symptom and found incidentally during imaging for other reasons. However, some of the patients may present with hematuria, flank pain or palpable mass. Symptomatic presentation occurs in less than 15% of the cases, and mostly related to spontaneous retroperitoneal hemorrhage or rupture. As in our patient, she was never diagnosed to have renal tumor before. Thus, her presentation with back pain and premature labor with a decompensated hemorrhagic shock and decompensated metabolic acidosis, increase the clinical suspicion of abruptio placenta. The undiagnosed renal AML in pregnancy misled the managing teams during the acute event. Even though the transabdominal ultrasound revealed no evidence of retroplacental clot, less attention was given to the other solid organs and intraabdominal region, making the renal mass and retroperitoneal clot being missed. This could be depicted if the ultrasound was performed by the radiology team. But again, the life-threatening condition requires acute measures for the most likely diagnosis deduced at that particular time.

Diagnosis and management of renal AML largely depends on the radiological assessment. Generally, sporadic renal AML can be radiologically classified as fat-rich; also known as classic AML and fat-poor AML. Presence of adipose tissue in a lesion or mass is the diagnostic criteria of a classic AML, thus making it possible to be diagnosed on CT or MRI. Fat-poor AML is a diagnostic challenge because the paucity of the adipose tissue, making it difficult to differentiate with renal cell carcinoma (RCC). However, the incidence is uncommon (2). In our patient, a thorough observation revealed a small portion of fat attenuation within the mass, in the background of the huge perinephric hematoma, making the diagnosis of renal AML the first in the list as compared to other tumors.

Tumor size of more than 4cm and presence of intralesional aneurysm are known to be the factor predictive of bleeding and ruptured renal AML, a complication prompt early intervention (2). In our patient, presence of pseudoaneurysm within the tumor with evidence of active bleeding, indicates the complication already occurred at the time of diagnosis.

Several factors affect management of renal AML in pregnancy, including gestational age, hemodynamic status of the mother, fetal well-being, and the facilities available in that particular center such as operation theatre, angiography unit and neonatal intensive care unit (4). Patients who is hemodynamically stable with no evidence of active bleeding or fetal compromise can be managed conservatively. However, the patient should be closely monitored at the center with adequate facilities to anticipate any acute complication that requires immediate definitive management. If the patient is in shock or shows evidence of active bleeding, immediate treatment is required by angio-embolization without or with nephrectomy. Both treatments have shown favorable results, although it is also determined by tumor suitability and availability of the resources (4). As in our patient, although angio-embolization is an option for nephron-sparing procedure, the primary team decided for emergency nephrectomy since the patient was in decompensated hypovolemic shock which required fast action to prevent morbidity and mortality.

CONCLUSION

Renal angiomyolipoma is a benign neoplasm which usually found incidentally on imaging for other causes. It has female predilection and known to grow rapidly during pregnancy causing complications such as hemorrhages from intratumoral pseudoaneurysm and tumor rupture. It is challenging for the managing team when the patient with previously undiagnosed renal angiomyolipoma presented with complication during pregnancy. As a take home message, non-obstetric clinical diagnosis should always be considered when the decompensated hypovolemic shock status does not

correlate with the obstetric clinical evaluation, hence requires appropriate radiological assessment.

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