CASE REPORT

Ascending cholangitis: rare presentation of a ruptured right-sided renal angiomyolipoma

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Abstract

We report the case of a 46-year-old female who presented to the Emergency Department with acute, painful obstructive jaundice, with evidence of secondary ascending cholangitis. Surprisingly, imaging revealed the clinical picture to be caused not by hepatobiliary pathology, but by external compression of the biliary tree from a ruptured renal angiomyolipoma (AML) of the right kidney. The patient remained haemodynamically stable and conservative management saw resolution of biliary obstruction. We believe this to be the first report of a renal AML presenting in this way. This report highlights the diverse spectrum of presentations of renal angiomyolipomas.

INTRODUCTION

Renal angiomyolipomas are typically found incidentally or present with pain, haematuria or a palpable mass. However, unusual presentations have been reported, especially in the context of tumour rupture. We present what we believe to be the first case in the literature of a ruptured renal angiomyolipoma (AML) presenting with biliary obstruction. This report outlines the potential of renal AMLs to present atypically, and demonstrates an apparently straightforward clinical presentation with an unusual aetiology.

CASE

A 46-year-old female with a distant history of cholecystectomy presented to A&E reporting right upper quadrant (RUQ) pain and jaundice of 5 days duration. She reported the onset of fevers and vomiting the previous day. There were no bowel or urinary symptoms.

On examination, she was apyrexial with scleral icterus. Her abdomen was soft but tender in the RUQ. Investigations revealed Hb of 10 g/dL, leukocytosis (14.6 × 10^9/L) and elevated C-reactive protein (299 mg/L). Liver function tests were elevated in an obstructive pattern, with bilirubin 46 umol/L, alkaline phosphatase 274 IU/L and gamma glutamyl transferase 142 IU/L. Renal function was normal.

Fluid resuscitation and IV antibiotics were commenced, and an abdominal ultrasound performed. It revealed the presence of a large heterogenous right renal mass. Further evaluation was performed with magnetic resonance cholangiopancreatography. Findings were of a 5.7 × 4.6 cm^2 exophytic mass of high T1 signal intensity arising from the posterior right kidney. Adjacent to the mass was a large 8.3 × 10 cm^2 retroperitoneal haematoma (Fig. 1). These findings were consistent with a ruptured angiomyolipoma. The adjacent common bile duct was dilated at 8 mm, secondary to mass effect (Fig. 2).

The case was discussed at urology MDT for possible selective renal embolization, however, in the context of improving...
More recent studies highlight a shift towards incidental presentations, likely a consequence of increased availability and sophistication of imaging performed for other indications, or as a screening modality in tuberous sclerosis. A retrospective UK study published in 2011 noted 86% of patients \( n = 102 \) to be asymptomatic at diagnosis of AML. Those with symptomatic presentations had pain (5.9%), haematuria (2.9%) or spontaneous life-threatening haemorrhage (5.9%). Patients with tuberous sclerosis were noted to be more frequently asymptomatic at diagnosis; possibly reflecting a referral bias [3].

A 24-year US study concluded in 2009 with similar results; 83.9% of patients \( n = 87 \) presented incidentally, while 16% were symptomatic with pain, haematuria or haemorrhage [4].

Less commonly, renal angiomyolipomas present as the eponymous Wunderlich’s syndrome, first described in 1856, with spontaneous rupture resulting in haemorrhage confined to perirenal/subcapsular spaces [5]. The literature also reveals a handful of apparently unique cases, including presentation as thrombus involving the right atrium [6], caudal rupture resulting in haemoperitoneum with abdominal compartment syndrome [7] and duodenal obstruction [8].

Our patient was managed with close clinical observation as an inpatient during the acute phase, and subsequently discharged on active surveillance. Management strategies for renal AMLs have been somewhat controversial, with debate regarding whether a particular tumour size threshold (usually 4 cm) serves as an absolute indication for intervention in the absence of symptoms or concern for malignancy. Recent studies suggest safety of active surveillance for asymptomatic renal AMLs > 4 cm [9] and those in the 4–6 cm bracket [10]. Whilst higher rates of requirement for delayed intervention are acknowledged with larger tumours, this approach potentially avoids overtreatment of a significant proportion of patients [9].

Renal angiomyolipomas are interesting tumours with a diverse array of potential presentations. Our case of an AML presenting as biliary obstruction further reiterates this potential to present atypically.

**CONFLICT OF INTEREST STATEMENT**

None declared.

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**PATIENT CONSENT**

Yes.

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