Spontaneous rupture of a retroperitoneal extrarenal angiomyolipoma initially misdiagnosed as acute cholecystitis

Ghassan Almaimani, Thomas Zoedler, André Schneider, Bayan Almaimani

ABSTRACT

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Case Report: We report the case of an 81-year-old woman with a right extrarenal retroperitoneal mass initially misdiagnosed as acute cholecystitis. A definitive diagnosis of angiomyolipoma is typically made after histopathologic examination of the operative specimen, which was not possible in this case. However, modern imaging modalities, particularly CT scan and MRI scan, have made it possible to identify these lesions in vivo.

Conclusion: This case highlights that the preoperative diagnosis of extrarenal angiomyolipoma can be challenging. Awareness of their clinicopathological and radiological features is essential.
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Keywords: Angiomyolipoma, Computed tomography scan, Hemorrhage, Hematoma, Retroperitoneum

INTRODUCTION

Extrarenal angiomyolipomas are extremely rare benign tumors that are sometimes difficult to diagnose preoperatively. Angiomyolipomas are most often detected incidentally during imaging for other reasons, although they occasionally present with abdominal pain when large in size or when complicated by hemorrhage [1–3]. Imaging is very useful for diagnosis and determining their characteristic composition, although histopathological assessment is ultimately necessary when possible to differentiate them from malignant fatty tumors such as liposarcomas. The exact incidence of extrarenal angiomyolipomas is unknown, but they are described in a small number of case reports. Herein, we present an educational case of extrarenal retroperitoneal angiomyolipoma with spontaneous rupture that was initially confused with acute cholecystitis.

CASE REPORT

An 81-year-old white woman with known gallstone disease presented to the emergency department with a
two-day history of right upper quadrant pain. There was no history of trauma and she was not on anticoagulant therapy. Her past medical history included myocardial infarction and stroke, but her past surgical history was unremarkable. On presentation, her vital signs were within normal limits and there was no fever. Her abdomen was tender with guarding in the right upper quadrant on examination. Murphy’s sign was positive.

Blood tests indicated that she was anemic with a hemoglobin level of 9.3 g/dL, but her coagulation profile and platelet count were normal. C-reactive protein (CRP) was elevated (Table 1). Abdominal ultrasonography revealed a slightly thickened gallbladder containing small gallstones (Figure 1). The patient was admitted to hospital with a diagnosis of acute cholecystitis and initially managed with parental ampicillin/sulbactam 1.5 g 8 hourly. Despite antibiotic therapy, the clinical symptoms did not improve on day-2 and, moreover, the CRP was further elevated.

A decision was made to perform laparoscopic cholecystectomy.

On diagnostic laparoscopy, the gallbladder was found to be unremarkable, but further exploration revealed a large retroperitoneal hematoma without evidence of active bleeding. The hematoma was managed by saline lavage and drain placement. It was decided to terminate the operation without removal of the lesion due to the patient’s age and comorbidities. However, postoperative abdominal computed tomography (CT) scan with intravenous contrast (Figure 2) demonstrated a right retroperitoneal fat-containing mass situated behind the liver. A diagnosis of extrarenal angiomyolipoma with spontaneous rupture was made based on: (i) direct visualization of the lesion; (ii) our prior experience with such lesions; (iii) the known propensity of angiomyolipomas to hemorrhage; and (iv) a lack of suspicious features on imaging. The patient improved over the postoperative period and she was discharged from hospital on the seventh postoperative day.

The patient was managed conservatively due to the patient’s age and comorbidities, the absence of active bleeding, and because there was no pressure effect on surrounding anatomical structures. She received regular follow-up every two months for half a year then yearly thereafter for two years. She remains symptom free and the lesion was sonographically stable at her last follow-up appointment, further supporting the benign diagnosis.

### Table 1: Biochemistry and hematology results

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Day of presentation</th>
<th>Day 2 after presentation</th>
<th>Units</th>
<th>Normal range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin</td>
<td>9.3</td>
<td>8.9</td>
<td>g/dl</td>
<td>12–16</td>
</tr>
<tr>
<td>White cell count</td>
<td>9.19</td>
<td>7.74</td>
<td>1000/ul</td>
<td>4–10</td>
</tr>
<tr>
<td>C-reactive protein</td>
<td>3.95</td>
<td>5.13</td>
<td>mg/dl</td>
<td>0–1</td>
</tr>
<tr>
<td>Bilirubin</td>
<td>0.68</td>
<td>0.57</td>
<td>mg/dl</td>
<td>0.10–1.20</td>
</tr>
<tr>
<td>Gamma-glutamyl transferase</td>
<td>63</td>
<td>61</td>
<td>U/l</td>
<td>35–104</td>
</tr>
</tbody>
</table>

The patient was managed conservatively due to the patient's age and comorbidities, the absence of active bleeding, and because there was no pressure effect on surrounding anatomical structures. She received regular follow-up every two months for half a year then yearly thereafter for two years. She remains symptom free and the lesion was sonographically stable at her last follow-up appointment, further supporting the benign diagnosis.

Figure 1: Ultrasound image of the gallbladder acquired at presentation in the emergency department. The gallbladder contains gallstones and has a slightly thickened wall.

Figure 2: Sagittal abdominal computed tomography view illustrating a fat-containing retroperitoneal mass (10x11x11 cm) behind the liver.
DISCUSSION

Angiomyolipomas are uncommon benign neoplasms with a characteristic composition of blood vessels, smooth muscle, and mature fat. The majority of angiomyolipomas are sporadic (80%) and most commonly found in adult women (mean age of presentation 43 years; F:M 4:1). The remaining 20% are seen in the hereditary setting of tuberous sclerosis, Von Hippel-Lindau (VHL) syndrome, and neurofibromatosis type 1 [1]. Angiomyolipomas are typically found in the kidney but are also commonly found in the liver and, less commonly, retroperitoneum, ovary, fallopian tube, spermatic cord, palate, and colon [2]. Although most angiomyolipomas are asymptomatic, 68–80% of patients develop symptoms when the tumor grows to 4 cm or more [3]. The most severe symptoms are associated with tumor rupture, with patients presenting with acute onset pain due to hemorrhage; up to 20% are in shock at the time of presentation. Although hemorrhage is a frequent complication, necrosis and calcification are rare [4].

A definitive diagnosis of angiomyolipoma is typically made after histopathologic examination of the operative specimen. However, modern imaging modalities, particularly CT scan and MRI scan, have made it possible to identify these lesions in vivo [4], with MRI most suitable for assessing the distinctive vascular and adipose components of these lesions [2]. In our case, the most important differential diagnosis was liposarcoma, which is characterized on CT scan by the presence of non–fat attenuating intratumoral nodules and calcifications, especially multiple, globular calcifications, which were not seen in our case [5–7]. Furthermore, angiomyolipomas are known to have a propensity for hemorrhage, unlike liposarcomas, which are hypovascular; we note that a single case of retroperitoneal liposarcoma complicated by hemorrhage is reported in the literature [8]. Angiomyolipomas are primarily treated by surgery or tumor embolization based on tumor size and symptoms. Many cases of renal angiomyolipoma with spontaneous rupture are reported in literature but, to date, only three cases of retroperitoneal extrarenal angiomyolipoma with spontaneous rupture (including our case) are described [9, 10].

CONCLUSION

This case highlights the need to be vigilant and aware of this infrequent entity, whose presentation can mimic other common conditions. A high index of suspicion is required for effective treatment of this lesion.

Acknowledgements
We are thankful the patient for allowing us to report her medical report as a case report.

Author Contributions
Ghassan Almaimani – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Thomas Zoedler – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
André Schneider – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Bayan Almaimani – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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