CASE REPORT

Acute haemorrhage from a retroperitoneal liposarcoma: a rare presentation

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SUMMARY

A previously healthy 61-year-old Caucasian woman presented to the emergency department after collapsing at home with associated abdominal pain radiating to her back. An urgent CT angiogram was requested to rule out a ruptured aortic aneurysm. This showed a large 21 cm fat-containing lesion arising from the mid-pole of the left kidney, with an adjacent 4 cm perirenal haematoma. An initial diagnosis of a ruptured angiomyolipoma was made. Her haemoglobin was 105 g/L, with a creatinine of 104 mmol/L and an eGFR of 47 mL/min. Her clotting profile was normal. Following resuscitation, she was taken to the operating room and underwent an emergency open left radical nephrectomy via a left flank incision. Her recovery was uneventful and she was discharged home after 6 days. The histopathology confirmed a well-differentiated liposarcoma.

BACKGROUND

The majority (80%) of retroperitoneal sarcomas present as asymptomatic abdominal masses. Symptoms are usually non-specific, and they do not appear until the tumour becomes very large. A mass effect or local invasion may lead to pain, gastrointestinal obstruction, weight loss, and neurological and muscular skeletal symptoms.¹ Acute haemorrhage from a retroperitoneal liposarcoma is rare. We would like to report a case of a large retroperitoneal liposarcoma that presented with acute haemorrhage at first diagnosis.

Figure 1 CT abdomen: liposarcoma, kidney and perirenal haematoma.

Figure 2 CT abdomen: retroperitoneal liposarcoma and haematoma.

CASE PRESENTATION

A previously healthy 61-year-old woman with no relevant family or medical history presented to the accident and emergency department after experiencing acute abdominal pain and collapsing at home. She was found to be tachycardic (heart rate 120) and had low blood pressure (90/50 mm Hg), and required immediate resuscitation.

INVESTIGATIONS

Baseline blood tests including a cross match were requested, and these showed that she had a haemoglobin of 105 g/L, with a creatinine of 104 mmol/L and an estimated glomerular filtration rate (eGFR) of 47 mL/min. Her clotting profile was normal. An urgent abdominal CT angiogram was performed, which identified a large 21 cm fat-containing lesion arising from the mid-pole of the left kidney, with an adjacent 4 cm perirenal haematoma (figures 1 and 2).

DIFFERENTIAL DIAGNOSIS

At presentation, a ruptured aortic aneurysm was suspected. However, following review of her CT scan, it was thought that she may have had a ruptured renal angiomyolipoma consistent with Wunderlich syndrome causing haemorrhagic shock.

TREATMENT

The patient underwent an emergency operation on the day of admission. A retroperitoneal approach to the kidney via a left flank incision was performed.

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Figure 3 Histopathological specimen.

Intraoperatively, a large mass was encountered, which seemed to be originating from the perinephric fat with a surrounding haematoma. Control of the left renal hilum was achieved with ligation of the renal artery and vein and division of the ureter. The left kidney and the surrounding mass were then retrieved and sent for histology.

OUTCOME AND FOLLOW-UP

The patient made an uneventful recovery and was discharged from hospital 6 days later.

Histopathological analysis demonstrated a lipomatous tumour composed of lobules of well-differentiated adipocytes with scattered intervening spindle to polygonal cells with hyperchromatic nuclei. There was no evidence of dedifferentiation. A diagnosis of well-differentiated liposarcoma was made with the benefit of a second opinion and later confirmed with molecular testing. She was referred to our regional sarcoma centre for continued follow-up.

She has not developed any recurrence in the first 6 months postoperatively.

Figure 4 Histopathological specimen.

Learning points

- ▶ Although rare, liposarcomas may present with acute haemorrhagic shock.
- ▶ Preoperative cross-sectional imaging is crucial in establishing a diagnosis and in aiding in the surgical planning; however, this has limitations, and a low index of suspicion for a differential diagnosis must be kept in mind especially for malignancy.
- ▶ In acute haemorrhage with haemodynamic instability, surgery is the mainstay of treatment.
- ▶ Complete resection of a liposarcoma should be the primary aim of treatment, which may involve resection of adjacent or involved organs.
- ▶ Once a histological diagnosis has been established, these patients should be referred to the regional sarcoma centre for follow-up and/or adjuvant treatment.

DISCUSSION

Haemorrhagic shock caused by an acute bleeding from a retroperitoneal liposarcoma at first presentation is extremely rare, and there are very few published cases in the literature. Guzmán Martínez-Valls *et al*² published a case report in 1996 outlining a similar case in which the patient underwent emergency surgery with excision of the ipsilateral kidney. The histology demonstrated a well-differentiated tumour, making acute haemorrhage even more unexpected.

A differential diagnosis should always be considered, and includes a primary neoplasm arising from a retroperitoneal visceral structure (eg, pancreas, adrenal glands, kidneys and duodenum), a retroperitoneal sarcoma, a lymphoma or a metastatic lesion.³ The optimal treatment for patients with localised, resectable retroperitoneal sarcomas is surgery with negative margins.⁴ Although negative microscopic margins (R0) are ideal, this is difficult to achieve technically from a surgery standpoint and to truly evaluate from a pathology standpoint for these often large tumours. The accepted goal of surgery is for a complete macroscopic resection (R0/R1).

Given the difficulty of achieving complete margin-negative resections, en-bloc resection of adjacent viscera is frequently required.⁵ The kidney is the most frequently resected organ (with rates varying from 36% to 80%), followed by segmental resection of the large bowel, pancreas and spleen (in left-sided tumours).^{6,7}

Other options to treat bleeding, such as angioembolisation, should be explored if available. In acute malignant haemorrhage with haemodynamic instability, surgery is almost certainly always required.

Contributors MAS was one of the consultants looking after the patient. He was integral in editing the case report and planning of the report. NS was the SHO in the treating team. He wrote the abstract and a large proportion of the case. OO-N was the registrar of the treating team. He gave important input into the report, specifically in getting radiological and pathological images. AQ is the overseeing consultant, operated on the patient and is responsible for the bulk of the information in the report.

Competing interests None declared.

Patient consent Obtained.

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