CASE REPORT

Delayed diagnosis of angiosarcoma of the spleen: clinically presenting as recurrent haemoperitoneum following embolisation

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SUMMARY

Angiosarcoma is a highly aggressive and extremely rare neoplasm of the spleen, with a limited number of reported cases worldwide.

We report a case of a 69-year-old man who presented with progressive symptoms of fatigue, abdominal and back pain, anaemia and haemoperitoneum 1 year following embolisation of the spleen for splenic rupture. Additional imaging and colonoscopy with biopsy revealed intra-abdominal fluid and a colonic adenocarcinoma. Diagnostic laparoscopy demonstrated a haemoperitoneum and colon carcinoma, after which splenectomy via laparotomy was performed. Pathology report revealed an angiosarcoma of the spleen with peritoneal tumour deposits.

Rupturing of a spleen may be due to an underlying disease. Therefore, the differential diagnosis of splenic rupture should include the possibility of a splenic tumour. The definitive diagnosis and treatment of a splenic angiosarcoma may require splenectomy, although prognosis is poor.

BACKGROUND

Angiosarcoma of the spleen is a rare mesenchymal malignancy and a highly aggressive tumour.^{1 2} So far, no more than 300 cases have been reported in the English literature worldwide since Langhans first described this disease in 1879.³ The prognosis is poor, regardless of the treatment strategies.^{2 4} We report a case of a 69-year-old man with a delayed diagnosis of splenic angiosarcoma (as second

diagnosis next to colon carcinoma) 1 year following coiling of the spleen due to splenic rupture.

CASE PRESENTATION

A 69-year-old man presented at our emergency department with progressive symptoms of fatigue, reduced fitness, shortness of breath on exercise, anorexia, and abdominal and back pain for the past few months. The patient had lost a few kilograms in weight during the last 3 weeks. Anaemia was diagnosed by the general practitioner a few months earlier.

The patient had a history of chronic obstructive pulmonary disease, hypertension and depression. Furthermore, he had undergone embolisation of the splenic artery 17 months earlier, due to a splenic rupture 6 weeks after minimal blunt abdominal trauma (figure 1).

Physical examination revealed stable vital signs and no abnormalities on examination of the head, neck, heart, lungs and extremities. There were no skin abnormalities. The abdomen was distended with bulging flanks, a small haematoma was visible around the umbilicus, bowel sounds were soft during auscultation and there was shifting dullness; there were no signs of abdominal tenderness or peritonitis.

Abnormalities on laboratory examination were a microcytic anaemia with a lowered haemoglobin level of 6.3 mmol/L (range 8.5–11.0), leucocytosis (white cell count $11.7 \times 10^9/L$ (range 4.5–11.0) and

Figure 1 (A) Abdominal CT scan (portal phase) showing contrast extravasations as a sign of active splenic bleeding (17 months earlier). (B) Angiography of the embolised spleen (17 months earlier).

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Figure 2 Abdominal CT scan (portal phase) at current presentation showing splenic artery coil embolisation with areas of focal necrosis and a large amount of intra-abdominal fluid.

thrombocytosis (platelets $550 \times 10^9/L$ (range 150–400)). Renal and liver functions were normal.

Chest X-ray showed right lower lobe atelectasis. Abdominal ultrasound showed a large amount of intra-abdominal fluid. Drainage of the fluid (11 L) revealed bloody ascites (exudate, without malignant cells on pathological examination). Abdominal CT (portal phase) demonstrated intra-abdominal fluid without evidence of its origin, a postembolisation spleen with focal necrosis and a renal cyst (Bosniak classification 4; [figure 2](#)). There were no signs of metastases. The carcinoembryonic antigen (CEA) in the blood was measured at $1.0 \mu g/L$. During the following days a colonoscopy was performed, which demonstrated a tumour at the hepatic flexure ([figure 3](#)). Biopsy revealed an adenocarcinoma. This tumour was not visible at the previous abdominal CT scan. Gastroscopy was normal, except for a sliding hiatal hernia.

We performed a diagnostic staging laparoscopy. A large amount of bloody ascites was seen in the upper abdomen with peritoneal abnormalities, which were biopsied and found to be benign on frozen section. The spleen could not be visualised well. Furthermore, there was a marking in the right colon, but

Figure 3 Colonoscopy: tumour at the hepatic flexure.

no evident colonic tumour was seen. There were no signs of liver metastases. The procedure was converted to a laparotomy. The spleen seemed to be fixed to the surrounding tissues and, because of the haemoperitoneum, it was decided to perform a splenectomy. The postoperative pathology report revealed a splenic angiosarcoma with peritoneal tumour deposits ([figures 4 and 5](#)). The postoperative course was complicated by an ongoing production of ascites fluid of 2–3 L/day, which was drained by an intra-abdominal surgical drain. Furthermore, the patient developed pneumonia for which antibiotic treatment was started. The patient was discussed in the multidisciplinary team and there was an indication for palliative systemic therapy. However, during hospital stay the patient clinically deteriorated and died 26 days following surgery.

OUTCOME AND FOLLOW-UP

The patient deteriorated during hospital stay and died before palliative systemic therapy could be given.

DISCUSSION

Primary angiosarcoma of the spleen is a rare malignant disease, but it is considered the most common non-haematopoietic splenic malignancy.¹ Splenic rupture is the most feared complication and may often be the presenting symptom.² Other frequently encountered symptoms, such as abdominal or back pain, fatigue or weakness, anorexia and weight loss are non-specific, and can start a few months prior to clinical presentation. Also, cases with symptoms of chest pain, shortness of breath and gastrointestinal bleeding have been described, as well as asymptomatic patients.^{2 5–7} Splenomegaly is the most common finding on physical examination. Laboratory findings include anaemia, leukocytosis and thrombocytosis.^{2 6 7} Most of these findings were present in our case.

At the time of presentation, our patient had collapsed after acute onset of left flank pain. Six weeks prior to this event, the patient had fallen down in his shed. Abdominal CT demonstrated intra-abdominal haemorrhage, signs of splenic rupture, and consolidated fractures of the eighth and ninth ribs. Although there was an inhomogeneous aspect of the spleen, there were no signs of a splenic mass, metastases or other intra-abdominal abnormalities. These findings in combination with the recent abdominal trauma were sufficient to perform no

Figure 4 Remains of the spleen after fixation and cutting for the histology. Weight: 230 g. Note the absence of a smooth capsule, the areas of necrosis (yellow) and tumour (whitish).

Figure 5 (A) Low-power magnification ($\times 5$) of an H&E-stained slide with tumour (centre) haemorrhage (left) and necrosis (bottom). (B) H&E stain ($\times 20$) of tumour with atypical blood vessels with erythrocytes in and between the vessels. (C) H&E stain ($\times 20$) of more solid growing tumour, consistent with a high-grade angiosarcoma. (D) Slide-stained with antibodies against CD31. Tumour cells were also positive with CD34 and somewhat less with factor VIII and podoplanin. (E) Slide stained with antibodies against human herpesvirus 8, negative, arguing against an HIV-associated tumour.

further diagnostics. The patient was treated successfully with acute embolisation of the spleen.

When reviewing the literature, no other cases were found reporting on the treatment of splenic rupture due to splenic angiosarcoma with arterial embolisation. Furthermore, a minority of patients present with delayed splenic bleeding, ranging from 2 days to over 4 weeks, following trauma.^{8 9} In a recent case, delayed splenic rupture even occurred 70 days following the initial trauma.¹⁰ While it is possible that trauma had caused a splenic rupture in our patient, it is also likely that the spleen ruptured due to the angiosarcoma itself. However, this was not proven at first presentation 17 months prior.

A systematic review by Renzulli *et al*¹¹ reported 845 patients with atraumatic splenic rupture and in 31 patients the aetiology was splenic angiosarcoma. Although the literature regarding splenic angiosarcoma mostly includes case reports, a few larger series (including up to 40 cases) described splenic rupture or haemoperitoneum on presentation in 8–29% of the cases.^{2 4–7 12 13} Splenectomy was the treatment of choice in all these patients.

At second presentation, the CT scan demonstrated an embolised spleen without signs of a splenic mass. There was a large amount of intra-abdominal fluid. The literature on abdominal ultrasound or CT in patients with ruptured spleens due to

angiosarcoma reports heterogeneous findings. Cases with intra-abdominal haemorrhage and poorly defined spleen masses have been described.^{7 14 15}

The diagnosis in our case was further challenged by the colonic carcinoma found at colonoscopy. Combined with the large amount of intra-abdominal fluid on CT scan, the differential diagnosis included a metastasised colonic tumour, with an embolised spleen as secondary diagnosis. However, there were no signs of (peritoneal or omental) metastasis on abdominal CT scan. Furthermore, ascites drainage revealed bloody fluid and pathology reports were negative for malignant cells in ascites, and serum CEA value was normal. Even the frozen section of the biopsied peritoneal mass during surgery showed no malignant cells. A recent case report clearly addressed this cytopathological challenge in the diagnosis of angiosarcoma since a diversity of histological growth patterns may be found and pathology reports may not always be conclusive.¹⁶

Concomitant second malignancies have been reported before. Falk *et al*² described a concomitant malignancy in 3 of 40 patients (basal cell carcinoma, renal cell carcinoma, non-Hodgkin's follicular lymphoma). The literature revealed only two other cases with synchronous colonic cancer and splenic angiosarcoma (table 1).^{17 18}

Table 1 Primary splenic angiosarcoma with concomitant colon carcinoma

Author	Kinoshita <i>et al</i> ¹⁷	Kamocki <i>et al</i> ¹⁸	Kornmann <i>et al</i>
Year	1988	2013	Present case
Male/female	Female	Female	Male
Age (years)	51	77	69
Symptoms	Anal bleeding, altered bowel habits	Elective surgery (haematochezia and fatigue)	Fatigue, anorexia, shortness of breath, abdominal and back pain, weight loss
Laboratory examination	Normal range	Normocytic anaemia	Microcytic anaemia, leukocytosis, thrombocytosis
Imaging	CT scan, coeliac angiography	Colonoscopy	Chest X-ray, abdominal ultrasound/CT scan, colonoscopy
Findings during surgery	Not reported	Enlarged spleen with tumours, purpura-like changes in the greater omentum/wall/mesentery small bowel, tumour above the colorectal junction	Bloody ascites, peritoneal abnormalities, a fixed spleen to the surrounding tissues
Disease	1. Adenocarcinoma (sigmoid) 2. Splenic angiosarcoma 3. Hepatic leiomyosarcoma	1. Adenocarcinoma (sigmoid) 2. Splenic angiosarcoma	1. Adenocarcinoma (hepatic flexure) 2. Splenic angiosarcoma
Pathology spleen	Haemangiosarcoma (2 cm solitary nodule)	Angiosarcoma (G3)	Angiosarcoma with peritoneal metastasis
Metastasis	Not reported	Lymph nodes (origin not noted)	Peritoneal (angiosarcoma)
Treatment	Left hepatic lobectomy, splenectomy, sigmoidectomy during the same operation	Colorectal resection, splenectomy	Embolisation, splenectomy
Prognosis	Free from signs of recurrent disease more than 3 years after surgery	Death 1 month following surgery	Death 26 days following surgery (about 19 months following splenic rupture)

Findings are reported as stated in the original case reports.

Angiosarcoma is a highly aggressive tumour with a poor prognosis. At presentation, metastases are present in the majority of patients. These mostly involve the liver, peritoneal surface, bones, lymph nodes, lungs, gastrointestinal tract, brains, soft tissues and adrenal glands.^{2 6 7} About 20% of patients are alive for a median time of 6 months after diagnosis.²

Although it is known that the outcome of patients with splenic angiosarcoma is poor, our patient presented with new symptoms more than a year following embolisation. In the other case of splenic angiosarcoma and sigmoid cancer, a colorectal resection with resection of the locoregional lymph nodes and a splenectomy were performed during the same procedure.¹⁸ No adjuvant chemotherapy was administered and the patient died approximately 1 month later. In our case, the deferred choice not to immediately perform a splenectomy in the first place was the unknown origin of the ascites and the presence of a colonic carcinoma. Despite the ongoing haemoperitoneum, none of the treating physicians had considered a splenic malignancy as primary cause for the splenic rupture. Therefore, it was decided to first perform a staging laparoscopy. Owing to intra-abdominal findings and patient performance, the decision was made to perform only a splenectomy. We would have performed the colectomy on second laparoscopy. However, after the diagnosis of angiosarcoma our patient deteriorated rapidly and died almost a month following surgery.

Unfortunately, the definitive treatment of angiosarcoma is still open to debate. It is not known whether adjuvant or palliative chemotherapy may be effective¹⁹ and further research may be necessary to answer this question.

In conclusion, embolisation for splenic rupture is effective. However, it should be noted that a ruptured spleen may be due to non-trauma-related causes. The differential diagnosis of splenic rupture should always include splenic tumours (benign or malignant). The definite diagnosis requires splenectomy. With its poor prognosis, splenectomy in angiosarcoma will often prove to be solely diagnostic.

Contributors All authors participated in the design of the study. VK drafted the manuscript, but all authors edited the manuscript, and read and approved the final manuscript. VK treated the patient at the emergency department. KR performed the surgery. PvR reported the CT scan. DM reported the pathology results.

Competing interests None.

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Learning points

- Splenic angiosarcoma is a rare condition with poor outcome.
- Embolisation for splenic rupture is effective.
- Always include non-trauma-related causes (including splenic tumours) in the differential diagnosis of splenic rupture.

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