

Spontaneous splenic artery rupture in a patient with an unclassified malignant spindle cell tumor of the spleen: report of a case

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Abstract

The purpose of this paper was to describe an unusual manifestation of a sarcoma of the spleen and to raise awareness for spontaneous rupture of the splenic vessels in patients with splenic tumors. A 70-year-old man was admitted to our institution, suffering from left upper quadrant abdominal pain. Upon physical examination, a large and tender abdominal mass was palpated. Abdominal computed tomography showed a heterogenous enlarged spleen with active contrast extravasation from the splenic artery, free intraperitoneal fluid, and a retroperitoneal hematoma. The patient was treated with angioembolization of the splenic artery. Because of rebleeding, splenectomy was performed. Pathology revealed the spleen to be involved with an unclassified malignant spindle cell neoplasm. We concluded that in the case of spontaneous rupture of the splenic artery, accompanied with a radiologic appearance of an enlarged spleen, the diagnosis of sarcoma should be included in the differential diagnosis.

Keywords: Spindle cell, Sarcoma, Spleen, Spontaneous rupture

Case presentation

A 70-year-old man was admitted to the emergency department, suffering from left upper quadrant abdominal pain and constipation for 3 days. The patient denied having sustained trauma to his abdomen. His medical history included essential hypertension, ischemic heart disease, chronic atrial fibrillation, aortic valve replacement in 2002, transient ischemic attack, and a previous laparotomy in 2008 due to intestinal perforation secondary to a 1.2 cm carcinoid tumor of the small bowel with non-malignant features. The patient's current medical treatment included Coumadin (warfarin).

Physical examination upon initial admission revealed an initial blood pressure of 132 mm Hg/79 mm Hg, heart rate of 122 bpm, and normal body temperature. A large tender abdominal mass was palpated in the left upper quadrant of his abdomen. Laboratory evaluation revealed an initial hemoglobin value of 9.8 g/dL and an INR of 5.2. The rest of the routine blood work results were within normal limits.

Upon admission, the patient was resuscitated with intravenous fluids, which included 2 units of packed red blood cells, 3 units of fresh frozen plasma, and intravenous vitamin K. The patient was then referred for abdominal computed tomography (CT), which revealed an enlarged heterogenous spleen. Furthermore, there was active contrast extravasation from the proximal splenic artery and a moderate amount of the free intraperitoneal fluid (Fig. 1). The radiologic findings were interpreted as spontaneous rupture of the spleen. Because of his severe comorbidities and exceedingly high operative risk, we chose to perform angiographic embolization. Hemorrhage was controlled following this procedure (Fig. 2). Sixteen hours later, the patient showed signs of recurrent bleeding, with the appearance of hypotension and a

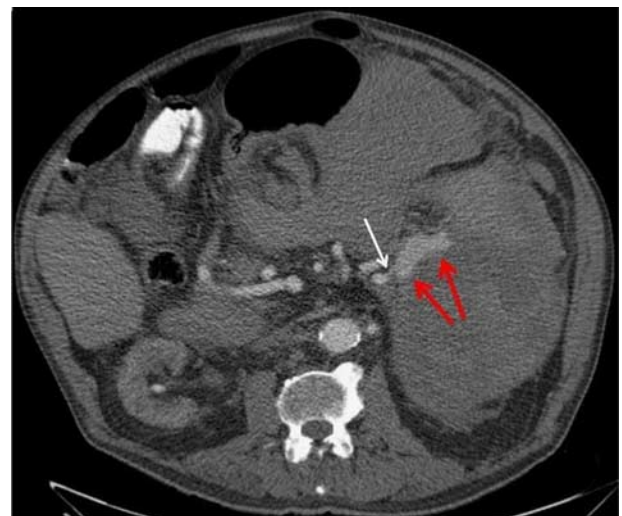


Figure 1. Abdominal computed tomography revealing a huge heterogenous spleen and active contrast extravasation (red arrows) from the proximal splenic artery (white arrow), and a moderate amount of the free intraperitoneal fluid.

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Figure 2. Contrast extravasation seen during fluoroscopy and angiographic embolization. The arrows denote contrast extravasation.

decrease of hemoglobin concentration to 6.4 g/dL. The patient was immediately taken to the operating room.

Exploratory laparotomy revealed free blood in the abdomen and a large retroperitoneal hematoma. The spleen was found to be massively enlarged, and heterogenous, but without any signs of the intraparenchymal hemorrhage (Fig. 3). Bleeding from the proximal splenic artery was found on exploration of the retroperitoneal hematoma. Hemostasis was achieved by ligation of the vessel, and splenectomy was performed. Following the operation, the patient gradually recovered and was eventually discharged home.

Macroscopically, the excised spleen's dimensions were 28 cm × 19 cm × 14 cm, and it weighed 2730 g. The microscopic findings revealed the splenic mass to be composed of sheets of spindle cells with large atypical nuclei, accompanied by mononuclear inflammatory cells (Fig. 4). The tumor was completely excised with the noninvolved surgical margins (negative margins). Although mitotic images were not conspicuous, the proliferative index by the Ki 67 marker was above 80%. Of all immunostains, the spindle cells stained positive only with vimentin, which did not help in differentiating between different



Figure 3. The surgical specimen. The spleen was found to be massively enlarged, heterogenous, but without any signs of the intraparenchymal hemorrhage.

tumor types. Accordingly, the specimen was reviewed by a well-known pathologist from Harvard University, who despite performing extensive immunostains, also failed to demonstrate a specific line of differentiation in the tumor cells. Examination of the ruptured splenic artery revealed no evidence of tumor. The patient was discharged with the final diagnosis of unclassified malignant spindle cell neoplasm of the spleen, most likely unclassified sarcoma. This case was discussed in our tumor board. Because of his comorbidities and unclear advantages of adjuvant treatment, it was decided to offer this patient clinical follow-up only.

Discussion

Most of the primary tumors of the spleen are benign and originate from the vascular endothelium^[1,2]. These include hemangioma, hamartoma, littoral cell angioma, lymphangioma, hemangioendothelioma, and hemangiopericytoma. With the exception of lymphoma involving the spleen, other primary and secondary malignant neoplasms are relatively rare. Malignant neoplasms in the

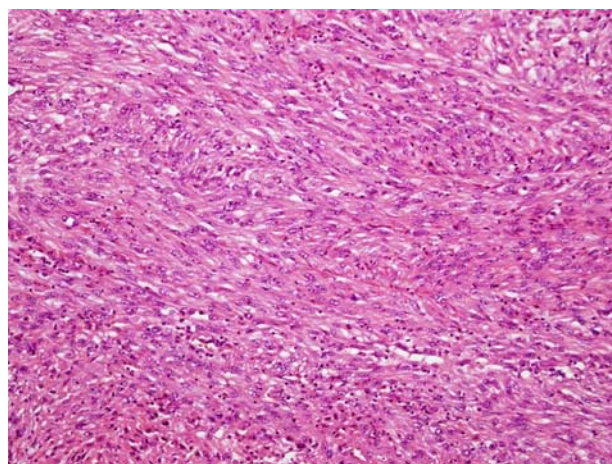


Figure 4. The microscopic findings revealed the splenic mass to be composed of sheets of spindle cells with large atypical nuclei, accompanied by mononuclear inflammatory cells.

spleen are more commonly metastatic. These are secondary to breast, lung, melanoma, colorectal, and ovarian carcinomas^[3,4]. Within this group of malignancies, solitary involvement of the spleen is rare but has been reported^[5]. Sarcoma is the most common primary, nonhematologic malignancy of the spleen. Nonetheless, its occurrence is rare. In 1912, Council first described primary sarcoma of the spleen^[6]. Before the introduction of immunohistochemistry stains, all these tumors were classified as “fibrosarcoma.” In 1982, Govoni et al^[7] published a first case of malignant fibrous histiocytoma. Of the subtypes known today, angiosarcomas seem to be the most aggressive, with 80% of the cases presenting with multiple metastases at the time of diagnosis^[8].

The common clinical manifestations of spleen sarcoma include weight loss, fatigue, anemia, and persisting fever. The most common physical finding is splenomegaly, appearing in 50% of cases^[7]. It is worth mentioning that this patient did not suffer from any of the above mentioned symptoms. His initial albumin was 3.8 g/dL, indicating a normal nutritional state. In splenic sarcomas, laboratory examinations can show cytopenia, leukocytosis, thrombocytosis, and elevated erythrocyte sedimentation rate. Anemia and thrombocytopenia are presented in > 50% of cases^[9,10]. Review of his previous medical history did not show any indications of splenomegaly. However, a revision of the CT scan performed at the time of bowel perforation, which occurred 4 years earlier, revealed a 2.8 cm tumor in the upper pole of the normal-sized spleen. A second CT scan, performed in the following 8 months due to renal colic, showed a similar splenic mass with the same dimensions. This observation suggested the slow development of this tumor in this specific patient.

The most dramatic manifestation of spleen sarcoma is spontaneous rupture. We did not find any clinical series describing similar cases in the relevant English literature. There are few case reports of such cases. In none of these reports was the diagnosis confirmed preoperatively. The prognosis is dismal. Wick and colleagues reviewed all spleen sarcoma cases treated in the Mayo Clinic during a 51-year period. In their series, which included 9 patients, the 6-month survival rate was 21%^[10]. Patients presenting with spontaneous rupture fared worse^[11–13].

The diagnosis of sarcoma in our patient was made only after pathologic evaluation of the resected spleen. During the operation the spleen appeared to be abnormal, suspicious for tumor, but not ruptured. Rather, the source of the bleeding was found proximally in the splenic artery. We speculated that the pathologic involvement of the splenic artery in the tumor would lead to

spontaneous vascular rupture and hemorrhage. However, pathologic examination did not reveal evidence of neoplasm involvement in the area of arterial rupture.

In summary, this case demonstrates the existence of an unclassified malignant spindle cell neoplasm in the spleen, which is extremely rare. We conclude that in cases of spontaneous rupture of the splenic artery in patients with solid splenic tumors, the diagnosis of sarcoma may be included in differential diagnosis.

Conflict of interest statement

The authors declare that they have no financial conflict of interest with regard to the content of this report.

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