Spontaneous Splenic Rupture Secondary to Angiosarcoma

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Angiosarcoma of the spleen is a rare tumor with a very poor prognosis. Review of the world literature reveals only 57 reported cases of this neoplasm. Sixteen of these presented with an acute abdomen secondary to splenic rupture. Patients with this tumor have a mean survival time of 14.4 months after detection, and this decreases to 4.4 months after splenic rupture. The only rational treatment is splenectomy prior to splenic rupture. This paper describes another case presenting as spontaneous splenic rupture.

Angiosarcoma is a rare, soft tissue neoplasm. It probably accounts for less than 1% of all sarcomas. Angiosarcomas of deep soft tissues are rare, constituting only 25% of all angiosarcomas. Deep tissues most often involved are breast, liver, and bone, with spleen being primarily involved less frequently. Until 1985, only 57 cases of primary angiosarcoma of the spleen had been reported. Of these, 16 presented as an acute abdomen secondary to spontaneous splenic rupture. This report describes an additional case and reviews the literature.

Case Report
A 27-year-old man presented to the emergency room complaining of severe abdominal pain of 2 hours duration. The patient had been admitted 2 weeks earlier with severe lower thoracic “back pain.” Evaluation with thoracic spine x-rays, CT scan of thoracic spine, and blood work revealed no abnormalities, and the patient was discharged. There was no history of recent trauma.

Past medical history included an astrocytoma of the brain 2 years prior to this admission. He was treated with craniotomy and radiation therapy with no known recurrence.

Physical examination revealed a white male with pale, clammy skin, hypotension, and tachycardia. The abdomen was diffusely tender, especially in the upper quadrants. Peripheral blood showed a normochromic anemia (Hgb-10.4gm/ml), leukocytosis (WBC-17,000cm³), and thrombocytopenia (Plt-116,000cm³). CT of the abdomen revealed a large amount of free intraperitoneal blood and a non-homogeneous spleen that looked suspicious for anterior rupture. The patient was resuscitated with intravenous fluids and blood, with rapid stabilization of vital signs. At laparotomy 4 hours after admission, the patient was found to have approximately 6 units of blood in the peritoneal cavity, with a rupture of the inferior pole of the spleen. Splenectomy was performed and the abdomen was explored. Multiple small hemangiomata on the anterior surface of the liver were noted.

Pathologic examination revealed a 117 gm spleen with multiple lacerations over the anterior surface and several hemorrhagic parenchymal cysts (Fig 1). Microscopic review noted multiple tumors containing cavernous vascular spaces lined by atypical cells with hyperchromatic nuclei. This pathologic picture
was consistent with primary hemangiosarcoma of the spleen (Fig 2).

The early postoperative course was uneventful, and the patient was discharged on the sixth postoperative day. Four weeks after discharge the patient returned complaining of increasing abdominal pain and was found to have enlargement of his liver. Despite a course of radiation therapy and aggressive chemotherapy, the patient deteriorated and died 112 days after diagnosis. The cause of death was thought to be exsanguinating intra-abdominal hemorrhage secondary to metastatic disease. Autopsy was not performed.

**Discussion**

Langhans first described primary angiosarcoma of the spleen in a 30-year-old man in 1894. Since then there have been 56 additional cases, making the total reported cases 57. The disease apparently affects males and females with equal frequency, and the average age of those affected was 47 years. Thirty patients were 50 years of age or older, and only 4 were less than 20 years of age.

The pathogenesis of this tumor is unknown. Thorium dioxide (thorotrust) has been implicated as a carcigenic agent closely linked to osteogenic sarcoma and Kupffer cell sarcoma of the liver. There has been no significant history of exposure in patients developing sarcomas of the spleen. Some think that angiosarcomas represent malignant transformation in preexisting hemangionas or hamartomas.

The most common presenting complaint is abdominal pain, frequently localized to the left upper quadrant and probably due to splenic enlargement. Approximately 1 of 3 patients present dramatically with splenic rupture, as in this case. Splenic rupture is an extremely bad prognostic event as the mean survival time in these patients is 4.4 months compared to 14.4 months in patients who have had splenectomy prior to rupture. Our patient survived 3.7 months after splenic rupture.

Hematologic findings in patients with splenic angiosarcoma frequently include anemia, thrombocytopenia, and leukopenia. The mechanism producing these abnormalities is not completely understood. Platelets may be consumed in the stagnant vascular spaces within the tumor. Anemia, found in 70% of patients, is thought to be due to a microangiopathic process resulting in red cell damage and hemolysis.

At laparotomy, both splenic and hepatic involvement are usually found. De Navasque has suggested that the organ with the largest gross tumor is the primarily involved organ. This case, with smaller hepatic lesions, represents dissemination via the splenic vein.

According to Chen and associates, metastases most commonly involved, in decreasing frequency, are liver, lung, bone, and lymph nodes.

Currently the best treatment for this tumor is early splenectomy, but the high incidence of metastases present when tumor is detected limits surgical therapy as a curative modality. The rarity of this tumor has made it difficult to assess the effect of radiation and chemotherapeutic agents on this neoplasm.

**Conclusion**

Splenic angiosarcoma is a very rare tumor with an extremely poor prognosis. The tumor is usually diagnosed very late, and the current modes of treat-
have been ineffective in dealing with this neoplasm. Approximately one-third of the tumors present with spontaneous splenic rupture. Splenic rupture is an extremely bad prognostic sign, and the only rational treatment is early diagnosis and splenectomy prior to splenic rupture.

References

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