

CASE REPORT

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Leiomyosarcoma of the Splenic Vein

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Abstract: Leiomyosarcomas arising from the wall of blood vessels are rare and aggressive neoplasm. We report a case of a previously healthy 66-year-old woman who presented with intermittent abdominal pain, progressive constipation, and weight loss. Abdominal computed tomography showed a 12 cm solid heterogeneous tumor in the tail of the pancreas. The patient subsequently underwent surgical resection of the pancreatic mass. Surprisingly, histological and immunohistochemical analyses revealed leiomyosarcoma arising from the smooth muscle of the splenic vein. After surgery, she received adjuvant chemotherapy. One year later, there was no evidence of local recurrence. In this paper, we discuss the available information about leiomyosarcomas of splenic vein and its management.

Keywords: sarcomas of the great vessels, leiomyosarcoma, splenic vein

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Background

Leiomyosarcomas (LMSs) are tumors of mesenchymal origin that usually appear in the abdominal cavity/retroperitoneum or uterus.¹ They are rare and aggressive neoplasms that arise from the blood vessels walls. Tumors of the inferior vena cava account for the majority of the cases and are often detected in middle-aged women.^{1,2}

Clinical manifestations depend on the anatomical site of origin and are related to tumor compression of the surrounding organs.^{3,4} Presentation includes palpable abdominal masses or symptoms related to thrombosis or embolism of the splenic vein as abdominal pain, nausea, and fever.³ Because of their rare occurrence, there is limited data in the literature, and most of it, regarding only aggressive surgical management that can improve long-term survival of these patients.⁵

In this paper, we describe a case of a 66-year-old woman with an LMS of the splenic vein simulating a pancreatic tumor.

Case Report

A 66-year-old Hispanic otherwise healthy woman presented with a 6-month history of intermittent, dull, and colic left upper quadrant abdominal pain, associated with progressive constipation and weight loss of 10 kg. She did not have any previous cancer history, had never smoked, and her family history was only positive for breast cancer in her mother.

The physical examination revealed a large non-tender mobile abdominal mass located in the left upper quadrant. Complete blood count, electrolytes, and comprehensive metabolic panel including liver enzymes were within normal limits.

The abdominal computed tomography (CT) showed a solid heterogeneous 12 × 10 cms mass located in the tail of the pancreas displacing the stomach and compressing the left kidney (Fig. 1). Whole body positron emission tomography (PET) scan did not show metastatic disease. The initial diagnosis of pancreatic cancer was considered, and surgery was scheduled. Intraoperative examination revealed a tumor attached to the posterior wall of the pancreas. For that reason, a distal pancreatectomy and splenectomy were performed, with complete excision of the tumor. After surgery, the patient did not have any surgical complication and a few days later was discharge



Figure 1. Abdominal computed tomography showed a solid heterogeneous tumor of 12 cm in diameter in the tail of the pancreas with central necrosis (blue arrow indicates the tumor).

home with a follow-up appointment in the outpatient oncology clinic.

Macroscopically, a 12 × 9 × 6 cm mass arose from the splenic vein smooth muscle. The mass was a solid and firm gray-brown color tumor, with focal areas of hemorrhage on the cut section (Fig. 2). The splenic vein did not show signs of thrombosis. Microscopically, fusiform cells predominated, with a high rate of atypical mitotic figures. The mitotic rate was 12/10 HPF (per high power field). Tumor cells were positive for smooth cell immunohistochemical markers (smooth muscle actin and desmin) and were negative for epithelial markers (keratin), neuronal markers (S100), and hematopoietic markers (CD34) (Figs. 3 and 4).

Once the patient recovered from surgery, she got adjuvant chemotherapy with 6 cycles of doxorubicin



Figure 2. Cut section of the 12 × 9 × 6 cm grayish-brown color tumor. It was solid and firm with focal areas of hemorrhage arising from the smooth muscle of the splenic vein.

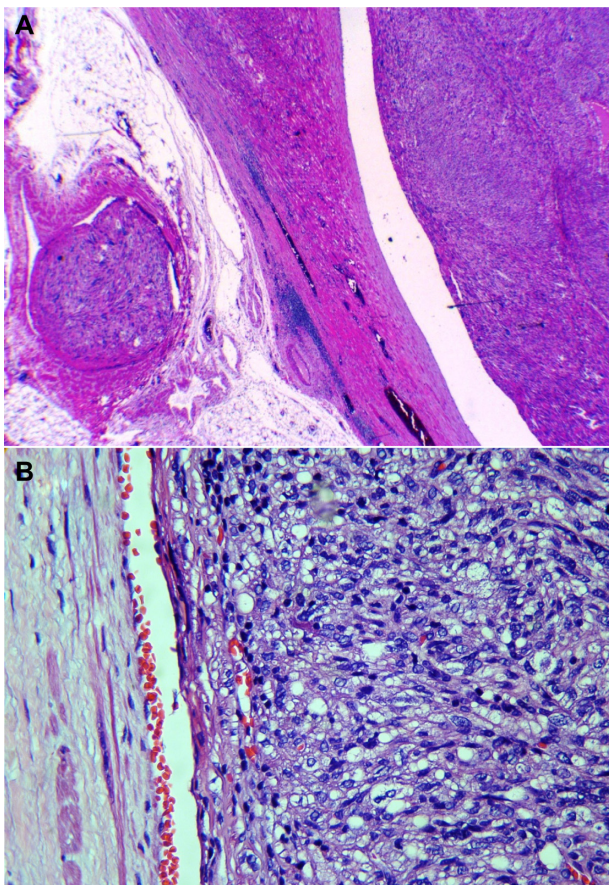


Figure 3. Hematoxylin and eosin stain (4 \times magnifications) revealed predominantly fusiform cells, with a high rate of atypical mitotic figures. (A) Intraluminal tumor growth of a leiomyosarcoma originating from the splenic vein (hematoxylin and eosin stain, $\times 40$ magnification). (B) Hematoxylin and eosin stain ($\times 200$ magnification) revealed predominantly fusiform cells, with a high rate of atypical mitotic figures.

and ifosfamide without any major toxicity. One year later, a whole body PET did not show any evidence of local or distant metastasis. Informed consent to publish this report was obtained from the patient.

Discussion

LMSs are rare malignant mesenchymal tumors derived from smooth muscle that essentially affect the inferior vena cava (IVC). They generally occur intra-abdominally in the retroperitoneum, mesentery, or omentum (40%–45%), in subcutaneous or deep soft tissue of the limbs (20%–30%), in the arrector pili muscle of the skin (15%–20%), and in the walls of blood vessels (5%).⁶ Major vessel LMSs are extremely rare, making up less than 2% of all LMSs and occur in veins approximately 5 times more often than in arteries.¹ The inferior vena cava accounts for 75% of large-vessel LMSs.^{2,4} The retroperitoneal

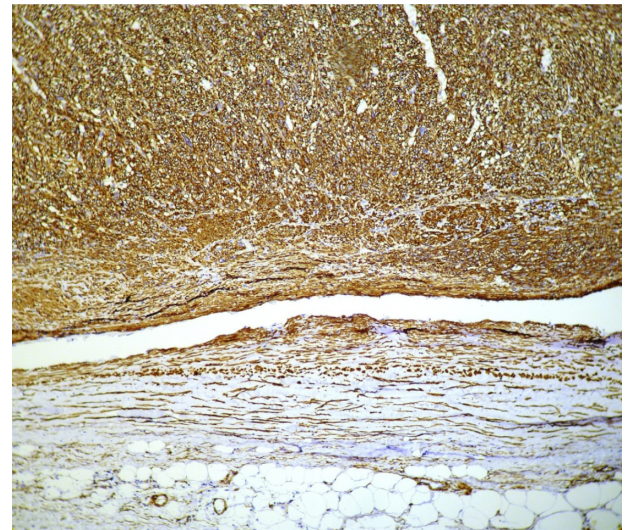


Figure 4. The tumor cells stained strongly positive for smooth muscle actin (100 \times).

veins near the inferior vena cava, including the renal, iliac, ovarian, and spermatic veins, may also be sites of occurrence.

Venous LMSs are the most common malignant vascular tumors. They usually present in middle-aged women between the 5th and 6th decade of life arising from the inferior vena cava or large size veins of the lower extremities.³ Those derived from the splenic vein are exceedingly rare with only 2 previous reported cases in the literature.^{6–8} This is the third reported case and the second in the English language. The previous 2 cases are to be described as follows: (1) A man presenting with epigastric pain and found to have a larger lesion on the CT scan, measuring 15 \times 6 \times 5 cm⁸ and (2) a 58-year-old woman complaining of abdominal pain, with a lesion on the abdominal CT of 3.5 \times 3 \times 3 cm (case of a single patient reported in 2 different articles).^{6,7} In our case, the patient was a 66-year-old woman with a tumor of 12 \times 9 \times 6 cm, the second largest tumor reported in the literature (Table 1).

The clinical symptoms and metastatic potential of these tumors are related to their position in the vessel wall, venous obstruction and compression of the surrounding organs by the mass.⁹ In addition, deep venous thrombosis and even pulmonary embolism have also been reported.¹⁰ According to the site of origin, sarcomas of the great vessels are classified into luminal and mural tumors.⁴ Luminal (intimal) sarcomas of the blood vessels are more common in

**Table 1.** Reported cases of leiomyosarcoma of splenic vein until today.

Author	Year	Patient	Location	Tumor size	Presentation	Treatment and outcome
Rödl ⁸	1988	--, male	Splenic vein	15 × 6 × 5 cm	Epigastric pain	–
Niver ⁶	2011	58, female	Splenic vein	3,5 × 3 × 3 cm	Epigastric pain	Distal pancreatectomy + splenectomy.
Gage ⁷	2012					No recurrence after 15 months
Reported case in this paper	2013	66, female	Splenic vein	12 × 9 × 6 cm	Epigastric pain	Distal pancreatectomy + splenectomy + adjuvant chemotherapy No recurrence after 12 months

arteries, and they are characterized by rapid growth and earlier occurrence of distant metastases due to intraluminal growth.^{2,4} On the other hand, mural sarcomas of the blood vessels are more common in veins, and, unlike the luminal sarcomas, they have a better prognosis, because of their extraluminal and slow growth.^{2,4} Tumor metastases of this slow growing malignant cancer to other organs are relatively infrequent but can be detected in liver, lungs, lymph nodes or bones.¹¹

Histologically, vascular LMSs are identical to those found in other nonvascular sites. Tumors are composed of spindle cells with eosinophilic cytoplasm and centrally placed elongated nuclei with blunt ends forming fascicles. In addition, they are positive for vimentin, smooth muscle actin, desmin, and h-caldesmon.^{1,4}

Regarding imaging techniques, ultrasound is often the first diagnostic tool because it does not expose patients to ionizing radiation, and it is widely available. However, the accuracy of ultrasound for diagnosing pancreatic tumors is only 50% to 70%.¹² For that reason, CT scan and magnetic resonance imaging (MRI) are very important because they allow us to determine the extension of the tumor to surrounding organs and the presence of vein thrombosis and distant metastasis.¹³ A highly sensitive and specific modality for detecting recurrence in posttherapy patients with sarcoma is 18F-fluorodeoxyglucose (FDG) positron emission tomography (PET-CT). However, it provides no significant advantage over CT scan or MRI for this purpose.¹⁴

Early diagnosis is very important for the treatment of this tumor because complete surgical resection is the only therapeutic modality that can cure or prolong patients survival.^{5,11,15} Wide surgical excision of the tumor with a resection margin of 2 to 3 cm is the only available treatment that can cure patients with

this condition. In our case, the surgical treatment was a distal pancreatectomy including splenectomy. Despite the aggressive behavior of this tumor, no consensus yet exists regarding adjuvant treatment. Radiotherapy has been suggested for high-grade soft tissue sarcomas located in the extremities and for intermediate grade tumors with positive margins.^{6,16} Furthermore, it is unclear whether chemotherapy would have made a difference in the prognosis of these patients.⁵ In our case, after surgery, the patient received adjuvant chemotherapy with doxorubicin and ifosfamide but no radiation therapy because the margins were clear. With this treatment, after 12 months, there was no evidence of local recurrence or distant metastasis.

As previously stated, given the rarity of venous-origin LMS, there are limited outcome data of this peculiar type of sarcomas, and, until today, free surgical margins have the most significant prognostic factor. In a study of 25 patients with primary inferior vena cava LMS over a 20-year period, patients who underwent complete resection had a 3-year survival rate of 76%, while there were no survivors at all after 3 years among those patients with incomplete resection. In these cases, a complete resection of the tumor was possible, and the IVC was repaired primarily via surgical means without a too high risk of postoperative edema.¹¹ Kieffer et al, based on the treatment outcomes of 22 patients with LMS inferior vena cava, recommended an aggressive surgical removal of the tumor by means of modern vascular surgery in combination with chemotherapy and/or radiotherapy.¹⁷ Reconstruction of the IVC using a prosthesis made of reinforced polytetrafluoroethylene (PTFE) facilitates a complete resection of the tumor and prevents clinical signs of venous congestion.¹⁸

To reduce tumor size and increase the resection rate, a preoperative neoadjuvant therapy can be undertaken. If, however, a complete tumor resection



is not possible, tumor reduction followed by radiation therapy represents a good palliative treatment option.¹⁹ In addition, a combination of chemotherapy and radiotherapy has been reported as being superior to radiotherapy alone with respect to an increase in survival rate.²⁰ In 3 different treatment series with sufficient case numbers and follow-up, a 5-year survival rate was achieved in 53%,²⁰ 35%,¹⁷ and 33%¹¹ of the cases, respectively, after radical resection followed by a curative approach using adjuvant therapy.

Although histological tumor grade has been demonstrated to have a profound influence on patient survival for most sarcomas, it has not been described to affect patient survival in inferior vena cava sarcomas.^{15,21}

Because of the insidious evolution of this neoplasm, follow-up imaging studies should be exhaustive, and is even more necessary than in other tumor types, to find out local recurrence or distant metastasis.^{1,2,4} The inferior vena cava LMS incidence is lower than that of the other sarcomas of the retroperitoneum, 11% versus 26% (*P* value not indicated in the literature), respectively.²⁰ However, large-vessel LMSs seem to follow the trend of other soft tissue sarcomas metastasizing primarily to the lungs, with the liver being the second most common location.²² In general, the incidence of metastases after surgical resection is reported to be approximately 40%.^{2,23}

Conclusion

LMS of the splenic vein is an extremely rare entity, occurring predominantly in middle-aged individuals. In this paper, we report the clinical presentation, imaging techniques, pathology, treatment, and outcome of the third case in the literature. Unfortunately until now, there are not enough cases to establish the best treatment, prognosis, and long-term results, perhaps due to the rarity of this tumor or because of a lack of case publications. Therefore, all these reasons mentioned above highlight the importance of the presented case.

Author Contributions

Read the pathology slides, diagnosed the patient: CA. Suggested case publication: CA. Searched the literature, reviewed the case history and contributed to the writing of the manuscript: FS, CA, JD, NG, GH. Obtained the pathology imaging: CA. Made critical

revisions and approved final version: FS. All authors reviewed and approved of the final manuscript.

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Competing Interests

Author(s) disclose no potential conflicts of interest.

Disclosures and Ethics

As a requirement of publication the authors have provided signed confirmation of their compliance with ethical and legal obligations including but not limited to compliance with ICMJE authorship and competing interests guidelines, that the article is neither under consideration for publication nor published elsewhere, of their compliance with legal and ethical guidelines concerning human and animal research participants (if applicable), and that permission has been obtained for reproduction of any copyrighted material. This article was subject to blind, independent, expert peer review. The reviewers reported no competing interests.

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