Leiomyosarcoma of the Inferior Vena Cava: Case Report and Review of Literature.

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Abstract

Leiomyosarcomas (LMS) account for 5–10% of soft tissue sarcomas (A). Leiomyosarcomas of the Inferior Vena Cava (IVC) are extremely rare. The first case was reported by Perl on 1875 and, since then, less than 30 cases have been reported worldwide [F]. About two thirds of all retroperitoneal leiomyosarcomas and three fourths of all vena cava leiomyosarcomas occur in women [A, G, I]. We present the case of a leiomyosarcoma of the inferior vena cava in Puerto Rico.

Introduction

A 54-year-old man presented severe, intermittent, dull, right flank pain for three weeks before admission. The pain became severe four days before hospitalization. General symptoms of weight gain, malaise, weakness and decreased exercise tolerance were also identified. The patient has a history of diabetes mellitus type 2, hypertension, diabetic retinopathy and neuropathy, strokes, and was a chronic smoker (30 packs/year) that quit approximately seven years ago. He has had an appendectomy, cholelithectomy and drainage of a toe abscess. He also has paternal and maternal history for hypertension and heart disease.

An abdominal and pelvic CT scan with IV contrast revealed a mass at the infrarenal inferior vena cava, measuring 80.82 mm and extending to the caudate lobe of the liver (Fig. 1). A needle biopsy of the mass confirmed the diagnosis of LMS. Further immunohistochemistry studies support a diagnosis of Leiomyosarcoma (See Table 1).

A biopsy of a mass in the right upper lobe of the lung identified by CT scan of the thorax, confirmed a metastatic leiomyosarcoma. Resection of the IVC tumor with ligation of the vena cava was performed. The resected segment of the vena cava measured 10 x 6.5 x 4.5 cm, with a mass measuring 10 cm. On cross section, the tumor presented areas of necrosis and hemorrhage and was growing inside the lumen.

Microscopic examination revealed a Leiomyosarcoma of FNCLCC histologic grade 2 (24 mitoses per 10 HPF - score 3; tumor necrosis <50% - score 1; differentiation - score 1). Peripherally oriented fascicles of spindle cells were observed with bright eosinophilic cytoplasm and blunted-ended nuclei. The tumor staging was pT2b NX M1 [C]. The post-operative course was unremarkable. In addition, the patient was treated with chemotherapy.

Discussion

Leiomyosarcomas of vascular origin comprise of a seemingly rare group [A]. They are most common in large vessels such as the inferior vena cava (A). IVC leiomyosarcomas occur during middle or late adult life, on average at 50 years of age, and are also most common in women.

The segment of the IVC involved is very important since it determines the symptoms and the possibility of resection (Figure 2). Most tumors arise in the lower or middle portion, while only a small number arise from the upper third. Patients generally present nonspecific symptoms, such as abdominal pain, tenderness, exhaustion and weight loss. In segment III tumors, patients may present Budd Chiari syndrome (hepaticomegalgy, jaundice and ascites). Conversely, tumors in segment II may result in renal dysfunction and hypertension in segment I cause lower extremity edema. Segment II tumors also produce right upper quadrant pain and tenderness, which may mimic biliary tract disease [A]. Our patient had middle portion (segment II) involvement but did not present renal dysfunction.

Pathologic staging of sarcomas is essential for therapeutic strategies. It consists of the determination of histologic type, grade, and tumor size and depth. As our case is a deeply situated tumor, measuring more than 5 cm, it is considered to be T3B and, regional lymph nodes could not be assessed (NX). A LMS of the IVC more frequently metastasize to the lung, as in our case [M1]. Grading and staging of soft tissue sarcomas is associated with disease specific survival. Our case was a pT2b Nx M3 (stage four) with a histological grade 2 (FNCLCC).

Modern imaging modalities allow for an early and accurate preoperative diagnosis, resulting in a higher rate of surgical resection and improved survival [I]. Surgical resection remains the current treatment of choice for primary leiomyosarcomas of the IVC [G]. In this case, resection of the tumor with ligation of the vena cava was the appropriate treatment, along with subsequent chemotherapy for prevention of recurrence and treatment of the metastasis found in the right upper lobe of the lung. General survival rates for IVC leiomyosarcomas are 5 years (50%) and 10 years (8%) [I]. The recurrence rate of these tumors is high and, even following a complete primary resection, more than 50% of patients develop a late recurrence [D]. The patient in this presentation, in his last follow up visit after surgery, was alive with no evidence of tumor recurrence, and only presented anemia.

Conclusion

1. Leiomyosarcomas of the inferior vena cava are very rare.
2. The tumors are most frequently present in patients over 50 years of age and in females.
3. Symptoms depend of the segment of the IVC involved.
4. The IVC segment most frequently involved is the middle segment.
5. Metastases are seen more frequently in the upper segment of the IVC.
6. The principal symptom is abdominal pain.
7. The use of appropriate immunohistochemistry panels is important to confirm the diagnosis of LMS.
8. A CT scan of the abdomen is the most useful study to determine the location of the tumor and treatment plan.
9. The preferred treatment is surgical resection of the tumor followed by chemotherapy or radiotherapy.
10. To our knowledge, this is the first case reported of Leiomyosarcoma of the inferior vena cava in Puerto Rico.

References