

Retroperitoneal Leiomyosarcoma of the Inferior Vena Cava: A Case Report

A Karami¹, S Azizi*², A Payombarnia², N Rezaee², N Azarpira³

¹Department of Surgery, ²Department of Pathology, Iranian Hospital, Dubai, UAE, ³Department of Pathology, Shiraz University of Medical Sciences, Shiraz, Iran

Abstract

Leiomyosarcoma of the inferior vena cava (IVC) is a rare sarcoma, but it is the most common primary malignancy of the IVC. It has an extremely gloomy prognosis. We describe a 40 year-old white female complaining of abdominal fullness for 7 weeks before she sought medical assistance. Initial work-up including sonography and computed tomography showed a huge tumoral mass in the abdominal cavity seemingly originating from the IVC with displacement of the right ureter and hydronephrosis. The patient underwent surgical resection of the tumor. Pathologic diagnosis was leiomyosarcoma. Postoperatively, she was placed on coumadin and adjuvant chemotherapy was started. Considering the aggressiveness of this tumor, early radical enblock resection with clear margins is still the only chance for long-term survival.

Keywords: Retroperitoneal; Leiomyosarcoma; Inferior vena cava

Case Report

A forty year-old woman from Nigeria was admitted in the Emergency Department of Iranian Hospital in Dubai, UAE affiliated to Iranian Red Crescent Society with severe abdominal pain. She had progressive abdominal distention with nausea during the last four months. In general abdominal examination, the patient had abdominal pain without tenderness. No evidence of ascitis was detected. There was lower extremity edema. Abdominal sonography showed a large abdominal mass containing cystic and solid parts. A computed tomographic scan revealed a large abdominal mass with multilocular mixed density pattern, occupying all parts of the abdomen with displacement of the right and left kidneys with grade II hydronephrosis in the right kidney (Figures 1A, 1B). The uterus and both ovaries were normal. Chest X-ray was normal.

Complete blood count and chemistry were within normal limit. She underwent laparotomy. A huge

retroperitoneal abdominal mass which seems to be originated from right sided IVC was attached to ascending, transverse colon, right ureter and kidney. The lesion had a fish fleshy, pinkish color and soft consistency with 10 kg weight. The specimen was fixed in 10% formalin. In the pathologic study, the tumor cells were arranged in clearly seen fascicles. Individual tumor cells were characterized by mild nuclear pleomorphism, central location of blunt ended, oval-shaped nuclei, and often perinuclear vacuolization. The mitotic rate was low (Figure 2). These findings were in favor of a sarcoma. Leiomyosarcoma, rhabdomyosarcoma, gastrointestinal stromal tumor (GIST), malignant peripheral nerve sheath tumor should be considered in differential diagnosis. The tumor was examined for S-100 protein, neurofilament, neuron specific enolase, desmin, smooth muscle actin (SMA), CD34, CD117 and vimentin (Dako, Denmark), using the Envision Kits technique (Dako, Denmark). Immunohistochemically, the cells were strongly positive for SMA (Figure 3) and vimentin. Based on both histological and immunohistochemical findings, the diagnosis was well differentiated leiomyosarcoma. The postoperative course was unremarkable. She received radiotherapy and adjuvant chemotherapy. The patient is free of recurrent disease 4 months after her surgery.

*Correspondence: Saeid Azizi, MD, Department of Pathology, Iranian Hospital, Postal code: 2330, Dubai, UAE. Tel: +971-5-09385022, Fax: +971-4-3440322, e-mail: saeed_azizi2003@yahoo.com
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1A



1B

Figs 1A, 1B: Computed tomography scan of the abdomen, showing large retroperitoneal mass with right sided hydronephrosis.

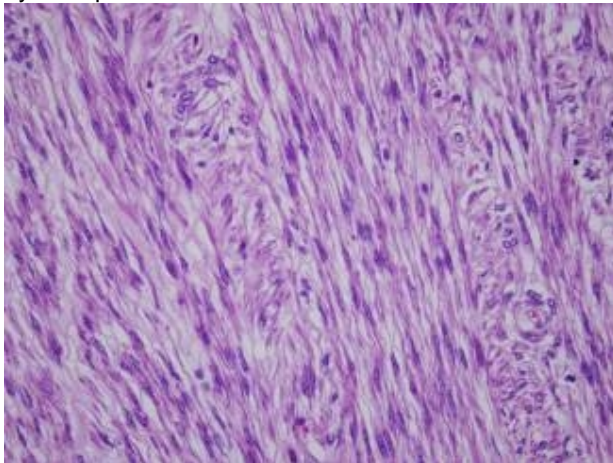


Fig 2: Tumoral cells in long fascicle with cigar shaped nuclei (H & E \times 100).

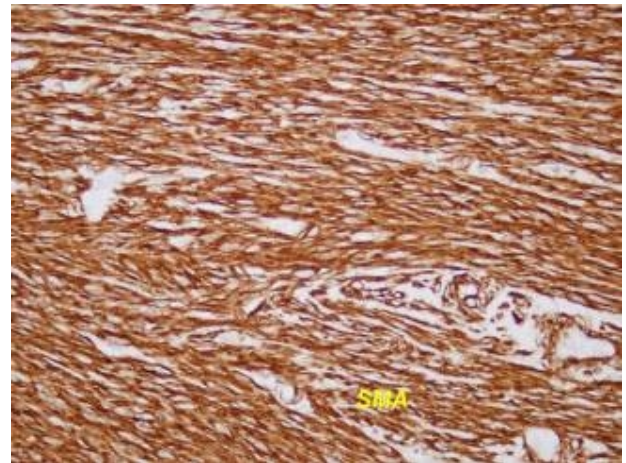


Fig 3: Positive immunochemical staining for SMA in tumor cells (IHC \times 400).

Discussion

Primary tumors arising from great vessels like the aorta, pulmonary artery or inferior vena cava (IVC) are rare.¹ The last one is the commonest site of its occurrence. It arises from the smooth muscle cells of the vessel wall. The first report of this tumor came in 1871 from Perl, who diagnosed this tumor at a postmortem examination.² Because of the limited case numbers, conclusions about the natural history and optimal treatment have been difficult to establish. We describe a case of primary inferior vena

cava tumor involving all three segments of the abdominal inferior vena cava including infrarenal, suprarenal and retrohepatic vena cava, along with the right kidney. It was resected without reconstruction of the IVC. The patient is doing well four months after the surgery without having any renal insufficiency, hepatic insufficiency or leg edema and has optimum quality of life.

In the literature review, there were a series reported by Hines et al. (14 cases),³ Hollenbeck et al. (25 cases),⁴ Dzsinič et al. (8 cases)⁵ and Kieffer et al.,⁶ of patients presenting with leiomyosarcoma of

the IVC. According to Kieffer et al.⁶ findings, the most common presenting symptom was pain in the right flank that was observed in all. This symptom was often long-lasting and misled the diagnosis. Our Findings are in agreement with the most data described in the register of Mingoli et al.¹ with regard to female predilection, frequency of abdominal pain as presenting symptom and association with edema of the lower extremities.

The goals of management of these tumors include the achievement of local control, maintenance of venous return, and prevention of recurrence. Diagnostic imaging techniques, especially modern computerized tomography and magnetic resonance imaging, have advanced and allow excellent preoperative imaging, evaluation, and preparation.^{7,8}

Tumors were graded by examination of the formalin fixed, paraffin embedded, hematoxylin and eosin sections from well-differentiated tumors (Grade 1) to poorly differentiated (Grade III).⁹ It is well known that grade impacts significantly other soft tissue sarcomas with regard to prognosis and therapeutic recommendations. However, grade did not predict recurrence or survival. A recent literature review of cases revealed a trend with a quoted distant recurrence rate of 31% for Grade 1 and 2 tumors and a rate of 43% for Grade 3 tumors.⁹ However, others have not found a correlation between mitotic activity and outcome.^{9,10} Age, gender, tumor size, tumor grade, and lymph node status did not impact survival in this series.¹¹⁻¹⁵ The pathway of distant metastatic disease in pelvic sarcomas is also

different from extremity sarcomas since the majority are non-pulmonary metastases.

The long-term outcome of surgery for leiomyosarcoma of the IVC has been disappointing. In the international register of Mingoli et al.,¹ actuarial survival rates at 5 and 10 years were 49.4% and 29.5%, respectively, but the cancer-free actuarial survival rates at 5 and 10 years were 31.4% and 7.4%, respectively. In the three series with sufficiently large populations and satisfactory follow-up, actuarial survival rates 5 years after radical resection with curative intent were 53.3%,³ 33%,⁴ and 34.8%.⁶ It is also noteworthy that some surviving patients present local recurrence or distant metastases responding to various degrees to adjuvant treatment.

Surgery, whether performed alone or in combination with chemotherapy and possibly radiation therapy, is generally not curative but it constitutes the only hope of prolonged survival. Long term survival is related to an extensive surgery, being surgical margins free of tumor as the only prognostic factor with significant influence on patient's survival.

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References

- Mingoli A, Cavallaro A, Sapienza P, Di Marzo L, Feldhaus RJ, Cavallari N. International registry of inferior vena cava leiomyosarcoma: analysis of a world series on 218 patients. *Anticancer Res* 1996; **16**:3201-5. [8920790]
- Perl L. Ein fall von sarkom der v cava inferior. *Virchow's Arch Pathol Anat* 1871; **53**:378-83.
- Hines OJ, Nelson S, Quinones-Baldrich WJ, Eilber FR. Leiomyosarcoma of the inferior vena cava: prognosis and comparison with leiomyosarcoma of other anatomic sites. *Cancer* 1999; **85**:1077-83. [10091791]
- Hollenbeck ST, Grobmyer SR, Kent KC, Brennan MF. Surgical treatment and outcomes of patients with primary inferior vena cava leiomyosarcoma. *J Am Coll Surg* 2003; **197**:575-9. [14522326] [doi:10.1016/S1072-7515(03)00433-2]
- Dzsinich C, Gloviczki P, van Heerden JA, Nagorney DM, Pairolo PC, Johnson CM, Hallett JW Jr, Bower TC, Cherry KJ Jr. Primary venous leiomyosarcoma: a rare but lethal disease. *J Vasc Surg* 1992; **15**:595-603. [1560548] [doi:10.1067/mva.1992.34346]
- Kieffer E, Alaoui M, Piette JC, Cacoub P, Chiche L. Leiomyosarcoma of the inferior vena cava: experience in 22 cases. *Ann Surg* 2006; **244**:289-95. [16858193] [doi:10.1097/01.sla.0000229964.71743.db]
- van Rooij WJ, Martens F, Verbeeten B Jr, Dijkstra J. CT and MR imaging of leiomyosarcoma of the inferior vena cava. *J Comput Assist Tomogr* 1988; **12**:415-9. [3366953] [doi:10.1097/00004728-198805000-00011]
- Bretan PN Jr, Williams RD, Hricak H. Preoperative assessment of retroperitoneal pathology by magnetic resonance imaging. Primary leiomyosarcoma of inferior vena cava. *Urology* 1986; **28**:251-5. [3750612] [doi:10.1016/0090-4295(86)90057-9]
- Kulaylat MN, Karakousis CP, Doerr RJ, Karamanoukian HL, O'Brien J, Peer R. Leiomyosarcoma of the inferior vena cava: a clinicopathologic review and report of three cases. *J Surg Oncol* 1997; **65**:205-17. [9236931] [doi:10.1002/(SICI)1096-9098(199707)65:3<205::AID-JSO11>3.0.CO;2-2]
- De Waade B, Van Belle S, Clas H, Goovaerts G, Delvaux G, Willems G. Retroperitoneal sarcoma. *Acta Chir Belgium* 1987; **87**:229-34.
- Jurayi MN, Midell AI, Bederman S, Gruen J, O'Brien PH. Primary leiomyosarcoma of the inferior vena

- cava: report of a case and review of the literature. *Cancer* 1970;**26**:1349-53. [5483665] [doi:10.1002/1097-0142(197012)26:6<1349::AID-CNCR2820260623>3.0.CO;2-#]
- 12 Kevorkian J, Cento DP. Leiomyosarcoma of the large arteries and veins. *Surgery* 1973;**73**:390-400. [4687797]
- 13 Bruyninckx CM, Derksen OS. Leiomyosarcoma of the inferior vena cava: case report and review of the literature. *J Vasc Surg* 1986;**3**:652-6. [3959263] [doi:10.1067/mva.1986.av0030652]
- 14 Griffin AS, Sterchi JM. Primary leiomyosarcoma of the inferior vena cava: a case report and review of the literature. *J Surg Oncol* 1987;**34**:53-60. [3807376] [doi:10.1002/jso.2930340114]
- 15 Bailey RV, Stribling J, Weitzner S, Hardy JD. Leiomyosarcoma of the inferior vena cava: report of a case and review of the literature. *Ann Surg* 1976;**184**:169-73. [952564] [doi:10.1097/00000658-197608000-00006]