Leiomyosarcoma of the inferior vena cava: report of a case

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ABSTRACT

Leiomyosarcoma of inferior vena cava is a rare soft tissue sarcoma presenting with vague abdominal pain. Complete surgical resection with clear surgical margin plays a central therapeutic role. The effect of chemotherapy and radiation therapy is controversial. We reported a female patient with invasion both renal veins by this disease. It was surgically excised and replaced with graft and right renal vein was anastomosed to graft without any complications. Histopathological examination of the specimen grade II leiomyosarcoma arising from the vena cava. She is alive at 36th month.

Key words: Graft, leiomyosarcoma, vena cava inferior

CASE REPORT

Our patient was a 63-year-old woman. During the last 3 months her main complain was colicky abdominal pain located in the right upper quadrant. Physical examination was unremarkable. Abdominal ultrasound reported a conglomerated lymph nodes mass of 66 x 48 mm$^2$ in size compressing portal vein and spleno-portal union at pancreatic level. Abdominal CT and MRI showed a heterogeneous, solid mass of 7 x 7 x 4 cm$^3$ which was located between inferior edge of quadrate lobe of liver and third portion of duodenum. It was posterior to left kidney and left adrenal gland, compressing IVC and right renal vein. The pathologist reported a diagnosis of low grades LMS after ultrasound-guided true-cut biopsy led us to a diagnosis of a retroperitoneal LMS. During operation, we recognized a 10 cm mass originated from subhepatic IVC, adjacent to caudate lobe of the liver, displacing the pancreas and duodenum to the anterior. Both renal veins were invaded by the tumor (Figure 1). Left renal vein had not function, but another inferior polar left renal

Figure 1: Intraoperative photograph of the tumor. VCI, vena cava inferior; RRV, right renal vein

Leiomyosarcoma (LMS) is 10–15% of soft tissue sarcomas, but represent 45% of all retroperitoneal malignancies.$^{[1]}$ LMS arising from inferior vena cava (IVC) are very rare and reported over 200 cases since by Perl’s in 1871.$^{[2]}$ It originates from media of the vein and can expand intraluminally, extraluminally or both. We report our surgically treated patient with LMS of the IVC.

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vein draining the kidney. It was cut. IVC was cross-clamped 2 cm proximal and distal to the tumor. A 10 cm segment of IVC containing the tumor was excised together with right and left renal veins. A dacron graft (Sulzermedica, Sulzer Vascutek Ltd, Renfrewshire, Scotland) with a 20 mm of diameter was placed instead of the vena cava (Figure 2). The right ven was anastomozed to graft. The pathologic specimen was (9 x 9 x 3 cm³) encapsulated LMS with grade II histological type. Surgical margins were negative. Adjacent lymph nodes have not contained any metastasis. Patient was discharged without any complication. She is alive at postoperatively 36th month.

DISCUSSION

The LMS of the IVC is a rare soft tissue sarcoma originates from media of vein, and can expand extra or intraluminally or bi-directional fashion, like our patient. Site of the tumor can be described in relation to hepatic and renal veins; as level 1 above the hepatic veins, level 2 between hepatic and renal veins, level 3 between renal veins and iliac vein confluence.[3] The tumor of IVC was classified as level 2 in our patient.

Like our 63-year old case, 80% of such patients are females, with a mean age of 58 years. Silent symptoms of such tumors are not warning for the patient in early stages so it remains asymptomatic and diagnosis of the retroperitoneal mass is delayed as like our patient. It was reported that abdominal pain (67%) and an abdominal mass (42.7%) are two main symptoms. Vomiting, weight loss, lower limb edema, Budd-Chiari’s syndrome have been reported.[3]

If IVC tumor grows as a large extraluminal extension, it is a diagnostic dilemma for its origin. Hemant et al.[4] verified CT and MRI as sensitive tools for establishing diagnosis. The United States is useful to obtain tissue samples for histological evaluation. We could not determine exactly the origin of the tumor preoperatively because of its major extension out of lumen trough to surrounding connective tissue.

In 1997, Mingoli et al.[5] recommended caval wall radical resection 1 cm around the macroscopic evidence of the tumor. Once complete resection is accomplished second point is to provide continuity of the vein. The IVC can be reconstructed by auto-graft or synthetic graft like Dacron or PTFE.[5] In our patient, another major problem is the reconstruction of renal veins. It is reported that right kidney should be removed en bloc with tumor when venectomy is necessary, but it can be preserved by pelvic auto-transplantation, or anastomosis of the renal vein to the graft.[3] Stuart and Beaker[6] reported successful anastomosis of right renal vein to caudal segment of transacted IVC. We preferred to anastomize the right renal vein to graft. On the other hand, adequate venous return from the left kidney can be obtained from collaterals. Implantation of the left renal vein to the distal or proximal stump, or to the graft is other options. Following resection, most common intraoperative complication is hemorrhage. Postoperative morbidity is reported as 15% and the most common one is phlebitis.[5]

Mean survival after palliative resection was reported as 19 months. The 5 and 10 years survival rates for patients undergoing curative surgery were 49.4 and 29.5%, respectively, while disease free interval was 31.4 and 7.4%, respectively.[8] Our patients have relatively long overall survival owing to radical total resection of a low-grade sarcoma without lymphatic metastasis.

The effect of radiation therapy is controversial, similar to chemotherapy. Evidence of efficacy exists, but the indications and the best method of using radiochemotherapy remain unclear.[8][4] We believe the LMS of the IVC is a rare malignancy for which complete surgical resection with free margins plays a central therapeutic role.

REFERENCES

