SHORT REPORT

Recurrent Asymptomatic Retrohepatic Leiomyosarcoma of the Inferior Vena Cava

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A patient operated on for a retrohepatic leiomyosarcoma of the inferior vena cava had an asymptomatic recurrence 7 years after the original operation. He underwent a radical resection of the recurrent tumor with prosthetic reconstruction of the vena cava. The patient is alive and free of recurrent disease 3 years after the second procedure.

Local recurrence after radical resection of leiomyosarcoma of the IVC is common. These tumors grow slowly and a recurrence occurs even several years after a primary resection. Radical resection is possible after a recurrence with good chance of cure. Follow up of these patients with serial CT scans should be continued indefinitely.

Keywords: Leiomyosarcoma; Retrohepatic vena cava; Recurrent vascular tumor.

Case Report

A 68-year-old patient was admitted with right upper quadrant pain. Diagnostic work-up demonstrated a large mass involving the suprarenal inferior vena cava (IVC). Through a right subcostal incision the mass was resected with a tangential suture of the IVC. The pathology report demonstrated a leiomyosarcoma. Surgical margins were free of tumour. Five years later a CT scan was negative for recurrence. Two years after, the patient, still asymptomatic, underwent a CT scan that demonstrated a 3 cm recurrence (Fig. 1). An inferior vena cavogram demonstrated a partial occlusion of the retrohepatic IVC with limited tumor protrusion into the lumen (Fig. 2). The patient was operated on through the same incision. The duodenum, the right lobe of the liver and the first hepatic segment were mobilized. The vena cava was isolated and a recurrent sarcoma of the retrohepatic vena cava was removed en bloc with the right kidney and pericaval lymphnodes. The right renal vein was involved with tumor up to the hilum. Vascular reconstruction was performed with an 18-mm PTFE graft and the left renal vein was reimplanted on to the graft, but was unaffected by tumor. Total clamp time was 70 min. The postoperative course was uneventful. Frozen sections demonstrated tumour free margins and the pathology report confirmed recurrent leiomyosarcoma. Twenty-eight months after this procedure a CT scan demonstrated no recurrence with patency of the graft (Fig. 3).

Discussion

Primary venous leiomyosarcoma (PVL) is a rare disease. The vena cava is the structure most often involved, in 60% of cases. After the autopsy report of Perl in 1871, more than 300 cases have been described, either as case reports or small series. PVL of the IVC strikes mainly adults (mean age 54 years) with a predominance of females (82%). It grows slowly and often becomes fairly large before giving symptoms. The tumor spreads locally to liver, pancreas and to the ‘porta hepatis’. Hematogenous and lymphatic spread is less common and it is reported in 9% of resected tumors and in 32.5% of non-resectable disease. Distant metastasis usually occur to liver, lungs and lymphnodes.

PVL of the IVC may involve any segment of the IVC: the lower third, from the origin of the IVC to
the renal veins; the middle third, from the renal veins to the hepatic veins, and the upper third from the hepatic veins to the right atrium. The middle segment is most often involved (30–100%). Extraluminal growth is more common than intraluminal, with thrombosis of the IVC occurring in 33–41% of cases.3,4 Clinical presentation is variable and depends on the segment involved. Tumors involving the middle third of the IVC most commonly present with epigastric pain, right upper quadrant and back pain (50%). In 10% of patients, the tumor is asymptomatic and diagnosis is incidental.4

The diagnosis is usually made by CT scan or MRI, while cavography is useful in tumors with intraluminal growth because it allows transluminal biopsy and helps in planning venous reconstruction after resection.1,6,7 Complete surgical resection is the only treatment option and is feasible in 57–84% of published series.5,8 Resection of the venous leiomyosarcoma of the middle and lower third may be performed with or without venous reconstruction. A literature review revealed that prosthetic repair of the IVC was performed in 40 cases, with ringed PTFE used in the vast majority (39 cases). Graft patency is 90% in the best series with a mean follow-up of 2.8 years.9 In a recent report reconstruction of the juxta-renal vena cava for PVL was successfully achieved using a ringed PTFE and concomitant left renal vein reimplantation.10 In our case the segment involved by the tumor was the retrohepatic portion of the IVC and prosthetic reconstruction was performed on reintervention after recurrence of the tumor. This is the first case reported where a prosthesis was used in reconstructing the IVC after resection for recurrent retrohepatic disease.

A ringed prosthesis is commonly used in retrohepatic IVC reconstruction since it is thought that compression by the liver in the supine position may endanger prosthetic patency. We have used a non-ringed PTFE prosthesis because we thought that its compliance, more similar to the native IVC, would allow a greater increase in flow velocity with respiratory movements.

Reimplantation of the left renal vein is usually not necessary, but it is advisable in case of concomitant right nephrectomy.
In conclusion, even in the absence of symptoms, follow up of patients operated for PVS of the IVC should continue indefinitely because these tumors grow very slowly and recurrence is possible even after several years. Recurrent tumor may be operated on with good long-term results. Non-ringed prosthesis may be used as an IVC substitute with mid term patency.

References


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