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Asymptomatic Thrombosis as a Late Complication of a Retrohepatic Vena Caval Graft Performed for Primary Leiomyosarcoma of the Inferior Vena Cava: Report of a Case

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Abstract
A 63-year-old woman successfully underwent a graft replacement of the retrohepatic inferior vena cava with a ringed polytetrafluoroethylene graft for primary leiomyosarcoma of the inferior vena cava (IVC). Although anticoagulant had been administered, a thrombus was found in the IVC just cranial of the downstream anastomosis 67 months after the operation. The patient remained free of symptoms, and she had no evidence of any tumor recurrence. She underwent a complete resection with a prosthetic reconstruction for leiomyosarcoma of the IVC and has since been able to enjoy a reasonably long-term survival. The occurrence of thrombosis must be kept in mind in the long-term follow-up of such cases.

Key words Thrombosis · Vena caval graft · Leiomyosarcoma · Inferior vena cava · Complication

Introduction

Leiomyosarcoma of the inferior vena cava (IVC) is a rare but well-recognized clinical entity for which the optimal treatment is considered to be a complete surgical resection.[1–6] Although case reports of graft replacement of the IVC for primary leiomyosarcoma have been published, few reports have focused on the long-term outcome. We herein report a patient with leiomyosarcoma of the retrohepatic IVC who was treated successfully by both a resection and graft replacement, but subsequently developed thrombosis of the IVC as a late complication.

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Case Report

A 63-year-old woman underwent upper abdominal computed tomography (CT) for an investigation of right-sided back pain and was found to have a mass in the retrohepatic portion of the IVC (Fig. 1). Vena cavography showed both the stenosis of this portion of the IVC and the collateral pathway through the inferior right hepatic vein to the right hepatic vein (Fig. 2a). Arteriography showed the mass to be hypervascular, and it was supplied by the right inferior phrenic artery.

An operation was performed on January 10, 1996. The liver was completely mobilized and separated from the tumor together with the IVC, and all of the short hepatic veins including the inferior right hepatic vein were divided. The confluence of the main hepatic veins with the IVC was free of the tumor on the cranial side. The confluence of the IVC with the right renal vein was intact but its confluence with the left renal vein was involved with the caudal extent of the tumor. The infrahepatic portion of the IVC was divided between the left and right renal veins. The tumor was completely removed together with the IVC and the right adrenal gland. Under a venovenous bypass using a centrifugal pump, the retrohepatic potion of the IVC was replaced, an end-to-end anastomosis was performed with a 16-mm ringed polytetrafluoroethylene (PTFE) graft measuring 10 cm in length, which was selected according to the diameter of the IVC and the length of the resected IVC. The left renal vein was reconstructed using end-to-side anastomosis to the lower one-third of the graft (Fig. 3). The bypass flow rate was maintained at about 1000 ml/min, and the total bypass time was 95 min. Anticoagulant therapy was not administered during the operation. The length of operation was 9 h 25 min, and the total blood loss was 2400 g. The whitish tumor measuring $7 \times 5 \times 6$ cm was observed to originate from the wall of the IVC. A diagnosis of leiomyosarcoma was established histologically.

The postoperative course was uneventful. Neither adjuvant radiation nor chemotherapy was administered either pre- or postoperatively. The patient has been followed in our outpatient clinic with vena cavography, magnetic resonance imaging, ultrasonography, or CT every 6–12 months (Figs. 2b, 4). Warfarin potassium 3–4 mg/day and ticlopidine hydrochloride 300 mg/day have been administered to control the thrombo test to less than 20% (normal 70%–140%) or the international normalized ratio of prothrombin time to more than 1.5 (normal 1.0–1.2) to prevent thrombosis of the graft. Vena cavography showed the prosthetic graft to be patent without any stenosis of the anastomosis (Fig. 2b). Doppler ultrasonography in the infrahepatic portion of the IVC revealed a continuous waveform without a swirl wave 55 months following surgery, but the blood flow velocity was not measured. Although the patient has been free from symptoms for 69 months, CT demonstrated a thrombus in the IVC just cranial to the downstream anastomosis on July 23, 2001 (Fig. 4a). The patient's performance status has remained good despite

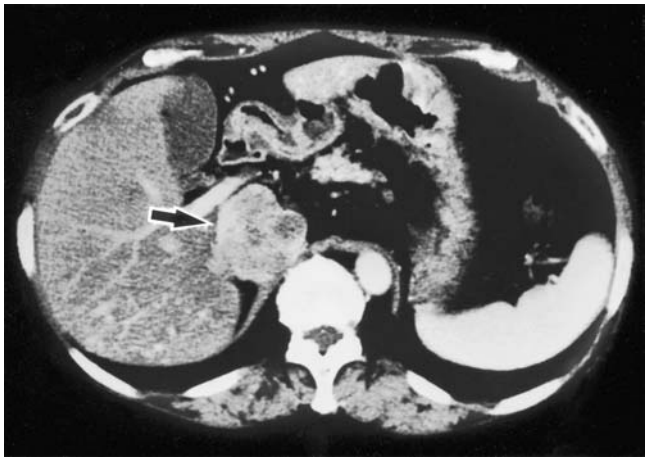


Fig. 1. Computed tomography (CT) shows a mass compressing the inferior vena cava (IVC) (*arrow*) to the right ventral side in the retrohepatic portion of the IVC

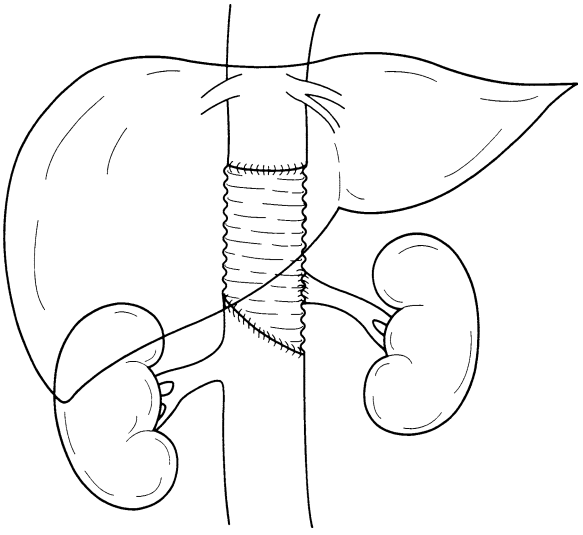
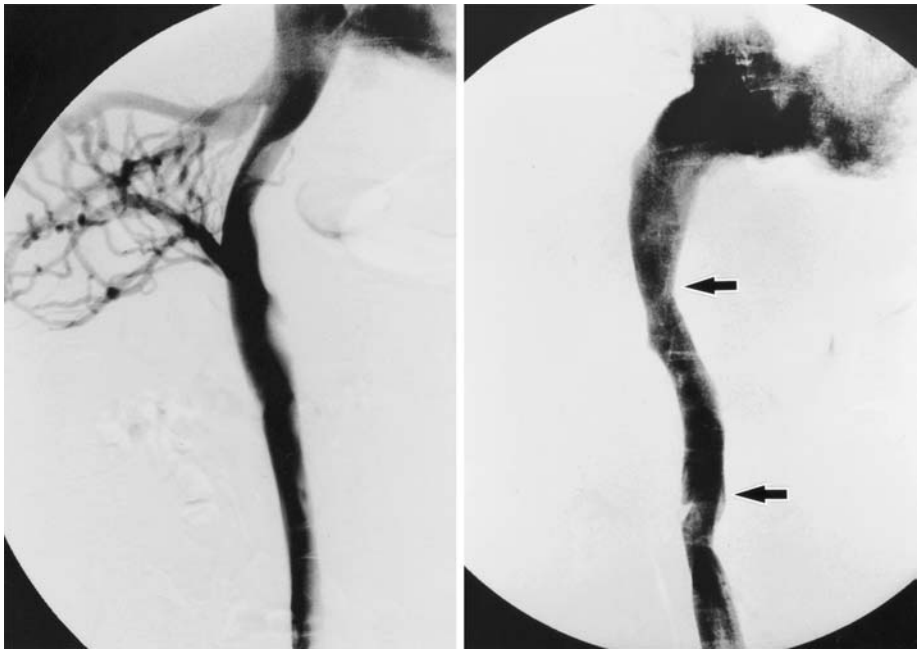


Fig. 3. The retrohepatic portion of the IVC was replaced with a 16-mm ringed polytetrafluoroethylene (PTFE) graft after a complete resection of leiomyosarcoma of the IVC. The left renal vein was then reconstructed



a **b**

Fig. 2a,b. Vena cavography. **a** Stenosis in the IVC and collateral pathway through the inferior right hepatic vein to the right hepatic vein on admission. **b** Patency of a prosthetic graft without stenosis of anastomoses (*arrows*) 5 months after surgery

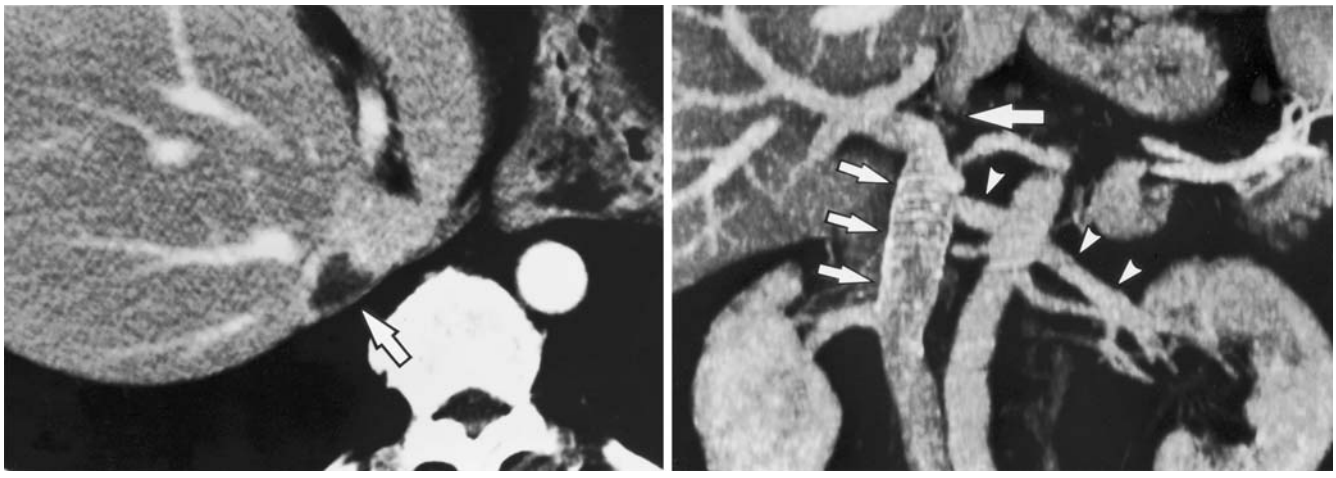


Fig. 4a,b. At 67 months follow-up, **a** enhanced CT shows the defect filled with thrombus just cranial of the downstream anastomosis of the IVC (*arrow*). **b** Three-dimensional CT shows both the ringed PTFE graft (*small arrows*) and the left renal vein (*arrowheads*) to be patent, and neither a recurrence of the tumor nor any collateral pathways are seen. The *large arrow* indicates the area of thrombosis

the development of this thrombus, and she has had no evidence of tumor recurrence (Fig. 4b).

Discussion

Primary tumors of the IVC may extend locally without distant metastasis.[5] The treatment of choice for primary leiomyosarcoma of the IVC is a complete resection of the tumor and surrounding tissue, including the IVC, both for long-term cure and palliation.[3–6] However, even a 5-year disease-free interval cannot be considered a cure, since recurrence still can occur after that time.[1,6]

Four methods for reconstructing the IVC following either a complete or partial resection have been reported: primary closure, patch repair, autogenous vein graft, and prosthetic graft.[1,2,4,7–9] The risk of death is higher for primary leiomyosarcoma of the proximal segment than of the distal segment of the IVC.[5] Hardwigsen et al.[9] reported that a prosthetic replacement of the IVC may be required more often following a suprarenal resection than after an infrarenal resection, and symptoms of IVC obstruction developed in three of six patients who underwent a resection of the IVC and primary closure without graft replacement. Since an IVC obstruction develops gradually, the opportunity exists for collateral channels to develop around the tumor. This usually permits the venous return to be maintained intraoperatively during IVC occulusion without the use of a bypass. A pump-driven venovenous bypass was used in this patient because we had to divide the inferior right hepatic vein, which served as one of the primary collateral pathways. We reconstructed the left renal vein using end-to-side anastomosis to the prosthetic graft, even thought it is generally believed that the left renal vein has an adequate collateral circulation to perform a ligation.[10] The graft replacement of the IVC may be performed safely with a low graft-related complication and good patency in selected patients, and PTFE has also been reported to provide good long-term patency rates.[1,8,9,11,12] However, late graft occlusion has been reported with and without tumor recurrence.[8,9]

The use of anticoagulant therapy following an IVC reconstruction remains controversial.[2,9,12] The local boundary layer infusion of heparin increased overall IVC graft patency, and markedly reduced downstream anastomotic neointimal hyperplasia and cell proliferation, in an experimental model.[12] This may represent an attractive strategy for anticoagulation in venous prostheses. Although good long-term patency has been reported for ringed PTFE grafts without anticoagulant,[2] thrombosis of the IVC developed just cranial of the downstream anastomosis in spite of anticoagulant therapy in this patient.

Patients who undergo a complete resection with a prosthetic reconstruction for primary leiomyosarcoma of the IVC can therefore obtain a reasonably long-term survival; however, thrombosis may occur as a late complication in an inferior vena caval graft.

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