Primary leiomyosarcoma of the Inferior Vena Cava and Interposition of a Bovine Pericardial Graft

R. Karmeli*, A. Eyal, S. Eldar and S. Fajer

Department of Vascular Surgery, Carmel Medical Center, Haifa, Israel

Primary leiomyosarcoma of the inferior vena cava (IVC) is a rare disease with fewer than 220 cases reported in the literature. Symptoms usually develop late in the course of the disease delaying the diagnosis and worsening the prognosis. Curative treatment consists of total surgical resection of the tumour with interposition grafting for the missing portion of the IVC. The reported 5 year survival rate ranges from 37 to 53%.

We report a case of a 58-year-old female with primary leiomyosarcoma of the juxtarenal IVC diagnosed incidentally on abdominal ultrasound. After further investigation by computerised tomography angiography (CTA), the tumour was removed completely with clear surgical margins. Bovine pericardium was used as an interposition graft for the resected portion of IVC. Some advantages of using this material for venous bypass include its versatile diameter options, malleability, minimal needle hole ooze and less potential for infection in comparison to synthetic grafts [Vasc. Endovasc. Surg. 37 (2003) 225]. Further research is needed regarding the patency rate, late infection and overall success.

We believe a high index of suspicion is crucial for the early diagnosis of this disease. An aggressive surgical approach is necessary for optimal survival.

Key Words: Leiomyoma of inferior vena cava; Bovine pericardial patch.

Introduction

Tumours of the inferior vena cava (IVC) can be primary or secondary in origin. Primary tumours arise from the vessel wall, while secondary ones originate in adjacent tissue and invade the IVC by direct spread.1,2

Primary leiomyosarcoma of the IVC is a rare finding with fewer than 220 cases reported in the literature.3 The first case was reported in 1871, identified on a post-mortem examination. A late diagnosis with metastases complicates the surgical treatment and leads to a relatively poor prognosis. Several recent cases have been published reporting the diagnosis and treatment of leiomyosarcomata of the IVC.3,4,5 The recommended treatment includes complete surgical resection with the role of adjuvant therapy remaining unclear. The 5 and 10 year survival rates of these tumours are 38 and 14%, respectively.3,4

Case Report

A 58-year-old female with a previous history of hypertension and hypercholesterolaemia presented with nonspecific right sided abdominal pain, a single episode of 38°C fever, fatigue and weight loss. Physical examination was unremarkable apart from mild right-sided abdominal tenderness. No abdominal mass could be felt and there was no sign of lower limb oedema or venous engorgement. An abdominal ultrasound ruled out biliary pathology but identified an abnormal juxtarenal mass in the IVC. Abdominal CTA revealed a central intraluminal mass arising from the dorsal aspect of the IVC, with dilation of the lumen, and possible involvement of the right kidney and the right adrenal gland (Fig. 1). Further investigation included a chest X-ray, echocardiogram and chest CTA, which ruled out cardiac or pulmonary artery involvement and metastatic disease. The patient was started on IV heparin to prevent vena cava thrombosis, and was scheduled for surgery.

The tumour was successfully removed with the involved portion of the IVC and the pre-vertebral
fascia, sparing both renal veins (Figs. 2 and 3). A bovine pericardial patch was used to create an interposition graft (Fig. 4).

On the first post-operative day, a decrease in haemoglobin levels was noted along with distention and rigidity of the abdomen. A CTA was performed which showed active retroperitoneal hemorrhage. The patient was re-explored and a bleeding lumbar artery was sutured. The post-operative course was otherwise uneventful. The patient was discharged on the 10th post-operative day with anticoagulant therapy.

Pathological exam showed that the resection margins were clear. A follow-up CTA conducted twelve months post-operatively demonstrated a patent graft with no evidence of local recurrence.

Discussion

Primary leiomyosarcoma of the IVC is a rare finding, more common in females, and usually presenting in the sixth and seventh decades.\(^\text{2-4}\) Diagnosis is often delayed because the disease is relatively asymptomatic. Symptoms and signs include abdominal pain (66%), abdominal mass (48%), lower limb œdema (39%), weight loss (30.6%) and Budd–Chiari syndrome (22.2%). Additional nonspecific symptoms reported include fever, weakness, anorexia, nausea, vomiting, nocturnal sweating and dyspnœa—some attributed to metastasis.\(^\text{2}\)

Diagnosis depends on specialised imaging techniques. Doppler ultrasonography usually enables visualization of the entire vena cava and defines patency as well as the presence and extent of an intraluminal thrombus. CT and MRI identify not only the presence of a tumour but give valuable information regarding localized spread and distant metastasis. Venography is important in cases of IVC occlusion in order to demonstrate collateral flow and plan surgical strategy. Pre-operative intraluminal biopsy is useful when differentiation from a thrombus is difficult.\(^\text{2}\)

For those patients diagnosed early, the optimal treatment consists of total surgical excision with prosthetic interposition grafting.\(^\text{6}\) Reconstruction of
the resected portion diminishes symptoms of lower limb venous engorgement following surgery. The response to chemotherapy and radiotherapy is variable, although some reports show benefit from radiotherapy for control of local disease. We prefer anticoagulant therapy following IVC resection, although the issue is controversial.

Resectability of the tumour is the primary factor affecting the prognosis. Late diagnosis and/or involvement of the suprarenal vena cava both complicate the surgical approach and worsen the prognosis. The reported 5-year survival of patients with primary leiomyosarcoma of the IVC is 53%, similar to that of retroperitoneal leiomyosarcomata.

In our case, early diagnosis and sole involvement of the juxtarenal IVC allowed total resection. A bovine pericardial patch was used as an interposition graft in the abdominal IVC.

A literature search revealed one other case where bovine pericardium was used as an interposition graft of the abdominal IVC. The pericardial patch has also been used in other cases but not as interposition grafts.

Our use of the pericardial patch as an interposition graft seemed more appropriate than using a synthetic graft due to the versatile diameter options. Other advantages include its friendly management and less frequent bleeding after needle puncture.

In the present instance a follow-up CTA showed patency of the graft 12 months after the operation, suggesting the adequacy of biological grafts in such a setting. We believe that a high index of suspicion for the early diagnosis of primary leiomyosarcoma of the IVC combined with an aggressive surgical approach improves the survival of these patients. The use of a bovine pericardial patch is an option in selected cases, offering some advantages over synthetic grafts.

References


Accepted 20 November 2003