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CASE REPORT

Juxta-renal Inferior Vena Cava Leiomyosarcoma

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Introduction

Leiomyosarcoma of the inferior vena cava is very rare,[1,2] with most of reports being isolated case studies or small series. This paper presents a case of a juxta-renal leiomyosarcoma of the inferior vena cava (IVC).

Case Report

A 37-year-old female with no significant past medical history presented with a 3-month history of right flank and iliac fossa pain radiating to her back. She also complained of intermittent abdominal distension, a loss of appetite and significant weight loss (10 kg) for the preceding 6 months. On examination an 8×8 cm, firm, tender mass was palpable in the right para-umbilical region. There was no peripheral oedema or any other abnormal findings. Blood chemistry was normal, but the erythrocyte sedimentation rate was raised to 80 mm/h. Abdominal ultrasound revealed a 10×10 cm mass in the lumen of the IVC at the level of the renal veins. Abdominal CT scan (Fig. 1) confirmed the presence of a well defined, solid lesion in the IVC arising at the level of the renal veins. The mass had an intra- and extramural component and demonstrated some non-homogenous contrast enhancement. A normal right adrenal gland was seen separately from the mass. An inferior vena cavagram demonstrated a near total occlusion of the infra-renal IVC with well developed collateral flow via the lumbar veins (Fig. 2). No obvious liver or lung metastases could be demonstrated on CT scan. Percutaneous biopsy was not attempted.

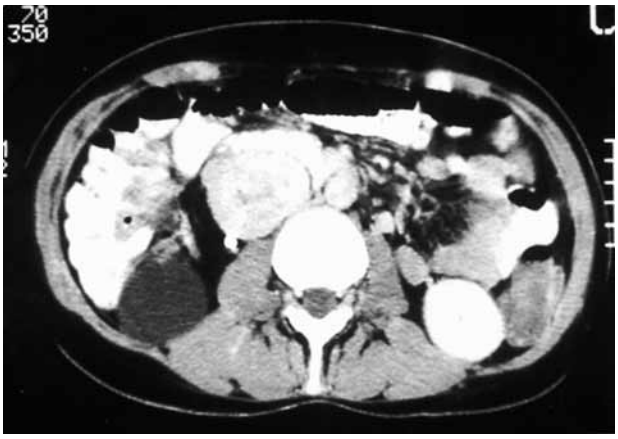

Fig. 1. Abdominal CT scan demonstrating a well defined solid lesion in the IVC rising at the level of the renal veins.

Laparotomy was performed via a long midline incision. Exploration confirmed the presence of a 10×10 cm tumour of the IVC extending from 8 cm above the iliac confluence inferiorly to the level of the renal veins. Two right renal veins were found, the more inferior being occluded by the tumour. Although the tumour had an extramural component it did not infiltrate adjacent structures and there was no sign of intra abdominal metastases. Exposure was obtained by mobilising the right colon and the duodenum by an extensive Kocher's manoeuvre. Dissection was complicated by extensive retroperitoneal lumbar venous collaterals. The right ureter was mobilised and control was achieved of the IVC 5 cm above and below the tumour. Lumbar collaterals were tied off and the lower and upper right renal veins and the left renal vein were divided with macroscopic tumour free margins. The IVC was then divided inferiorly and superiorly

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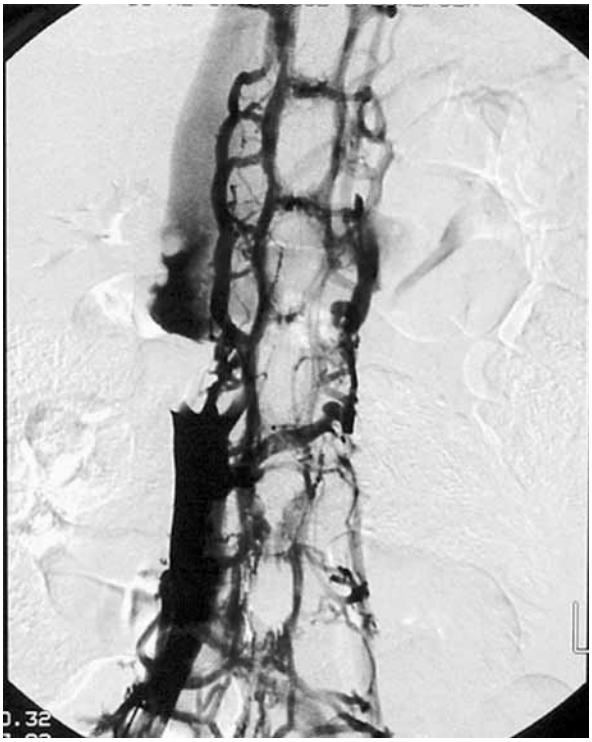

Fig. 2. Inferior vena cavagram demonstrating a near total occlusion of the infra-renal IVC with well developed collateral flow via lumbar veins.

with tumour free margins and the tumour was dissected free and removed. No macroscopic retroperitoneal infiltration was present. Reconstruction of the IVC was then achieved by a 18 mm e-PTFE interposition graft with direct re-implantation of the upper right renal vein. The left renal vein was re-implanted on the 18 mm graft via a 10 mm e-PTFE interposition graft. Postoperatively the renal function was monitored and was not affected. The patient was also treated with low dose subcutaneous heparin and graded pressure stockings to prevent venous thrombosis and promote graft patency. No complications occurred and she was discharged 6 days post-operatively. Histology confirmed the tumour to be a leiomyosarcoma (of intermediate grade) of the inferior vena cava with clear proximal and distal resection margins. At some places the tumour budded on the adventitial surface.

At 2-year follow-up the patient was well and no signs of tumour recurrence could be demonstrated on clinical examination, chest X-ray or abdominal CT scan. The scan (Fig. 3) clearly demonstrates the patent e-PTFE inferior vena cava graft with the side-graft from the left renal vein and the directly implanted right renal vein.

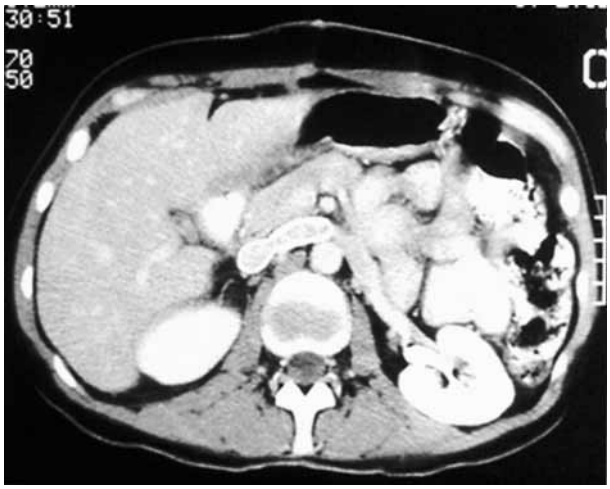

Fig. 3. Abdominal CT scan demonstrating the patent e-PTFE inferior vena cava graft with the side graft from the left renal vein and the directly implanted right renal vein.

Discussion

Perl[3] first described a leiomyosarcoma of the inferior vena cava in 1871 at a postmortem examination. The first successful surgical resection was reported in 1928.[4] Since the establishment of an international registry for these cases in 1992, only about 220 cases had been registered.[2]

Leiomyosarcomata of the inferior vena cava are usually slow growing tumours and may, like other retroperitoneal tumours, reach a large size before becoming symptomatic. This is responsible for significant problems of late presentation and delayed diagnosis and treatment, with resultant poor prognosis. The majority of patients are female (mean age at admission 54.4 ± 13.6 years) with a male:female ratio of 1:4 to 1:5.[1,5,6] Tumours can arise in the upper (above and including the hepatic veins), middle (retrohepatic to the origin of the renal veins), and lower (below the renal veins) IVC.[2] They most commonly involve the middle segment (42%) with the lower (34%) and the upper segment (24%) less frequently involved.[1] Presenting features depends on the level of involvement of the IVC and can include abdominal pain, a palpable mass, lower limb oedema, Budd Chiari syndrome and weight loss. Less frequently patients can present with abdominal distension, anorexia, nausea, vomiting, jaundice and nocturnal sweating. In 10% of patients the tumour can be asymptomatic and will only be diagnosed incidentally on routine physical examination or during imaging or laparotomy for other indications.[7] Most tumours exhibit a predominantly extraluminal growth pattern (73%), with a minority (27%) growing intraluminally.[1] The tumour is usually

slow growing, depending on the grade and differentiation, and metastasise late in the course of the disease. Haematogenous spread is mainly to the liver and lungs and lymph node metastases occur much less frequently. Metastatic disease is present in approximately 9% of resected and 33% of non-resected tumours respectively.[7] Diagnostic studies include ultrasonography, Doppler ultrasound, computerised tomography (CT scan) and nuclear magnetic resonance (NMR).[8,9] Angiography (arterial and venous) supply information on the blood supply of the tumour, its intraluminal extent and the option of transluminal biopsy.[9,10] Percutaneous needle biopsy allows histological diagnosis but does not indicate the organ of origin.[11] This also carries the risk of tumour dissemination.[12] The mainstay of therapy for these tumours remains complete surgical excision. Resection with a tumour free margin appears to be the only therapeutic modality that prolongs patient survival.[1,2,7] Reconstruction of the IVC is not always necessary as gradual occlusion of the IVC allow venous collaterals to develop. It is most often required for the upper and middle segments of the IVC to facilitate venous drainage of the liver and right kidney.[1,7,13] Reconstruction, when necessary, can be achieved by autogenous or synthetic graft replacement. The left renal vein can be safely ligated provided that collateral venous drainage via the adrenal, gonadal and lumbar veins is preserved. Renal veins, when both involved, can be reimplanted directly or via a graft limb on to the IVC-graft. The role of adjuvant therapy is unclear. It can be given preoperatively for tumour downstaging or postoperatively in an attempt to prevent recurrence and systemic metastases. Despite individual successes,[14] the data is inconclusive because of the small number of cases. On balance adjuvant therapy does not seem to influence outcome.[15,16] If complete resection was not possible debulking with radiation therapy provides good palliation.[5,14] The 5- and 10-year survival rates of IVC leiomyosarcoma are 37.8% and 14.2%, respectively, confirming the aggressive nature of these tumours.[1,2]

To summarise, leiomyosarcoma of the inferior vena cava is a rare tumour most commonly affecting females. The clinical manifestations are usually non-specific resulting in late presentation and diagnosis with advanced tumour stage. A high index of suspicion and more common use of new non-invasive diagnostic techniques will allow earlier diagnosis and hopefully a better prognosis. Complete surgical resection is the only proven therapeutic modality that prolongs survival.

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