SHORT REPORT

Endovascular Repair of Spontaneous Non-aneurysmal Aortocaval Fistula

M. S. Duxbury¹, I. P. Wells², C. Roobottom², A. Marshall³ and A. W. Lambert¹

¹Vascular Surgical Unit, Departments of ²Radiology and ³Cardiology, Derriford Hospital, Plymouth, Devon, PL6 8DH, U.K.

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Introduction

Spontaneous aortocaval fistula (ACF) is rare and usually associated with an abdominal aortic aneurysm.¹ It presents with characteristic haemodynamic disturbances,¹ and has significant morbidity and mortality.² We report the first case of an ACF due to a retroperitoneal sarcoma successfully treated by endovascular stenting.

Case Report

A 61-year-old woman with type 2 diabetes was admitted with acute dyspnoea, orthopnoea, peripheral oedema, right upper quadrant pain and nausea. She had no other specific cardiovascular risk factors or past medical history. She was tachycardic and tachypnoeic with a raised jugular venous pressure and warm peripheries. The chest was clear. There was tender, smooth hepatomegaly and no abdominal aortic aneurysm. This initial presentation was suggestive of constrictive pericarditis.

Trans-thoracic echocardiography revealed right ventricular dilatation with functional tricuspid regurgitation and a small pericardial effusion. Abdominal ultrasound demonstrated transient reverse flow in the portal vein, but was otherwise normal. No abdominal aortic aneurysm was identified. Urgent femoral angiography revealed a left to right shunt at the level of right common iliac artery to vein causing high output right heart failure: the “bursting heart syndrome”.³ Computed tomography (CT) confirmed an arterio-venous fistula at this level with loss of the fat plane between the distal aorta and inferior vena cava (IVC) but no mass. CT pulmonary angiography was normal, as was serum biochemistry and haematology.

In view of the patient’s life threatening heart failure, it was decided to place an iliac covered stent-graft which was available to us. The right femoral vessels were exposed surgically under local anaesthetic revealing pulsatile femoral veins. A covered straight stent-graft (Separate limb of an Excluder™ Endoprosthesis, WL Gore & Associates, INC., Flagstaff AZ, U.S.A.) was placed in the right common iliac artery (Fig. 1). Angiography demonstrated almost complete cessation of fistula flow and the massive backflow via arterialised pelvic veins resolved. The patient made marked symptomatic improvement with complete resolution of orthopnoea.

Ten days post stenting there was sudden clinical deterioration with worsening dyspnoea and recurrent
orthopnoea. Angiography showed partial prolapse of the proximal end of the stent-graft into the IVC, enlarging the arterio-venous fistula (Fig. 2). A bifurcated stent-graft (Excluder™ Bifurcated Endoprosthesis, WL Gore & Associates) was placed (Fig. 3) via bilateral femoral cut-downs without complication. This excluded the fistula completely.

A repeat CT scan revealed a filling defect in the IVC, consistent with thrombus. The patient was established on warfarin anticoagulation and discharged home. One month later the patient’s symptoms had not returned. A CT at this time demonstrated a well-placed stent but development of a soft tissue mass in the region of the distal IVC where the fat plane had been lost on the initial CT. A right hydroureter was also noted. The kidneys appeared normal. An open retroperitoneal biopsy was performed under general anaesthetic. Histological examination revealed a high-grade spindle cell pleomorphic sarcoma with a focal haemangiopericytoma pattern. Immunohistochemistry showed weak expression of desmin but no cytokeratin, S100 or smooth muscle actin. A clinical oncology opinion was sought and chemo-radiotherapy instituted.

**Discussion**

Eighty percent of primary ACFs are due to erosion of an abdominal aortic aneurysm into the IVC, which complicates 1% of such aneurysms.² Traumatic and iatrogenic³ causes account for the majority of the remainder. The clinical presentation of ACF may resemble constrictive pericarditis, although trans-thoracic echocardiography may be helpful in differentiating this condition. The first successful ACF repair was reported by Cooley in 1954.⁵ Despite advances in surgical and anaesthetic technique, surgery still

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Fig. 1. Transfemoral angiogram demonstrating prompt filling of the IVC due to a spontaneous fistula at the organ of the right common iliac artery.

Fig. 2. Plain X-ray showing prolapse of the proximal end of the iliac stent through the fistula into the IVC.

Fig. 3. Exclusion of the fistula after deployment of a bifurcated stent graft.
Endovascular stent-grafting is an attractive approach, and indeed there is one previous report of bifurcated stent-grafting of an ACF associated with an abdominal aortic aneurysm. We are not aware of a previous report of ACF associated with a retroperitoneal sarcoma.

In this case, emergency treatment was required for control of severe cardio-respiratory symptoms. Insertion of the straight stent-graft served as a temporising measure which, despite incomplete exclusion of the fistula, markedly improving the patient’s cardiovascular status until a definitive procedure could be performed with a bifurcated stent-graft. This case illustrates an unusual cause of ACF and its successful endovascular treatment.

References


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