The Effect of Acute Thrombosis of an Inflammatory Abdominal Aortic Aneurysm on Progression of Retroperitoneal Fibrosis: Case Report

K.K. Varadhan, T.A. Ojimba and I.S. Osman*

Suffolk Vascular Unit, Ipswich Hospital, Ipswich, Suffolk, UK

Keywords: Inflammatory abdominal aortic aneurysm; Thrombosis; Retroperitoneal fibrosis

Introduction

Inflammatory abdominal aortic aneurysms (IAAA) represent 3–10% of all abdominal aortic aneurysms (AAA). The pathogenesis is poorly understood. Although they are considered to present technical challenges during open repair, the indications for surgical intervention and the results of surgery are similar to those of ordinary AAAs. Thrombotic occlusion of an IAAA is said to occur in 0.7–2.8% of cases. Mortality rates of up to 60% after thrombosis have been reported, due mainly to myocardial infarction and renal failure, with high rates of limb loss. A search of MEDLINE revealed no reports of spontaneous occlusion of an IAAA. As such there is little in the literature regarding the effect of thrombotic occlusion of an IAAA on the natural history of any associated retroperitoneal fibrosis (RPF). We report such a case in which the peri-aortic inflammation continued post thrombosis and discuss the management and potential implications of this problem.

Case Report

A 63-year-old man was admitted to hospital with renal failure. Ultrasound and CT scans showed bilateral hydronephrosis and ureteric obstruction. A 4.8 cm infrarenal AAA was noted along with a 1.5 cm rind of peri-aortic fibrosis suggesting an IAAA (Fig. 1). Due to the relatively small diameter of the aneurysm he was managed conservatively with ureteric stent placement, steroids and six-monthly surveillance CT scans.

The initial six-month scan showed near complete resolution of the periaortic inflammation in response to the steroid therapy, with marked improvement in the hydronephrosis (Fig. 2). His renal function had also returned to normal. The IAAA had not increased in size and so he was left on low dose steroid treatment to be rescanned in 6 months. The ureteric stents were subsequently removed.

He presented again 3 months later with rapidly worsening bilateral intermittent claudication. A repeat CT scan showed thrombotic occlusion of the IAAA. Over the next 3 months his symptoms progressed to critical limb ischaemic and he therefore underwent axillo-bifemoral grafting. Steroid therapy was discontinued on discharge.

Over the next 2 months a gradual decline in renal function was noticed. Further CT imaging (Fig. 3) showed significant recurrence of the peri-aortic inflammation. He has required another ureteric stenting procedure and has been recommenced on steroids. No further procedure has been carried out regarding the thrombosed IAAA although he is being considered for bilateral ureterolysis. He has no ischaemic symptoms.
Discussion

It was felt that two aspects of this case merited reporting and discussion.

Firstly, thrombotic occlusion of an AAA is rare. We were unable to find any reports of acute thrombotic occlusion specifically in an IAAA after a MEDLINE search. Therefore experience in the management of this specific complication is likely to be limited.

The mechanisms leading to spontaneous occlusion of abdominal aortic aneurysms include the presence of multiple stenotic/occlusive lesions in the iliac arteries, a hypercoagulable state and sudden embolisation of a large mural thrombus.4,5 This patient had a small IAAA in the presence of occlusive iliac disease as seen on contrast CT.

Following thrombosis of an AAA, surgical intervention is advised.6 Standard in-lay graft repair is considered best but in the surgically unfit patient extra-anatomical reconstruction such as axillo-bifemoral bypass grafting is a good alternative.4,7 In our patient the decision to proceed to an extra-anatomical bypass was governed by the extent of retroperitoneal fibrosis (up to the level of the renal and superior mesenteric arteries) rather than the fitness of the patient. We therefore suggest that in the presence of extensive retroperitoneal fibrosis (up to the level of the renal and superior mesenteric arteries) rather than the fitness of the patient. We therefore suggest that in the presence of extensive retroperitoneal fibrosis (up to the level of the renal and superior mesenteric arteries) rather than the fitness of the patient.

We did not consider elective repair at the time of original presentation as we felt the peri-operative mortality risks at 4.8 cm diameter would be greater than conservative measures.8

The second notable aspect of this case is the rapid recurrence of the RPF on withdrawal of steroids despite cessation of blood flow through the IAAA due to thrombosis. We had erroneously anticipated that this event would lead to amelioration of the RPF process. This assumption was based on the popular theory for IAAA as put forward by Rose and Dent in 1981 through histological grading, which views all aneurysms as inflammatory, and IAAA as an extreme of this process.9 The theory also postulates that humoral factors can leak through diseased intima/arterial wall and provoke the peri-aortic inflammation and fibrosis.

Open repair of IAAA leads to regression of peri-aortic fibrosis in a large proportion of cases. Resolution of the RPF, however, is less certain following...
endovascular repair (EVAR). One of the differences between open AAA repair and EVAR is that the sac is almost completely cleared of thrombus in open repair while it stays in situ in EVAR.

It seems credible therefore that the continued presence of a critical mass of intra-sac thrombus provides the stimulus for persistence of RPF in our patient and possibly in those post EVAR patients in whom RPF persists despite adequate graft stenting. It remains to be seen what impact, if any, open aneurysm thrombectomy would have on the course of RPF in this patient.

References


Accepted 6 July 2004