Case report - Aortic and aneurysmal

A diagnostic odyssey and successful repair of a progressive aneurysmal expansion after complex aortic surgery and stent grafting of the descending aorta

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Received 13 April 2010; received in revised form 27 May 2010; accepted 28 May 2010

Abstract

A 48-year-old patient with Marfan syndrome presented with an ascending aortic and arch periprosthesi s hematoma (14.0 × 9.0 cm) and a descending aortic aneurysm (8.7 × 9.1 cm) developed over the last 2.5 years, after several operations on the ascending aorta and arch as well as stent-graft placement in the descending aorta. After a diagnostic odyssey, the small true aortic lumen was identified as the source for the ascending aortic periprosthesi s hematoma and descending aortic aneurysm expansion. The present case report underlines several important issues in the combined surgical and interventional management of patients requiring multiple and complex procedures on the thoracic aorta.

Keywords: Aortic surgery; Marfan syndrome; Aortic dissection

1. Introduction

A 49-year-old male patient with Marfan syndrome was referred to our clinic after routine CT angiography revealed a progressive enlargement of an ascending aortic and arch periprosthesi s hematoma (now 14.3 × 9.0 cm) and a descending aortic aneurysm (now 8.7 × 9.1 cm) (Fig. 1). The patient had undergone several operations in chronological order: aortic root conduit for Stanford Type A aortic dissection, aortic arch replacement, right coronary artery reimplantation for the treatment of a coronary aneurysm, ascending aorta and aortic arch re-replacement with the elephant trunk technique and a Cabrol shunt for the treatment of coronary aneurysm re-occurrence and finally a transabdominal descending aorta stent-graft implantation, the latter intervention taking place two years prior to referral.

The patient had also undergone various diagnostic procedures prior to referral; however, no definite source of leak could be identified. Computed tomography (CT) aortography did not reveal any definite source of arterial leak; however, imaging of the level of the aortic root was problematic due to the presence of the mechanical aortic valve which produced significant artifacts. To better examine the level of the aortic root, a conventional angiography was performed prior to referral, which also did not reveal any signs of leak at this level, nor at the level of the ascending aorta and arch. A contrast enhanced transesophageal echocardiography revealed no signs of paravalvular leak or any other definite signs of arterial leak, whereas Doppler examination revealed normal flow parameters and characteristics downstream of the aorta. The patient was referred to our clinic with the tentative diagnosis and suspicion of a leak in the area of the ascending aorta/arch, for further evaluation and eventual re-replacement of the ascending aorta and arch.

Upon referral we examined all diagnostic procedures performed in the last four years. We deemed that the slow–over three years–expansion of the hematoma of the ascending aorta was incompatible with a direct arterial leak. Such a connection would most probably lead to a rapid expansion of the hematoma, with signs of organ compression or tamponade. This hypothesis could explain the negative findings of all the above-mentioned diagnostics. We decided to perform a de novo complete CT evaluation of the aorta at our institution. The arterial phase (Fig. 1a) of the CT angiography performed revealed similar findings with the referral diagnostics, with no definite signs of any arterial leak. Upon careful examination, however, in the late venous phase (portal venous phase) of the CT aortography, a small lumen at the level of the descending aorta was identified, starting at the distal end of the stent-graft. This slit-like lumen could be identified only in the portal venous phase of the CT aortography.
(Fig. 1b), as contrast medium outside the stented segment of the descending aorta that could be followed retrogradely up to the level of the proximal descending aorta. The timing of the contrast enhancement of this lumen also indicated a retrograde perfusion of this lumen. With the patient on anticoagulation therapy due to the mechanical aortic valve a spontaneous seal was considered unlikely to occur.

We decided to intervene in the descending aorta where the slit-form lumen was identified, which we suspected to be the source of the progressive enlargement of the descending aortic aneurysm and ascending aortic periprosthetic hematoma. After a left lateral thoracotomy, femoral artery cannulation and low flow hypothermic perfusion state, the aneurysm was longitudinally opened and the thrombus was removed. The two prostheses were identified, without pathological findings. No leakage from the stent grafts could be identified. A small bleeding source right beside the stent grafts was identified. Preparation of this area revealed the small lumen which correlated to the CT findings. The longitudinal opening of this lumen showed that this was the true lumen of the chronic descending aortic dissection, a fact that was verified by the identification of two ostia of the intercostal arteries. This led to the conclusion that both stent grafts, as well as the elephant trunk, had been placed in the false lumen. Without signs of peripheral malperfusion, and with the stents intact we decided to avoid removing the stent-grafts and replacing the descending aorta. The small true lumen was surgically sealed with two U-stitches armed with Teflon pledgets. The patient’s postoperative course was uneventful and was discharged 19 days after the procedure. No event of visceral or spinal cord ischemia was observed at any time. Routine CT angiography nine months thereafter revealed a great regression of the descending aorta aneurysm and no progression of the ascending aortic periprosthetic hematoma up to 15 months after the procedure (Fig. 1).

2. Discussion

Surgical interventions in the aorta can be challenging especially in patients with Marfan syndrome who may require multiple and often complicated interventions. The presence of non-compliant graft material in the compliant aorta has been shown to produce increased tension in the residual aorta [1], which may lead to the formation of new aortic aneurysms further downstream in the aorta [2]. In the present case, during the second aortic arch reoperation (the fourth consecutive open surgery) the elephant trunk was placed in the much larger false lumen, a fact which also led to the positioning of the descending aorta stent-grafts in the false lumen. At the distal anastomosis of the aortic arch graft, the elephant trunk was attached to the dissection membrane only (Fig. 2). Thus, either the true lumen was not completely sealed in the first place or late dehiscence of the suture line facilitated this communication. Either way this allowed for retrograde flow from the distal descending aorta to the aortic arch, causing the expansion of the descending aorta aneurysm and of the ascending aortic periprosthetic hematoma (Fig. 2), beginning immediately after the elephant trunk procedure (Fig. 1). Sealing the true lumen led to a regression in the diameter of the descending aorta aneurysm and stopped the expansion of the ascending aortic periprosthetic hematoma, the dimension of which remained stable up to 18 months postoperatively (Fig. 1).

This case report points out several important issues in the combined surgical and interventional management of complex cases. First, the crucial role of the elephant trunk graft placement in the true lumen, particularly in the setting of chronic dissection, in which the true lumen can be much smaller than the false lumen. Second, the importance of distal sealing after stent-graft placement in the descending aorta (especially in patients with connective tissue disorders, such as Marfan syndrome), in order to avoid blood flow outside the stent-graft prosthesis. In patients with Marfan syndrome, conventional surgery may provide more durable results. Third, a complete surgical
sealing of the distal arch anastomosis in the setting of chronic aortic dissection with a large false lumen is a requirement for a successful subsequent interventional stent-graft placement. Finally, the concept of surgical fenestration during placement of an elephant trunk should be taken into consideration for later stent-graft reconstruction of the distal thoracic aorta.

References
