Case report

Presentation of a pseudoaneurysm as a supravalvular aortic stenosis 20 years after aortic root reconstruction

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Abstract

A 36-year-old patient was referred because of fatigue and decreased exercise tolerance 20 years after separate aortic valve replacement and aortic root reconstruction. The presence of a loud systolic ejection murmur and persistent left ventricular hypertrophy led to the diagnosis of severe supravalvular aortic flow obstruction by indirect compression of a large pseudoaneurysm. © 1998 Elsevier Science B.V. All rights reserved.

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1. Introduction

Resection of a part of the ascending aorta and replacement by a synthetic graft has been common practice since the early 1970s. Late complications include progressive dilatation of the graft, acute occlusion of the aorta or true or false aneurysms at the anastomotic site [1,2]. A severe supravalvular flow obstruction by the indirect compression of a pseudoaneurysm however, has seldom been reported.

2. Case-report

A 36-year-old man was referred to our out-patient department because of progressive decrease in exercise tolerance and fatigue. At the age of 16, he had undergone aortic valve replacement because of severe aortic stenosis together with resection of a grossly dilated part of the ascending aorta and non-coronary sinus vasa. A Bjork–Shiley prosthetic valve was implanted. A few centimetres above the origin of the coronary arteries up to the beginning of the aortic arch a 20 mm ASCII woven Dacron graft was used to replace the resected aneurysm. An extension of the same Dacron graft was used to close the resected aneurysmatic sinus valesalvae and this extension was sutured to the valve prothesis. The patient’s native proximal aorta was closed around the synthetic graft. The postoperative recovery and follow-up were uneventful.

The patient was free of symptoms for 20 years and worked full-time as a carpenter. In May 1996, he visited the out-patient department earlier than scheduled because of fatigue and decreased exercise tolerance. Auscultation of the heart revealed a loud systolic murmur radiating to the carotid arteries and back of the chest. No diastolic murmur was heard. The opening and closure clicks of the prosthetic valve appeared normal. The electrocardiogram showed unchanged left bundle branch block. The chest X-ray showed a slightly enlarged heart shadow with intrapulmonary edema. Transthoracic echocardiography showed severe left ventricular hypertrophy, a good systolic function of the
left ventricle and normal opening of the valve prosthesis. Doppler findings of the aortic valve prosthesis were within normal limits. However, placing the pencil Doppler probe in the right supraclavicular position, a systolic flow was measured in the proximal aorta of 6 m/s, reflecting a pressure gradient of 150 mmHg. Suprasternal echocardiography revealed a ‘coarctation’-like stenosis of the proximal aorta with a highly turbulent flow using colour Doppler imaging. A magnetic resonance imaging (MRI) scan of the thoracic aorta was performed and it was shown that the stenosis was located at the proximal anastomosis site (Fig. 1). Flow measurements calculated a pressure gradient of 100 mmHg. This was confirmed at catheterization and angiography, where a pressure drop was recorded of 105 mmHg by pulling the catheter back through the stenotic site.

2.1. Surgical findings

A false aneurysm was found originating from the proximal anastomotic site near the closed sinus valsalvae aneurysm. Outward expansion of this pseudoaneurysm was prevented by the ‘sheath’ of native aortic tissue wrapped around the synthetic graft during the operation in 1975. Filling of the false aneurysm therefore led to compression of the graft leading to severe flow obstruction (Fig. 2).

3. Discussion

Aortic valve replacement at a young age because of congenital aortic valve disease is performed with good long term results, although life expectancy in these patients is restricted [3]. However, long term complications of aortic root replacement by a synthetic Dacron graft are regularly seen. The formation of a pseudoaneurysms occurs in 5–15% of patients within a period of 10 years [2,4]. However, most of these pseudoaneurysms expand to the outer periaortic space. This case is unique because the pseudoaneurysm led to inward compression of the proximal vascular graft. Surgery finally led to the dissection-like mechanism of the obstruction: each cardiac cycle led to filling of the pseudoaneurysm (false lumen), resulting in compression of the aorta (true lumen). Further increase in cardiac output led to a more severe obstruction, starting a vicious circle, which led to progressive left ventricular hypertrophy, instead of the expected regression of hypertrophy.

In our patient, the persistence of left ventricular hypertrophy and the increasing intensity of the systolic murmur on auscultation were reasons to perform detailed echocardiography, including Doppler flow measurements of the proximal aorta. This finally revealed the pressure gradient and led to further diagnostic procedures. Review of patient data showed that, during
the yearly visits at the outpatient clinic, an ejection murmur was present from the moment he was operated. This complication might have been prevented when a composite valve graft prothesis was used. A recent study concluded that especially in young patients and in patients with dilated coronary sinuses a composite graft is recommended instead of a separate valve and graft. Furthermore, careful and regular imaging of patients with synthetic grafts of the aorta is recommended and especially Doppler echocardiography and MRI has shown to be of value to detect early and late complications [5].

4. Conclusion

This case describes a seldom reported complication 20 years after separate valve and aortic root replacement. A large pseudoaneurysm originating from the proximal anastomosis caused severe obstruction of aortic flow by indirect inward compression of the aorta. This complication might have been prevented by using a composite valve graft and might have been detected earlier when regular imaging of the proximal aorta was performed.

References