CASE REPORT

Large aortic pseudoaneurysm after Bentall procedure in a patient with Marfan's syndrome

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ABSTRACT

An 11-year-old male with Marfan's syndrome and aortic root dilatation underwent an uneventful Bentall procedure to replace his aortic root and valve. Five months later, surveillance echocardiogram revealed a slowly enlarging pseudoaneurysm arising from the ascending aorta. This finding was subsequently confirmed by computed tomographic angiogram. The patient had successful open surgical repair and paraaortic hematoma evacuation.

Keywords: Bentall procedure, pseudoaneurysm, Marfan's syndrome

CLINICAL SUMMARY

An 11-year-old male patient with Marfan's syndrome had progressive aortic root dilation by echocardiogram, measuring up to 5.3 cm on his most recent study, with mild aortic insufficiency. He underwent a Bentall procedure (aortic root and valve replacement) with a 23 mm On-X mechanical prosthesis and a 30 mm Valsalva graft. The coronaries were reimplanted. His operation and initial postoperative course were uneventful. Five months later, transesophageal echocardiogram was performed before a planned optic surgery for a separate issue. This revealed a mild-to-moderate paravalvular leak at the aortic valve prosthesis [Figure 1a and b]. The patient had no cardiothoracic symptoms at that time. Computed tomographic angiogram was requested on the same day to further evaluate the leak. This demonstrated a paraaortic hematoma measuring 5.8 cm × 5.4 cm × 3.7 cm, with a contrast jet arising from the right posterior aspect of the aortic root, approximately 3 cm above the valve plane [Figures 2b and 3b]. Per surgeon preference, and in light of the patient's clinical stability and the family's desire to attempt conservative management, the decision was made to pursue close imaging observation. Subsequent computed tomography (CT) scans demonstrated a chronic expanding pseudoaneurysm. Nine months postoperatively, the hematoma measured 10 cm × 8.5 cm × 4.7 cm with mass effect on the superior vena cava and atria [Figure 2a, 3a and c]. The pseudoaneurysm neck measured 3 mm × 4 mm in diameter. Findings were confirmed on echocardiogram, with pulsatile flow through the neck [Figure 4a and b, and Video 1]. The neck of the pseudoaneurysm was within 1 cm from the sinotubular junction and reimplanted coronary arteries. In light of the patient's known Marfan's syndrome, there was a concern for high risk of dissection at the landing zone if endovascular treatment was attempted. Due to the patient's young age, the short distance between the origin of the coronary arteries and the neck of the pseudoaneurysm, and risk of dissection, a decision was made to excise the pseudoaneurysm and not to use

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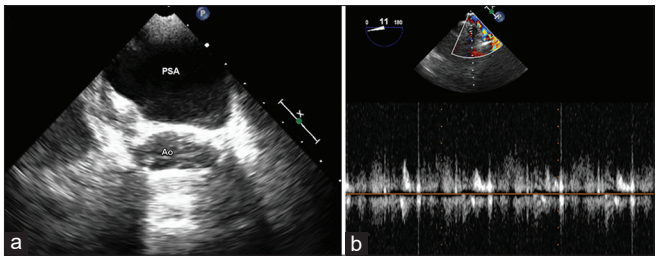

Figure 1: (a and b) Transesophageal echocardiogram performed 6 months postoperative demonstrates fluid collection anterior to the aorta (Ao) which was suspected to be a pseudoaneurysm (PSA). The pseudoaneurysm measured 4.1 cm × 3.8 cm with pulsatile flow on Doppler

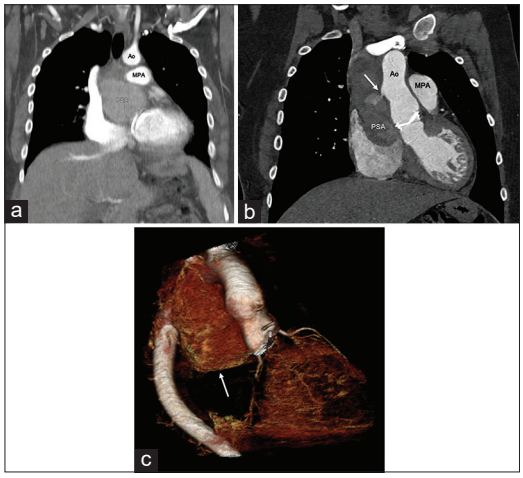

Figure 3: (a-c) Coronal contrast-enhanced computed tomography scan demonstrates the pseudoaneurysm 6 months postoperative (a) and 9 months postoperative (b) with a jet of contrast (arrow) into the pseudoaneurysm posterior to the aorta and the main pulmonary artery. Volume rendered image 9 months postoperative (c) demonstrating the pseudoaneurysm (arrow)

endovascular repair. An open surgical repair was deemed a safer option.

During surgery, the pseudoaneurysm was noted to be from a small opening toward the midportion of the ascending aorta near the distal anastomosis which was at the point where the aorta was transected during prior Bentall procedure. The inherent aortic wall abnormality related to the patient's known Marfan's syndrome rendered him more susceptible to developing pseudoaneurysm at the suture line due to the lack of strength of the native aortic tissue. The area of aortic transection and the portion of the pseudoaneurysm were therefore excised. The rightward aspect of the aortic wall was noted to be extremely thin. This area was repaired by performing an end-to-end anastomosis and buttressing the rightward aspect of the aorta with a double thickness of the wall as well as a long patch of bovine pericardium. The patient had an uncomplicated postoperative course.

Differential diagnosis

Differential diagnosis was anterior mediastinal mass, ductus diverticulum, mycotic aneurysm.

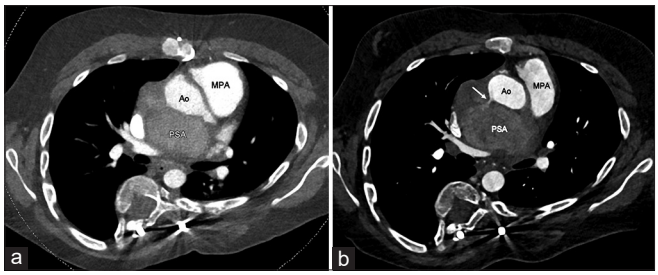

Figure 2: (a and b) Axial contrast-enhanced computed tomography scan demonstrates the pseudoaneurysm 6 months postoperative (a) and 9 months postoperative (b) with a jet of contrast (arrow) into the pseudoaneurysm posterior to the aorta and the main pulmonary artery (MPA)

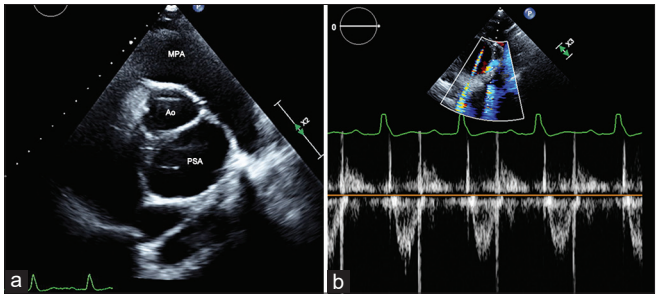

Figure 4: (a and b) Transthoracic echocardiogram performed 9 months postoperative, demonstrates fluid collection corresponding to an enlarging pseudoaneurysm posterior to the main pulmonary artery and aorta. The collection measured 4.5 cm × 4.8 cm with pulsatile flow on Doppler

DISCUSSION

A pseudoaneurysm is a focal dilation of an artery that does not contain all three of the normal arterial wall layers but is instead only contained by an outer adventitial layer or thrombus and fibrous tissue. Although often asymptomatic, symptoms of pseudoaneurysm include a pulsatile suprasternal mass, evidence of myocardial ischemia, chest pain, dyspnea, or stridor.[1] Echocardiography with color-flow Doppler and CT angiogram is extremely useful for diagnosing complications of composite aortic reconstruction, especially pseudoaneurysm formation.[2] Echocardiographically, the false aneurysm typically shows an area of flow extending a variable distance beyond the lumen of the aorta into the perigraft space. They may form as a sequela of suture line tension, graft infection, excessive use of biologic glue, or persistent bleeding into the space surrounding the aortic graft.[3] Postoperative pseudoaneurysms preferentially develop at aortotomy sites, cannulations sites in the setting of cardiopulmonary bypass, needle puncture sites, cross-clamping sites, and surgical anastomoses (such as at the location of reimplanted coronary ostia in aortic root reconstructions).[4]

The Bentall operation involves composite graft replacement of the aortic valve, aortic root, and ascending

aorta. It is commonly performed for the aortic root or ascending aortic aneurysm with aortic valve pathology, and reported complication rates are relatively low. In a recent large meta-analysis, the rate of early postoperative mortality was reported at 6% and the annual rates of the aortic root or aortic valve reoperation were 0.46% and 0.3%, respectively.[5] The development of aortic pseudoaneurysm after the Bentall procedure remains a potential complication both in the early and late follow-up periods, with incidence reported in historical publications of up to 7%–25% of graft replacements.[6] Operative techniques have since changed, but there is less data on the rate of pseudoaneurysm development in the recent era, and the exact current incidence is not entirely clear. In one case series, postoperative pseudoaneurysm was detected in 5% of patients after aortic root replacement procedures, which included the Bentall procedure as well as several other operative techniques.[7] Comparison of the advantages and disadvantages of different aortic reconstruction techniques is beyond the scope of this article, but it is important to monitor for pseudoaneurysm formation in any patient who has undergone prior aortic root surgery. More specifically, rates of in-hospital mortality and postoperative stroke after thoracic aortic pseudoaneurysm repair have been reported at 6.7% and 3.3%.[8]

Treatment options for aortic pseudoaneurysms include surgical grafts, ligation, pericardial roll graft replacement, embolization with coils, and the use of endovascular stent grafts combined with surgical treatment.[9] The common problem with endovascular repair is the presence of an inadequate short proximal and distal landing zone. In our patient, coil embolization and endovascular injection of embolic agents were not options because of the caliber of the pseudoaneurysm. It is critical for both clinicians and imagers to have a high index of suspicion for pseudoaneurysm formation after aortic root replacement because presentation can be asymptomatic (as in this case) or delayed, and clinical consequences for the patient can be severe.

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Conflicts of interest

There are no conflicts of interest.

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