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CASE REPORT

Diagnosis of a Postsurgical Aortic Pseudoaneurysm in Shone Syndrome by Cardiovascular Magnetic Resonance

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A 34-year-old male was admitted with increasing dyspnea on exertion for the past 3 years. He was born with shone syndrome. Shone syndrome was first described by Shone et al. in 1963 and consists of four obstructive, or potentially obstructive conditions of the heart.[1] These are a "parachute" deformity of the mitral valve, supravalvular ring of the left atrium, subaortic stenosis, and aortic coarctation. He had repair of a coarctation of aorta within the first year of his life. At 7, he underwent mitral and aortic valvuloplasty and at 15 had aortic valvulotomy and aortoplasty. Five years later, he underwent repair of an ascending aortic aneurysm with a Dacron graft, reimplantation of the coronary ostia by a button technique, and valve replacements with a Bjork–Shiley mitral valve prosthesis and St. Jude aortic valve prosthesis.

He was asymptomatic until age 31 when he was admitted with hemoptysis but discharged after negative bronchoscopy and V/Q scan. He continued to become progressively more dyspneic on exertion. He underwent right and left heart catheterization and transthoracic echocardiography, which showed pulmonary hypertension (Mean PA = 46 mm Hg), increased right ventricular pressure and the presence of an aortic pseudoaneurysm (PSA) extending from the proximal ascending aorta and compressing the left atrium. On angiography it was noted that the left circumflex originated from the right coronary artery but it was not possible to visualize left main or left anterior descending artery. Cardiovascular MR (CMR) was performed including MR angiography with gadolinium DTPA. This showed the previously placed tube graft in the ascending aorta connected to a large PSA

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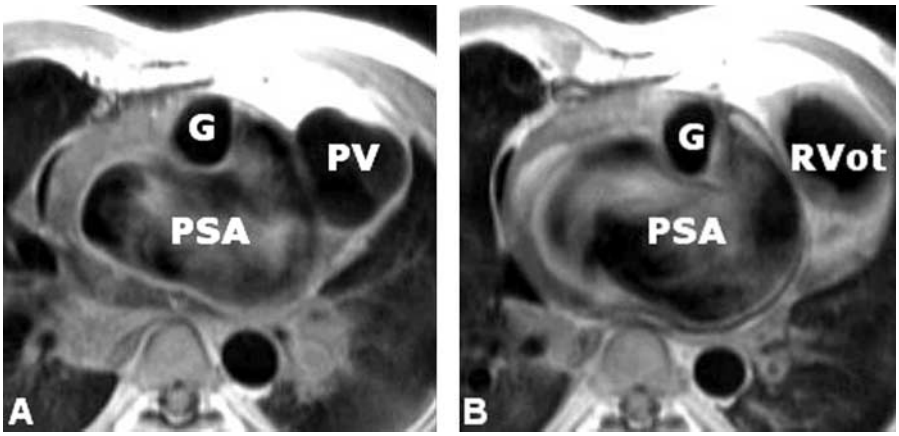

Figure 1. Axial double IR Black blood fast spin-echo images at the levels (A) of the pulmonary valve and (B) the right ventricular outflow tract (RVot). The PSA appears as a large fluid filled cavity, posterior to the ascending aortic prosthetic graft (G), the pulmonary valve (PV), and the RVot. Note the slow flow of blood producing signal within the PSA lumen.

(Fig. 1). A clear communication site was observed between the aorta and the PSA immediately above the aortic valve at the usual site of left main coronary artery (Fig. 2). However, the left main coronary artery was not visualized. Slow flow and extensive turbulence was visualized in the lumen of the PSA with concentric layered thrombus lining its wall. The PSA compressed the left and right atria and the SVC, and displaced and mildly compressed the superior right pulmonary artery, RPA (Fig. 3). The main pulmonary artery was enlarged (43 mm). Moreover, an aortic coarctation (12 mm) was seen again distal to the origin of the left subclavian artery without any associated turbulence.

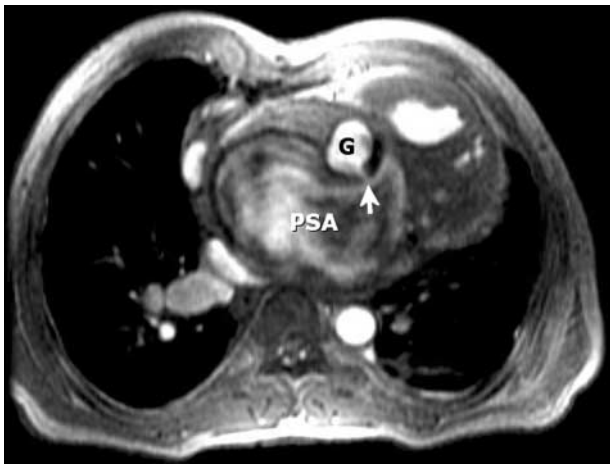

Figure 2. The axial fast gradient-echo sequence shows turbulent flow inside the lumen of the PSA. The communication between the graft (G) and the PSA is seen (arrow).

The patient underwent the repair of the ascending aortic PSA. The left coronary artery anastamosis had completely dehisced and there was a 5 mm separation between the hole (button) in the tube graft and the left coronary ostium. The left main coronary artery was then anastamosed to the ascending aortic graft using a 7 mm woven graft. The patient recovered completely and was discharged.

This case underscores the value of cardiovascular MRI in the assessment of postoperative complications of aortic repairs. The initial presentation with hemoptysis in the setting of multiple previous thoracic operations including aneurysm repair should raise concern about a possible aortopulmonary fistula. There have been many reports of hemoptysis as the initial manifestation of fistulized thoracic aortic aneurysm or PSA after an aortic repair into the bronchial tree.[2–9] Many times self-limited episodes of hemoptysis of lesser intensity precede a life-threatening massive hemoptysis. Fortunately, this patient had a diagnosis and surgical intervention prior to a catastrophic event.

The diagnostic approach is critical. This patient underwent bronchoscopy and aortography before CMR. Ramakanton et al. reported a case of false aneurysm rupture after aortographic pressure injection.[10] They concluded that in such cases, especially in PSAs, extreme caution should be exercised in performing aortography. Also, in cases of massive hemoptysis, bronchoscopy is not very useful in delineating the source of blood and caution should be exercised in performing this procedure on

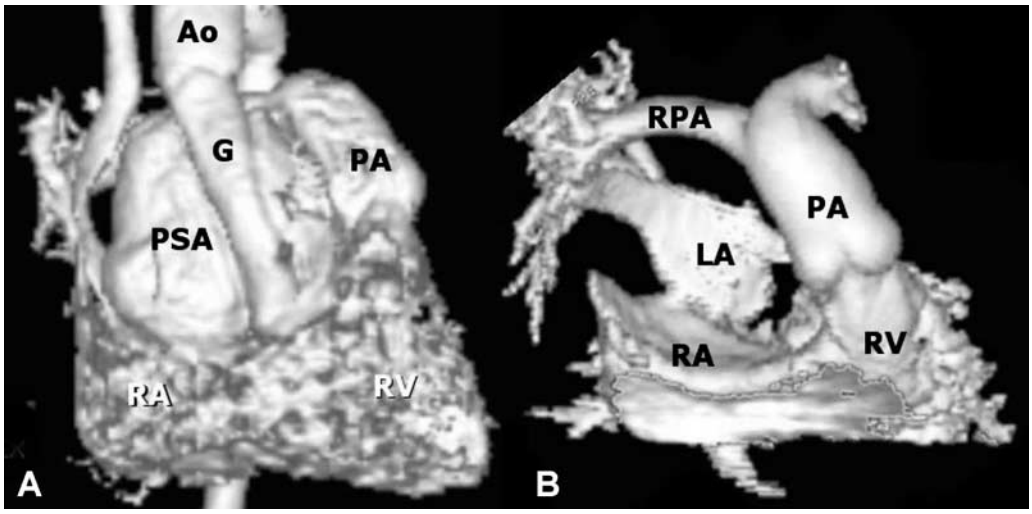

Figure 3. (A) The 3D contrast-enhanced MRA surface rendered image shows the blood filled PSA posterior to the ascending aorta prosthetic tube graft replacement (G). AO, ascending aorta; RA, right atrium; RV, right ventricle; PA, pulmonary artery. (B) In an earlier acquisition the PSA has not been opacified. Both atria are compressed and the RPA is compressed and mildly dislocated superiorly. LA, left atrium; RA, right atrium; PA, pulmonary artery; RV, right ventricle.

anyone with active hemoptysis. CMR is a noninvasive, safe, and highly accurate method to study these patients.

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