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Case Report

Aortic Fistula Plug: A Moving Target

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ABSTRACT

A middle aged male with history of Fallot type Double outlet right ventricle and coarctation of aorta who underwent multiple surgeries since childhood. With RV to pulmonary artery conduit and end to end coarctation repair initially, followed by aortic valve, pulmonary valve and RV-PA conduit replacements later. He subsequently developed significant flow through a fistula between a previously coiled aortic pseudoaneurysm and LVOT. This was percutaneously closed with a vascular plug and additionally a PFO, detected, was closed with a device.

Patient returned to the clinic with decompensated heart failure. Updated imaging demonstrated that the plug had migrated and together with the coil, which was impinging on the AVR, resulting in severe supra-valvar AS and continuous significant persistent residual flow across the Aorta- LVOT fistula via false aneurysm.

Our case highlights the importance of using multimodality imaging for complex congenital heart diseases. In view of high surgical risks associated with previous multiple sternotomies and patient's preference, percutaneous approaches were adopted initially: however, it was unfortunate to see the migration of vascular plug. After successful deployment of AVP, migration or recanalization is rare, but should not be completely discounted.

Background

A 48-year-old gentleman with history of Fallot type Double outlet right ventricle (RV) and coarctation of aorta underwent surgical repair as a child with RV to Pulmonary artery (PA) conduit and end to end coarctation repair. He had suffered recurrent episodes of endocarditis requiring aortic valve, pulmonary valve, and RV-PA conduit replacements with multiple sternotomies.

In 2016, a false aneurysm was noted in the distal aortic suture line and successfully treated with coil embolisation (Figure 1). However, subsequent transthoracic echocardiogram (TTE) revealed new significant aortic regurgitation (AR); caused by flow through a fistula, connecting his aortic pseudoaneurysm with his left ventricular outflow tract (LVOT) (Figure 2A). The fistula was closed percutaneously using an Amplatzer® Vascular Plug (AVP). Peri-procedurally (Figures 2B & 2C), a patent foramen ovale (PFO) was identified with left to right shunting and was also closed percutaneously using 35 Cribriform occlude (Figure 2D).

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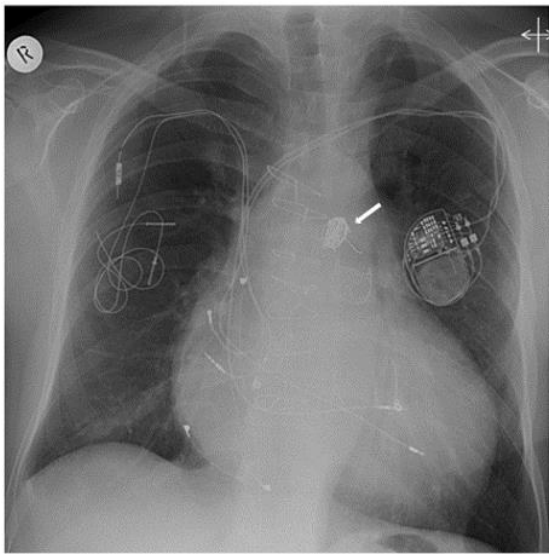

Figure 1: CXR showing cardiomegaly and coil embolisation of the false aortic aneurysm (white arrow).

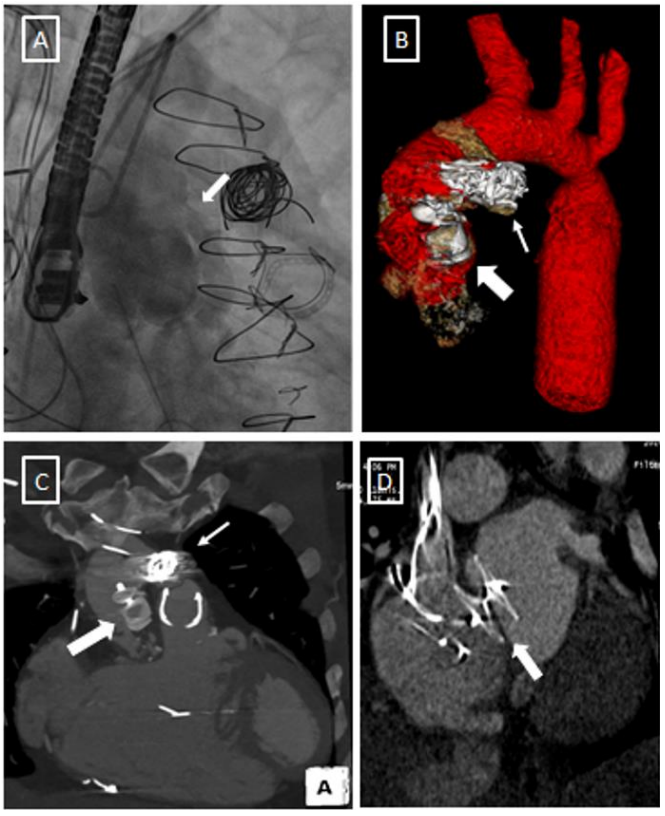

Figure 2: A) Angiogram showing the flow through the fistula connecting the aortic pseudoaneurysm and LVOT. **B)** 3D volume rendered CT showing the vascular plug (big arrow) and coil (small arrow). **C)** CT scan showing location of vascular plug (bit arrow) and coil (small arrow). **D)** CT showing location of the Amplatzer device in the intra-atrial septum (arrow).

Case Presentation

Three months later, he presented to our cardiology services with progressively worsening shortness of breath (NYHA class IV) along with orthopnoea, paroxysmal nocturnal dyspnoea and episodes of presyncope. On admission he was hemodynamically stable. Clinical examination revealed harsh ejection systolic murmur with a prominent diastolic component in the aortic region, abdominal ascites, hepatomegaly and pitting oedema up to the mid thighs. Blood tests showed significantly raised BNP 1466 ng/L (normal 0-20), mildly raised bilirubin and chronic thrombocytopenia. The rest of his bloods were unremarkable.

TTE showed moderate to severe AR (Video 1). There was holo-diastolic flow reversal in abdominal and descending aorta (Video 2). There was a moderately dilated Left ventricle (LV) with low normal Ejection Fraction (EF) 53%, and hypokinetic septal motion in the setting of significant AR. The RV was severely dilated with impaired systolic function and a severe low velocity Tricuspid regurgitation (TR) (Video 3). High continuous wave (CW) Doppler velocity was recorded at the right sternal edge with peak gradient of 86mmHg, mean gradient 49mmHg most likely at the level of coil in ascending aorta resulting in significant supra-valvar AS (Figure 3). To elucidate the mechanisms of the AR further, a TOE performed a few days later revealed a displaced vascular plug impinging onto the prosthetic aortic valve right coronary cusp causing significant obstruction (Vmax 3.7mmHg, PG 55mmHg, MG 30mmHg-under GA) (Figure 4). The AR was, in fact, a significant persistent residual flow across the Aorta- LVOT fistula via false aneurysm where the vascular plug had displaced (Video 4, Video 5, & Video 6). There was poor apposition in the PFO closure device discs with a mobile septum seen in between (Video 7). Hence a contrast bubble study was performed revealing new moderate to severe continuous shunting from right to left (Grade III) via the PFO (Video 8). This was due to increased right heart pressures secondary to impaired RV function and torrential TR. Cardiac CT demonstrated persistent contrast to suggest persistent patency of the LVOT- aorta pseudo aneurysm. (Figure 2A).

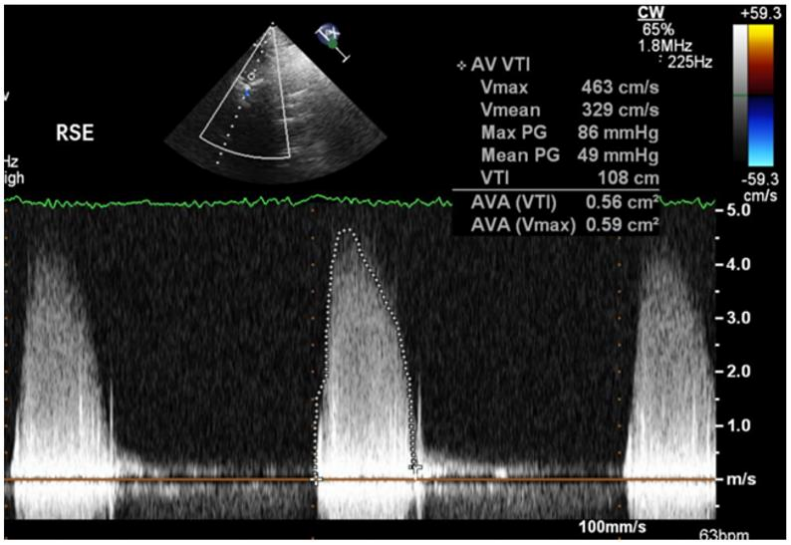

Figure 3: TTE showing severe aortic stenosis captured from the right sternal edge.

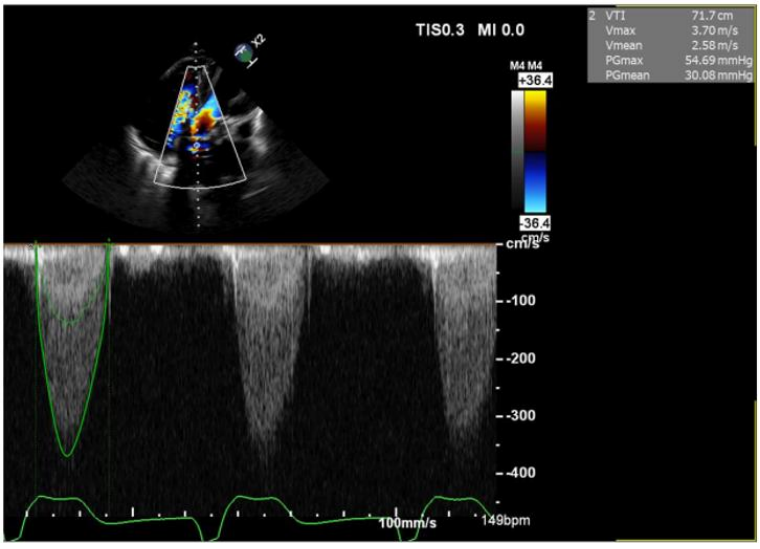

Figure 4: TOE showing significant obstruction at the aortic valve level.

Due to the extreme complexity of the case and the presentation of decompensated heart failure, a multidisciplinary approach was adopted, and a consensus was reached to add the patient on the Heart Transplantation list along with medical heart failure treatment, and patient was discharged home with a planned date for surgery.

Discussion

Due to advances in the treatment of patients with congenital heart disease, more patients are surviving into adulthood. Despite previous 'corrective' surgeries, this group of patients are more likely to require further reoperations and intervention. Although evidence is scant,

multiple sternotomies have been associated with increased early mortality in patients with congenital heart disease [1].

In order to reduce the risk of repeat sternotomy, a decision was made to undertake closure of the fistula with an AVP. Persistent patency, as in this case, is a recognised complication of the AVP. Incomplete embolization can be caused by a vessel that never occludes or a vessel that was occluded but later recanalized [2]. The AVP encourages clot formation by resisting blood flow, however, there is a recognised risk of lack of thrombus, although this is less common with newer version as the manufacturer increased the number of layers for the later versions. Vessel recanalization is a serious but rare complication, as in this case. Despite this, in studies reviewing embolisations in pulmonary arteriovenous malformations, the rates of recanalization using the AVP was less than the rate using coils [3].

In this case, the displacement of the AVP causing impingement of the prosthetic aortic valve leaflet was the most significant complication, requiring urgent sternotomy. Device migration is rare, however, in the treatment of aortic pseudoaneurysms, this appears to be a relatively common complication with an incidence around 12%. This is likely due to the flows, large necks, and very short landing zones in these cases [4].

Follow-Up

Unfortunately, multiple factors delayed admission for classical surgery, in particular intensive care staffing concerns secondary to the COVID-19 pandemic. A second opinion for heart transplantation was sought from another regional heart transplant center where he was transferred and is awaiting transplantation workup.

Conclusion

We present a complex adult congenital heart disease case who presented with decompensated heart failure secondary to multiple cardiac surgeries and aortic fistula device closure migration. Our case highlights the importance of utilizing multimodality imaging in assessing complex cardiovascular anatomy in specialist centers. AVP vascular plugs have a technical success rate of 92-100% and while device migration, embolization or recanalization is rare, they remain potential complications which need to be considered and communicated to patients ahead of planned percutaneous procedures.

Conflicts of Interest

None.

Funding

None.

Abbreviation

AVP: Amplatzer® Vascular Plug
Ao: Aorta
AR: Aortic Regurgitation
AVR: Aortic Valve Replacement
LV: Left Ventricular
LVOT: Left Ventricular Outflow Tract
PA: Pulmonary Artery
PFO: Patent Foramen Ovale
PPM: Permanent Pacemaker
PVR: Pulmonary Valve Replacement
RV: Right Ventricle
TOE: Transoesophageal Echocardiogram
TR: Tricuspid Regurgitation
TTE: Transthoracic Echocardiogram

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