Aorto-right ventricular fistulas are defects of the aortic wall in the area above the right coronary cusp, where it separates the aorta from the right ventricular outflow tract. Often, these injuries are due to trauma, infective endocarditis, rupture sinus of valsalva or occasionally following Aortic valve replacement. We describe aorto right ventricular fistula in a patient 3 months following aortic valve replacement and type A aortic dissection repair.

Keywords: Aortic dissection; Aorto-Right ventricular fistula; Aortic Valve Replacement

Abbreviations: CT: Computed Tomography; TEE: Transesophageal Echocardiogram; ARV: Aorto-Right Ventricular; RCA: Right Coronary Artery; RV: Right Ventricle; TAVI: Transfemoral Aortic Valve Implantation; RVOT: Right Ventricular Outflow Tract

Case History

A 56-year-old male patient with long standing history of essential hypertension on treatment. Sustained type A aortic dissection with severe Aortic regurgitation requiring urgent surgery. He had type A aortic dissection repair. He also received a mechanical aortic valve prosthesis three months prior to presentation to our institute. He was requiring follow up. He denied symptoms of chest pain, shortness of breath or palpitations. His medical therapy consisted of Asprin, Lisinopril, Atorvastatin, Metoprolol and Warfarin. His biochemical profile revealed a controlled LDL cholesterol. His blood pressure and heart rate were well controlled. An ejection systolic murmur and click were auscultated over the aortic area. Electrocardiogram showed sinus rhythm and left ventricular hypertrophy. A transthoracic echocardiogram showed normal biventricular size and systolic function, well-seated mechanical aortic prosthesis with acceptable hemodynamics. An abnormal systolic flow was noted by color Doppler at the right coronary sinus directed towards the right ventricular outflow tract (Figure 1 & 2). A transesophageal echocardiogram (TEE) demonstrated a well-seated aortic prosthesis with physiological trivial intra-valvar regurgitation and a small defect measuring four millimeters between the aorta and the right ventricle with abnormal systolic flow demonstrated at the right coronary sinus directed to the right ventricular outflow tract in keeping with aorto-right ventricular (ARV) fistula (Figure 3). No echocardiographic evidence of endocarditis is seen.

Cardiac computed tomography (CT) demonstrated patent left coronary arteries, a massive dissection flap extending from beyond the aortic graft to involve the descending aorta and a right coronary artery (RCA) dissection extending from the origin to the distal segment of RCA (Figure 4 & 5). The coronary dissection was not flow limiting and there was no evidence of obstructive coronary artery disease. The aorto-right ventricular fistula (ARV fistula) was detected by CT as a small defect four millimeters in size between the aorta and right ventricle (Figure 6).
Uncommon Findings Post Repair of Type A Aortic Dissection

Figure 2: Parasternal short axis view at the level of the aorta in TTE demonstrating flow to the RVOT with a measured neck of ~0.3 cm.

Figure 3: Mid esophageal long axis view in TEE demonstrate by color Doppler the shunt to the RVOT.

Figure 4: Curved MPR image demonstrates the dissection flap in RCA (arrow), ascending aorta graft (*) and the residual dissection flaps in the aortic arch and descending thoracic aorta.

Figure 5: Curved MPR image demonstrates right coronary artery dissection (arrow).

Figure 6: CT Axial image demonstrates a small defect in the aortic root and contrast jet directed from the right coronary sinus towards the right ventricle (arrow). The RCA dissection is seen in the same image (curved arrow).

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Multiple factors were considered in the management plan of our patient. The percutaneous treatment of the coronary dissection deemed unnecessary as the patient was stable without signs of ongoing ischemia. A two years follow up coronary CT angiography showed the same extent of the disease with no progression.

We think the small ARV fistula is likely to be related to the surgery as there was no evidence of infective endocarditis demonstrated. Redo Surgery for this fistula deemed unnecessary but unfortunately declined by the patient. The heart team decision was to carefully follow up the patient with consideration of percutaneous closure of the ARV fistula if there is progressive increase in the size of the right ventricle (RV) or worsening of its function. A serial follow up echocardiographic studies after 12, 24 and 36 months intervals revealed no change in the size or function of the RV. The patient continues to do well.

**Discussion**

Acute aortic dissection is not a very uncommon condition that could be catastrophic with a high mortality rate [1]. Extension of the dissection to the coronary arteries is a rare complication, but significantly increases the mortality rate. Hirst et al. reported coronary dissection as an extension of type A aortic dissection in less than 8% of cases [2]. The right coronary artery is more commonly involved [2,3]. Acute coronary involvement due to aortic dissection is not always associated with a flow limiting disease. Patients with coronary involvement are usually much younger, have a higher aortic regurgitation rate and less commonly, have intramural hematoma [3].

Aorto-right ventricular fistula (ARV fistula) is an exceedingly rare finding. It is mostly an acquired abnormality caused by infection, trauma or iatrogenic post surgical or percutaneous interventions. The most common iatrogenic causes for ARV fistulas are aortic valve replacement and to a lesser degree type A aortic dissection repair [4,5]. Few cases have been reported post transfemoral aortic valve implantation (TAVI) [6]. A well recognized rare cause of ARV fistula is rupture of sinus of valsalva aneurysms whether congenital or acquired [7]. There are some exceptional reported cases of ARV fistulas related to fractured sternotomy wires and pectus bar migration [8,9].

Diagnosis of aorto-right ventricular fistula was almost exclusively done by echocardiography. Confirmation of aorto-right ventricular fistula with cardiac CT was reported previously in only seven cases in English literature (Table 1) [5,10-14], four of these seven cases were congenital, two post aortic valve replacement and the last one was due to type A aortic dissection.

**Table 1:** Cases of Aorto-right Ventricular fistula followed up by Cardiac CT.

<table>
<thead>
<tr>
<th>Case</th>
<th>Year</th>
<th>Age</th>
<th>Gender</th>
<th>Modality</th>
<th>Cause</th>
<th>Management</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fonseca et al. [5]</td>
<td>2014</td>
<td>68</td>
<td>Male</td>
<td>TTE, CT</td>
<td>10 years following Type A aortic dissection</td>
<td>The patient was rejected for surgery due to very high surgical risk</td>
<td>Sudden hypotension and death</td>
</tr>
<tr>
<td>Al-Maskari et al. [10]</td>
<td>2014</td>
<td>59</td>
<td>Female</td>
<td>TTE, TEE, CT, Cath</td>
<td>12 years following aortic valve replacement</td>
<td>Percutaneous device closure</td>
<td>Doing well after 6 months follow up</td>
</tr>
<tr>
<td>Masri et al. [12]</td>
<td>2013</td>
<td>24</td>
<td>Male</td>
<td>TTE, CT</td>
<td>Congenital</td>
<td>Surgical repair</td>
<td>Smooth post-operative course</td>
</tr>
<tr>
<td>Dwivedi et al. [13]</td>
<td>2012</td>
<td>15</td>
<td>Male</td>
<td>TTE, CT, Cath</td>
<td>Congenital</td>
<td>Surgical closure</td>
<td>Not provided</td>
</tr>
<tr>
<td>Pinaud et al. [7]</td>
<td>2009</td>
<td>51</td>
<td>Male</td>
<td>TEE, CT, Cath</td>
<td>Congenital</td>
<td>Surgical closure</td>
<td>Not provided</td>
</tr>
<tr>
<td>Amabile et al. [14]</td>
<td>2008</td>
<td>82</td>
<td>Male</td>
<td>TTE, TEE, CT</td>
<td>10 years following aortic valve replacement</td>
<td>Surgical closure</td>
<td>Died</td>
</tr>
</tbody>
</table>

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Spontaneous closure of ARV fistula has not been reported and thus, careful follow-up of all patients with ARV fistula is prudent [15,16]. Treatment of aorto-right ventricular fistula depends on the cause, concomitant hemodynamics status and degree of left to right shunts.

Samuels et al. [17] In their series of 40 patients with traumatic ARV fistula reported a satisfactory surgical outcome in all of the 38 patients who underwent surgical repair. The mean interval between the time of injury and definitive repair was 1.5 years. Percutaneous closure with septal occluders have been recently reported in patients who are poor surgical candidates [18].

**Conclusion**

Imaging in patients post repair of type A aortic dissection and aortic valve replacement should search for complications of the original disease like the RCA dissection in our case that can be clinically silent and overlooked in the sitting of urgent surgery or complications related to surgery like the ARV fistula. The combination of coronary dissection and ARV fistula post type A aortic dissection repair is a very rare occurrence. The case highlights the additional values of non-invasive multimodality imaging in diagnosis, risk stratification and management planning.

**References**