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Case Report
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Aorto-Right Atrial Fistula of Unknown Etiology: A Case Report

*Abiah S Jacob[1], Sanjeev Bhattacharyya[2], Thomas A Treibel[2] and Wael I Awad[1]**

[1]Department of Cardiothoracic Surgery, Barts Heart Centre, St. Bartholomew's Hospital, UK

[2]Department of Cardiology, Barts Heart Centre, St. Bartholomew's Hospital, UK

Abstract

Aorto-Atrial Fistulas (AAF) is rare but potentially life-threatening conditions. Acquired AAF is most commonly secondary to endocarditis. Treatment modalities include medical management, percutaneous device closure, or surgical intervention. We report a case of AAF in a 54-year-old male treated surgically.

Keywords: Aorto-atrial fistula; Fistula; Connective tissue disorders; Cardiac surgery

Introduction

Aorto-Atrial Fistulas (AAF) is rare and complex pathological conditions which are potentially life-threatening. They can either be congenital or acquired. Acquired cases of AAF are most commonly secondary to another pathological condition. Understanding of the underlying disease is paramount in enabling planning of the optimal therapeutic strategy [1]. Treatment modalities include medical management, percutaneous device closure or surgical intervention.

Case Presentation

A 54-year-old Caucasian male presented with symptoms of worsening dyspnea, dizziness, central chest Paimon exertion and fatigue. He was of short stature and on examination had pectus excavatum, short malformed thumbs, and a para-umbilical hernia. Investigations confirmed aortic and mitral valve regurgitation and a fistula between the aorta and right atrium.

His past medical history included hypertension and dilated cardiomyopathy. He also had anal atresia at birth, unilateral renal agenesis, and bilateral inguinal hernia repairs. He had a family history of valvular pathologies and aortic rupture.

Cardiac angiogram showed a wide necked fistula between the non-coronary sinus and the right atrium with contrast leak into the right atrium above the tricuspid valve (Figure 1A, 1B). There was no aneurysmal dilatation of the sinuses of Valsalva. CMR confirmed a shunt between the non-coronary sinus and right atrium with continuous flow with Qp:Qs of 1.2 (Figure 1C). Echocardiography also confirmed a Barlow-type mitral valve with thickened leaflets and bileaflet prolapse (Figure 2). On follow up, he had worsening symptoms and subsequent transthoracic echocardiograms showed worsening moderate mitral regurgitation. There was a mobile mass in the aortic root area (5 mm × 9 mm), arising from the fistula. His left ventricular function was preserved.

At operation, the aorta and right atrium were opened to identify the fistula tract above the annular plane of the non-coronary sinus. A probe could be passed through the non-coronary sinus into the right atrium.

His aortic valve was trileaflet with thin, friable leaflets and multiple fenestrations. The fistula was excised and repaired directly. An attempted patch closure was abandoned due to proximity of the tricuspid valve and anatomical distortion. The mitral valve was not suitable for repair and hence was replaced with a mechanical prosthesis. The patient underwent mitral valve replacement with a 27 mm mechanical prosthesis and aortic valve replacement with a 21 mm mechanical prosthesis (On-X, Cryolife Inc).

He developed a pericardial effusion 15 days postoperatively with echocardiographic features of cardiac tamponade, drained electively *via* a subxiphoid incision. Post-operative TTE confirmed well-functioning valves and a preserved left ventricular systolic function. Histopathology of the excised fistula tract showed fibromyxoid disease. Mitral and aortic valve histopathology showed thickened

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Correspondence:
Wael I Awad, Department of Cardiothoracic Surgery, Barts Heart Centre, St. Bartholomew's Hospital, West Smithfield, London EC1A 7BE, UK, Tel: +44 02037658744; E-mail: wael.awad@nhs.net

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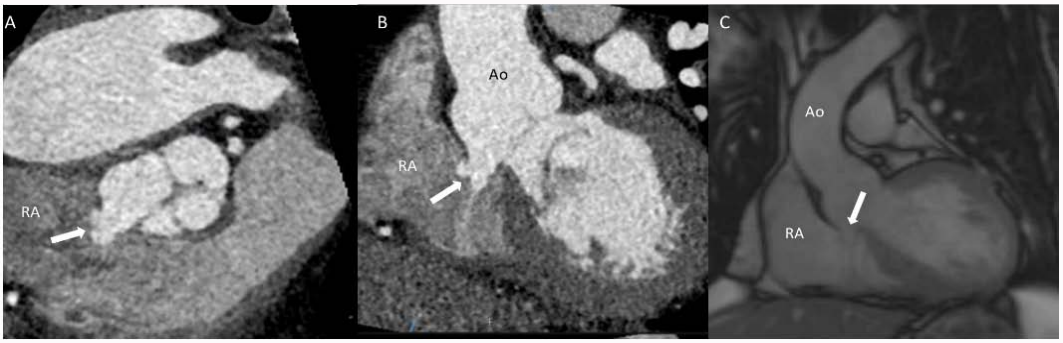

Figure 1: Panel A and B. Computed tomography. Aorta to Right Atrial (RA) fistula (arrow). Panel C: Cardiac Magnetic Resonance Imaging. Aorta to RA fistula (arrow).

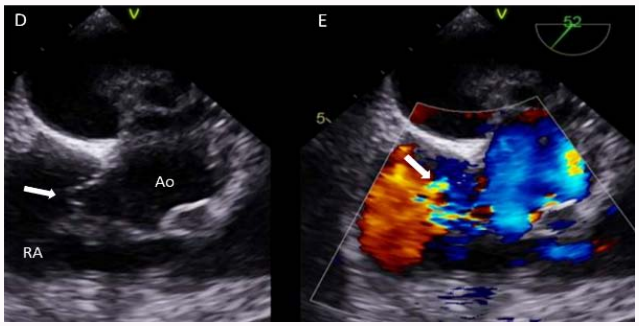

Figure 2: Panel D: Transesophageal Echocardiogram. Short axis view, Aorta to RA fistula (arrow). Panel E: Color Doppler showing flow from aorta to RA (arrow).

leaflets and dense fibrosis with mucopolysaccharide deposition, suggestive of fibrosis and myxoid degeneration. Microbiological examination showed no evidence of endocarditis.

Discussion

AAF is an uncommon but complex pathological condition that can be congenital or acquired, most commonly acquired following endocarditis. It is characterized by abnormal intracardiac flow between the aorta and the left or right atrium. Congenital AAF can be aorto-atrial tunnels or coronary cameral fistulas. Aorto-atrial tunnels are congenital anomalies originating above the sinotubular ridge, caused by inherited weakness in the aortic wall leading to tunnel formation into one of the atria [1]. Coronary cameral fistulas occur when one of the coronary arteries forms a shunt directly into the atria [1,2]. The incidence of AAF in the setting of endocarditis ranges from 1.6% in patients with native valve disease up to 5.8% in patients with a prosthetic valve [3]. In patients with no history of cardiac surgery or infection, aortic dissections or aneurysms are the major cause of AAF formation.

Patients with inherited connective tissue disorders are predisposed to vascular and digestive ruptures, typically arterial ruptures. In connective tissue disorders coronary artery rupture leads to intramural fistulas with the atrium [4].

Few cases of idiopathic aortic root-atrial fistulas like our case have been reported [5]. Patients developing AAF present with signs and symptoms pertaining to the underlying cause. If left undetected, AAF may lead to endocarditis, cardiac overload, and eventual death.

Diagnosing patients with AAFs can prove challenging depending on the size of the shunt. In our case, AAF was found incidentally on cardiac imaging. Our patient had no history of endocarditis or prior cardiac surgery predisposing to AAF. His clinical presentation, past medical history, family history, and histopathology reports suggest an etiology of connective tissue disorder leading to AAF. Connective tissue disorders should be considered in patients with AAF and aortic root or valvular pathology. Early diagnosis may prevent complications like endocarditis and progression to heart failure.

Author Contributions

Abiah Jacob: Writing - original draft, Writing - review & editing; Sanjeev Bhattacharya, Thomas A Triebel: Resources; Wael I Awad: Supervision, Writing - original draft, Writing - review & editing.

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