

Aorta-Right Atrial Tunnel Causing Heart Failure in a Young Female

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Abstract Aorta–right atrial tunnel is a rare entity characterized by a vascular fistula, which connects one of the sinuses of Valsalva in the ascending aorta and the right atrium. It is frequently associated with congestive heart failure. This abnormality is usually diagnosed by echocardiography or CT angiography. It requires immediate intervention due to high risk of complications as congestive heart failure. In this report, we present a case of a 33-year old female with no significant past medical history presents to the hospital with progressive shortness of breath found to have a congenital Aorta–right atrial tunnel.

Keywords: aorta-right atrial tunnel, aorta-right atrial fistula, heart failure

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1. Introduction

Aorta–right atrial tunnel is a vascular fistula which connects one of the sinuses of Valsalva in the ascending aorta and the right atrium. It was first reported in 1980 [1]. The etiology is largely congenital or secondary to endocarditis. The connection leads to shunting systemic circulation with right atrium. The clinical presentation usually includes symptoms of heart failure or on occasions, the patients is asymptomatic [2]. It is usually diagnosed by transthoracic or transesophageal echocardiography, computed tomographic angiography. Urgent treatment of this condition should be provided, giving high risk of complications mainly congestive heart failure and endocarditis. The definite treatment is closure of the shunt either by surgical approach or by cardiac catheterization. Here, we present an interesting case of a 33-year old female with no significant past medical history presents to the hospital with progressive shortness of breath found to have a congenital Aorta–right atrial tunnel.

2. Report of the Case

33-year old female from Peru with no significant past medical history presents to our hospital with progressive

shortness of breath for the last four weeks. Up until one month ago, she was active and healthy until she had flu-like symptoms of rhinorrhea, sore throat, and muscle fatigue that lasted for 2 weeks. She remained afebrile with no chills or rigors. The symptoms were alleviated with over the counter medications, acetaminophen and diphenhydramine. One week before presentation, she experienced palpitations and progressive dyspnea with significant decrease in exercise tolerance. On the day of presentation, physical examination was significant for tachycardia, continuous murmur over precordium and lower extremity edema. Electrocardiography showed right ventricular hypertrophy with repolarization abnormality (Figure 1). Transthoracic echocardiography showed ejection fraction estimated to be 65% with moderate paradoxical motion and moderate systolic flattening of the septum; and dilated right atrium and ventricle. These changes are consistent with right ventricle volume and pressure overload; and noncoronary cusp of aortic valve demonstrated a perforation extending to the right atrium with a large mass associated with the site of extension to the right atrium which may represent a vegetation with left to right atrial shunting noted (Figure 2, Figure 3). Transesophageal echocardiography confirmed the connection between non-coronary aortic cusp and right atrium with a suspicion of endocarditis (Figure 4 A,B). Computer Topography of the chest with intra venous contrast showed a connection between aorta and right atrium (Figure 5). She was started

on antibiotics for possible endocarditis and scheduled for surgery. She was treated with surgical closure of the Aorta-right atrial tunnel with aortic valve replacement. Cultures from the blood and the valve were negative for infection that confirmed congenital aorta-right atrial

tunnel. Hence, antibiotics were stopped. The surgery was complicated by transient third degree heart block (Figure 6) likely due to perivalvular inflammation. Follow-up reveals significant functional recovery of shortness of breath and exercise tolerance.



Figure 1. EKG of the patient at presentation demonstrates sinus tachycardia Right ventricular hypertrophy with repolarization abnormality

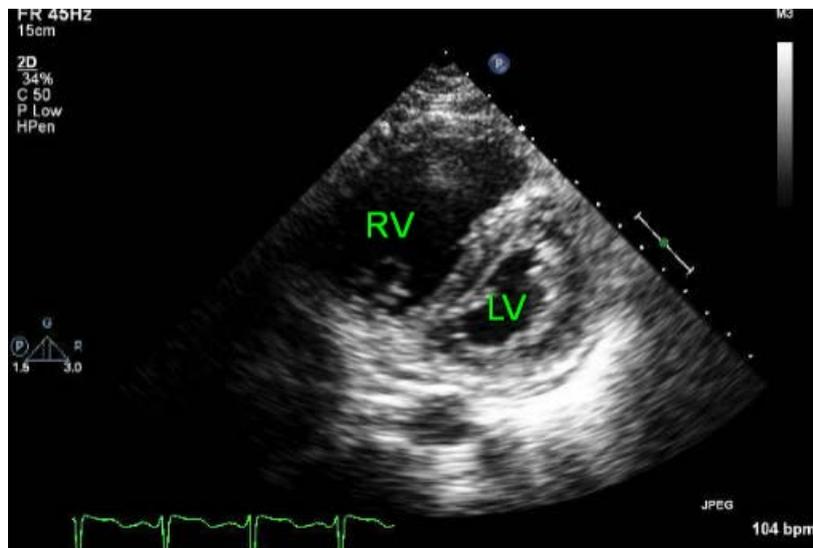


Figure 2. Transthoracic echocardiography, short parasternal view demonstrates moderate systolic flattening of the septum; the ventricle was moderately to markedly dilated with moderately to markedly reduced systolic function

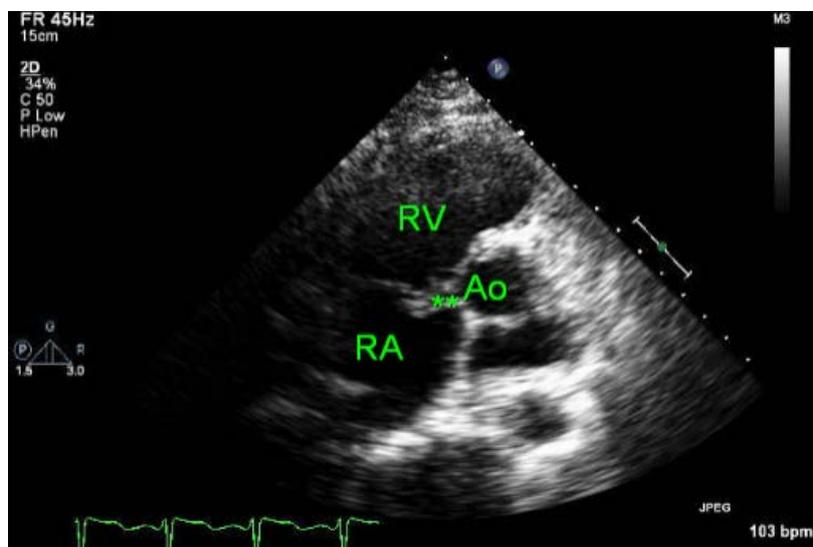


Figure 3. Transthoracic echocardiography: Non-coronary aortic cusp noted to be ruptured or perforated into the right atrium just above tricuspid septal leaflet. There is blood entering the right atrium from the noncoronary sinus. The stars are indicating the area with defect

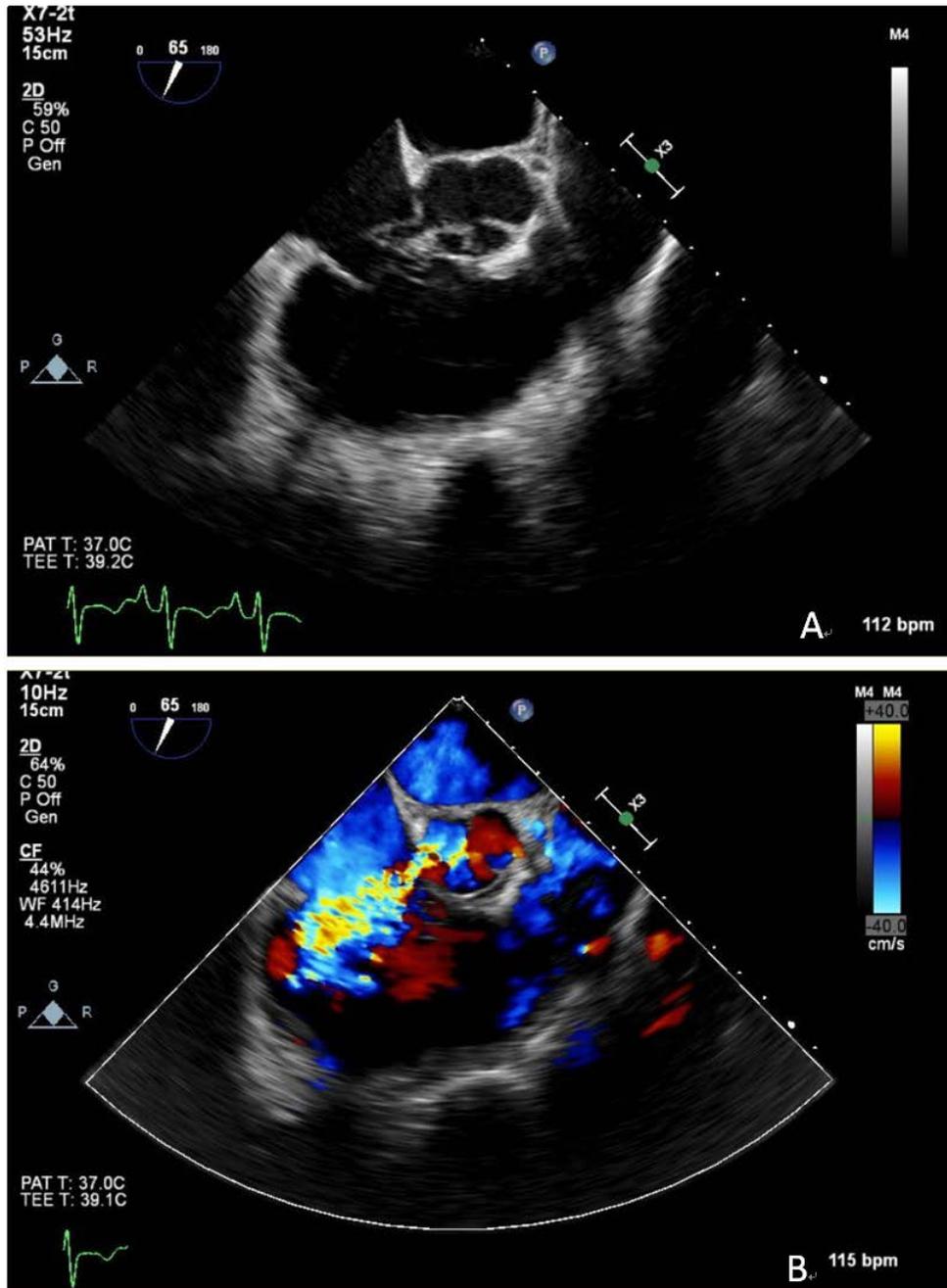


Figure 4 A,B: transesophageal echocardiography (A) with colored Doppler (B): The noncoronary cusp demonstrated a perforation/rupture extending to the right atrium with a large mass associated with the site of extension to the right atrium and significant left to right shunting noted

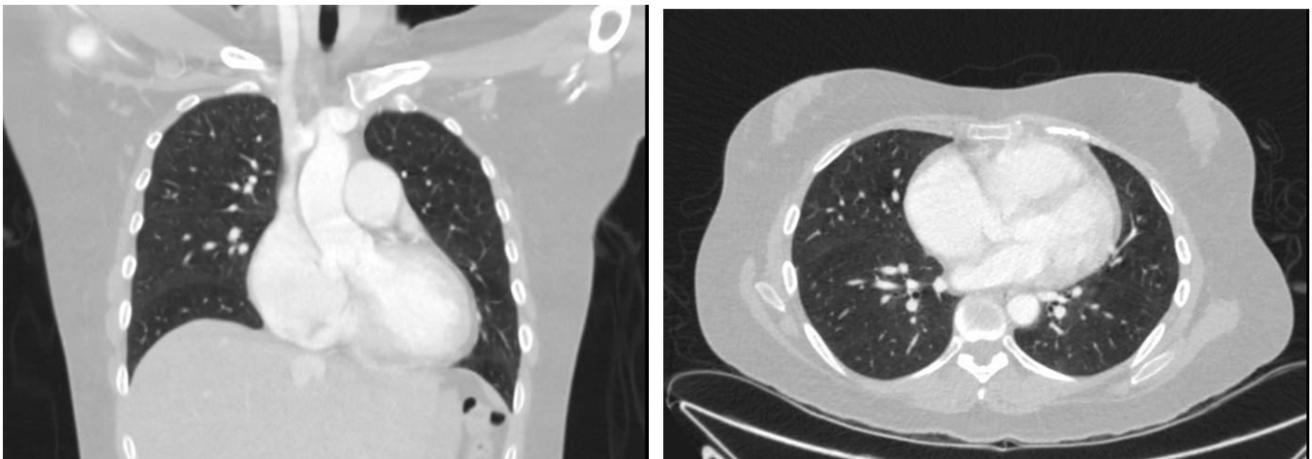


Figure 5. CT chest with IV contrast: a connection between aorta and right atrium

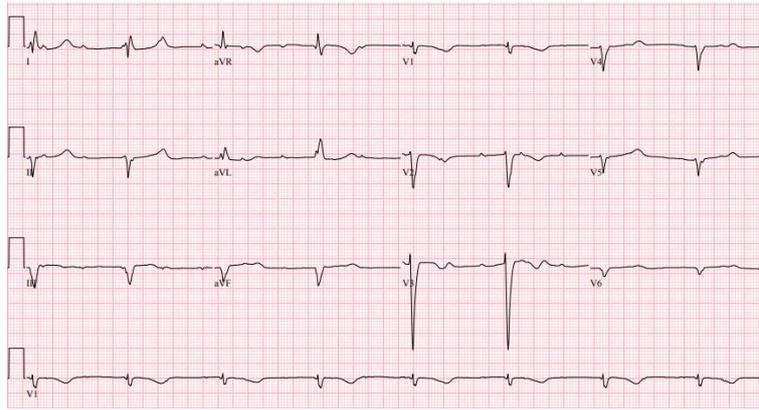


Figure 6. EKG of the patient after surgical intervention demonstrates third degree heart block

3. Discussion

Aorta–right atrial tunnel is a rare entity characterized by a vascular fistula, which connects one of the sinuses of Valsalva in the ascending aorta and the right atrium. It is classified according to its relation to the ascending aorta to anterior or posterior types [2]. The etiology of aorta–right atrial tunnel is usually congenital [3]. It was proposed that the origin of this tunnel is an abnormal embryonic formation of the supravalvular ridge and persistent mesocardial cysts and weakness of aortic intimal layer [1].

The clinical presentation ranges from asymptomatic continuous murmur on precordium to the symptoms of right sided heart failure. It is usually diagnosed by transthoracic or transesophageal echocardiography, computed tomographic angiography of the heart, ascending aortography, and cardiac catheterization [4]. It is important to differentiate aorta–right atrial tunnel from more common abnormalities like rupture of the aneurysm of the sinus of Valsalva as the former originates from above the sinotubular ridge and from coronary-cameral fistula by the absence of myocardial branches [5].

The treatment of this condition is important even in asymptomatic cases due to the risk of aortic valve dysfunction, likely regurgitation, congestive right sided heart failure followed by left sided heart failure, calcification, risk of infective endocarditis, aneurysmal dilatation, infective endocarditis, and pulmonary hypertension [4]. Treatment options are surgical closure under cardiopulmonary

bypass, coil embolization by cardiac catheterization, and external ligation under controlled hypotension. The choice of management is based on patient condition, the size of the tunnel, the presence of endocarditis, concomitant valvular lesion, and operator expertise [4].

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