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IMAGES IN PEDIATRIC CARDIOLOGY

Congenital Aorta Right Atrial Fistula: Successful Transcatheter Closure With the Amplatzer Occluder

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Case Report

A 12-year-old girl with a 4-year history of effort dyspnea and palpitations was referred to our institution for evaluation. Physical examination of the patient showed a continuous grade 3/6 heart murmur best heard along the right upper sternal border. A two-dimensional echocardiogram exhibited a small interatrial septal defect (diameter, 4 mm) with a bidirectional shunt and a high velocity jet in the dilated right atrium. A computed tomographic (CT) angiogram of the patient showed anomalous tortuous communication between the ascending aorta and the right atrium arising adjacent to the right coronary artery (Fig. 1).

An ascending aortogram combined with selective angiography confirmed the presence of a large fistula with a broad origin (diameter, 5.6 mm) from the right aortic sinus anterior to the right coronary artery and a narrow termination into the posterior wall of the right atrium (Fig. 2). Cardiac catheterization showed the presence of a left-to-right shunt with a pulmonary-to-systemic blood flow ratio (Q_p:Q_s) of 3:1. Coronary angiography demonstrated normal coronary arteries arising from the respective sinuses. Because the fistula had a separate anterior origin and a narrow terminal ending into the right atrium, a decision was made in favor of transcatheter device closure.

The fistula was hooked antegradely using an Amplatzer right 1 (AR-1) catheter (Medtronic Inc., Minneapolis, MN, USA), and a standard percutaneous transluminal coronary angioplasty (PTCA) guidewire balance middle weight (BMW) was passed through the tortuosity of the fistula toward the right atrial end. The wire then was retrieved from the right atrial end with a snare and exteriorized from the right femoral vein. A long sheath available with the device then was passed through the right femoral venous route to the right atrium. An Amplatzer duct occluder (AGA Medical Corporation, Plymouth, MN, USA) size 8 × 6 mm was chosen for closure of the fistula and passed through the sheath with the help of the Amplatzer 180° delivery system (Fig. 3). The fistula was successfully closed using the occluder with its large end placed through the fistula toward the aortic side. After deployment of the device, no residual flow through the fistula was demonstrated (Fig. 4).

Discussion

The aorta right atrial fistula lies in a group of abnormal vascular connections of the aorta named aortocameral fistulas. Aortocameral fistulas of the heart are rare extracardiac vascular channels, and their natural history, clinical presentations, and suitability of treatment options are unclear. The fistulous vascular extracardiac communication can have an origin from any of the three sinuses of Valsalva, although they are extremely rare from the noncoronary sinus. Although the ascending aorta is reported to be the most common site of origin for aortocameral fistulas, reports have described fistulas emanating from the descending thoracic aorta [2, 4].

Acquired aortocameral fistulas, which seem to be more common than the congenital form, have been described in association with prosthetic valve endocarditis after root repair and transcatheter closure of septal defects [1]. Commonly described associated cardiac anomalies with

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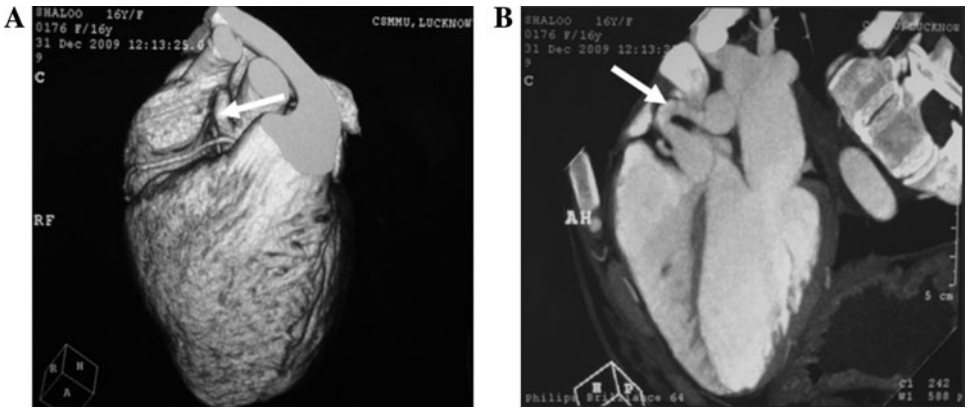

Fig. 1 Aorta-right atrial fistula (*white arrow* in panel A&B) as visible on CT cardiac angiography passing anterior to the right coronary artery

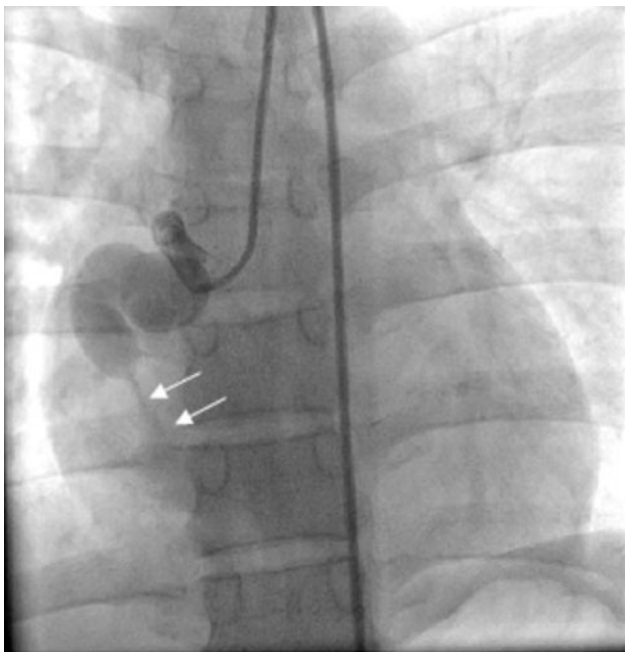

Fig. 2 Demonstration of aorta-right atrial fistula and its tortuous course after selective hooking on invasive angiography. Jet in right atrium is visible (*white arrows*)

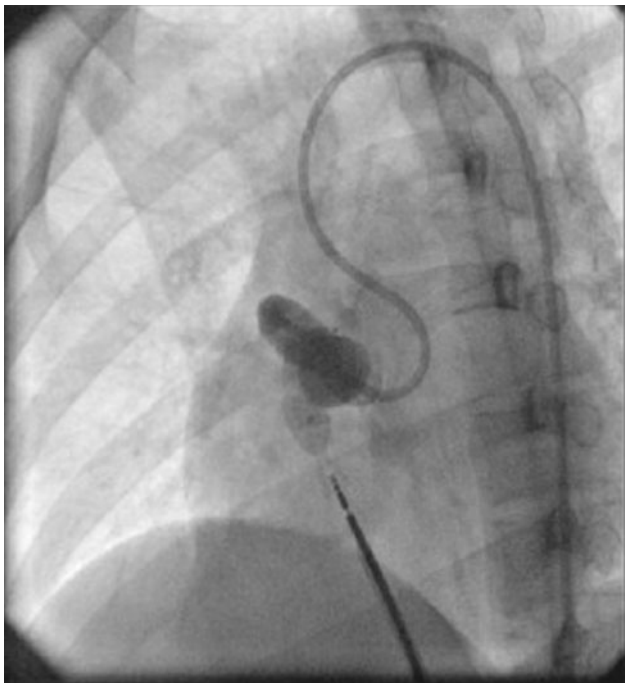

Fig. 3 Passing of Amplatzer device (8 x 6 mm) using 180° delivery system

this condition include the secundum type of atrial septal defect and persistence of the left superior vena cava [3].

Most of the patients described in the literature are reported to be asymptomatic, but a patient can have symptoms of palpitation, effort dyspnea, and recurrent respiratory tract infections [3]. Although fistula can be readily identified by two-dimensional echocardiography, retrograde aortography combined with selective coronary angiography is essential to a demonstration of its course and the coronary ostia.

Aorta right atrial fistulas can be classified as anterior or posterior according to their origin and course in relation to the ascending aorta [3]. The development of symptoms mandates fistula closure. Closure of an aorta right atrial fistula in asymptomatic patients is recommended [5] due to the low rate of procedural complications and the risk for volume overload of both ventricles, bacterial endocarditis, pulmonary vascular disease, aneurysm formation, and spontaneous rupture because of continued patency.

Various treatment options are available according to the type of fistula, its caliber, tortuosity, calcification, course, and relation of the coronary ostia to the aortic orifice of the fistula. Transcatheter closure of the fistula can be a suitable management option provided the opening of the right atrial end is small, the appropriate device according to the size is available, and there is a constriction in the course of the fistula [3].

Surgical treatment has the benefit of reconstructing the dilated sinus of Valsalva or minimizing the likelihood of

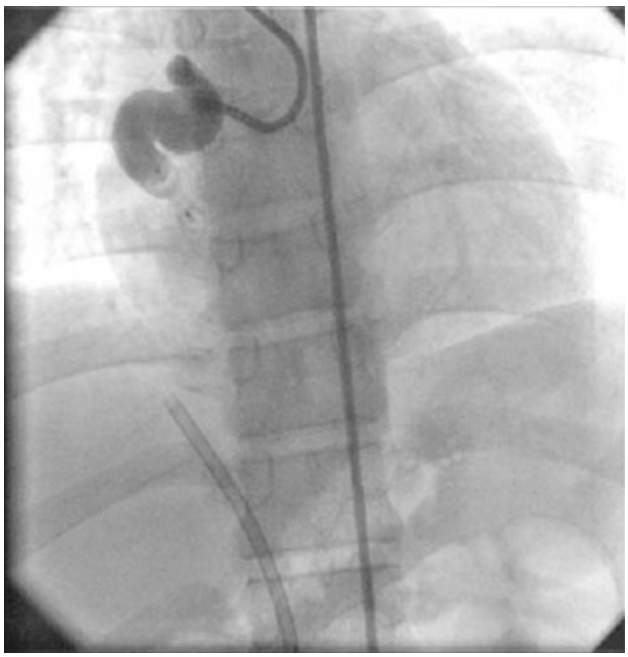

Fig. 4 Successful deployment of Amplatzer device (no residual flow is demonstrated)

further dilation by ligation of the fistula close to the sinus. Surgical closure of the aorta right atrial tunnel includes ligation near the aortic end for anteriorly located fistulas and between the superior vena cava and aorta as close to the aorta as possible for posteriorly located fistulas.

Whether fistula should be closed at both ends and what will be the risk of thromboembolism to systemic circulation if only the distal end is closed still remain unanswered. Transcatheter treatment is a useful management option for selected cases. However, persistence of the dilated sinus of Valsalva with transcatheter device closure constitutes concern and demands further follow-up evaluation for a determination of its evolution.

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