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Congenital Aortocaval Fistula from Right Subclavian Artery to Superior Vena Cava in an Adult with Tetralogy of Fallot

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ABSTRACT

Congenital aortocaval fistula in association with complex congenital heart disease has never been described before. We represent an adult with tetralogy of fallot and an undiagnosed subclavian artery to superior vena cava fistula in previous catheterisms. He underwent surgical correction, successfully. After 8 months post operation he was doing well with improved functional capacity and no cyanosis.

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Introduction

Congenital aortocaval fistula is a very rare condition which resembles anomalies causing right-to-left shunting (1-4). This condition occurs at different ages, although it is most commonly detected in infants and children (2) by a continuous murmur over the chest; this condition is highly mistaken for patent ductus arteriosus.

Congenital aortocaval fistulas are often asymptomatic. However, if present, the symptoms are mainly associated with the size, location, and duration of the Arteriovenous communication (5) and could initially manifest with heart failure. In such cases, angiography (conventional computed tomography and magnetic resonance imaging) is the best tool for diagnosis (6).

Case report

A 32-year-old male patient with a prior history of Tetralogy of Fallot (TOF), who had undergone Waterston shunt 20 years ago, was referred to our Adult Congenital Heart Disease Clinic (ACHD) for further evaluation. Severe cyanosis and clubbing, along with continuous murmur along the upper right sternal border, were the prominent findings of physical examinations. In addition, echocardiography showed TOF with a major aortopulmonary collateral artery (MAPCA) and no Waterston shunt.

Catheterism had been already performed twice and TOF had been approved. The first catheterism, performed 21 years earlier, had indicated the need for total repair. However, the surgeon at the time of surgery had decided to perform a shunt instead of total repair, given the

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small size of main pulmonary arteries (PAs) and PA branches. In the second catheterism, performed 6 years earlier, a closed shunt had been indicated, although the patient refused to undergo any procedures.

Afterwards, a third catheterism was planned, showing TOF. The contrast media, injected in the aorta, had opacified the dilated superior vena cava (SVC) and right atrium, where fistulous connection between the right subclavian artery and SVC was anticipated (Figure 1); nevertheless, aortic and central venous oxygen saturations were similar (76%).

Total repair of TOF with a pulmonary transannular pericardial patch and pericardial patch for ventricular septal defect (VSD), in addition to fistula closure, was performed. The patient was healthy enough to be discharged after 6 days with no cyanosis or continuous murmur. Postoperative echocardiography revealed a small residual VSD with severe pulmonary regurgitation and peak transpulmonary pressure gradient of 80 mmHg, which decreased to 30 mmHg after 8 months.

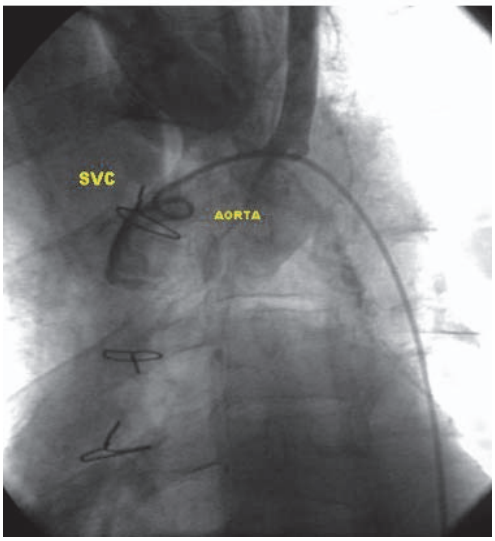

Figure 1. Connection between the right subclavian artery and the SVC was anticipated

Discussion

We presented a patient with a prior history of TOF, who was diagnosed with congenital aortocaval fistula. Continuous murmur over the patient's chest, indicated in the physical examination, had been disregarded since MAPCA could also present with the same signs. The echocardiographer missed to search for extra-cardiac pathology, considering the rarity of congenital aortocaval fistulas.

In such cases, it is preferable to consider all possible differential diagnoses including traumatic or congenital aortovenous fistula, coronary arteriovenous fistula, pulmonary arteriovenous fistula, sinus of valsalva aneurysm, pseudotruncus intercostal collateral vessels with aortic coarctation, VSD with aortic insufficiency, absence of pulmonary valve, aortopulmonary window, pulmonary arterial coarctation, mammary soufflé and venous hum (2). In the current case, the patient's deep cyanosis explained all his symptoms and decreased his functional capacity in a way that no other causes were noted.

Very few similar cases have been reported in the literature. A fistula between the aberant right subclavian artery and SVC has been described by Wong and colleagues (6). Guirezz et al. also reported three cases of communication between the brachiocephalic artery and SVC (7). All of these communications functioned as a left-to-right shunt and were all found in children without any other congenital cardiac diseases. It is generally recommended that these conditions be surgically repaired to avoid more significant hemodynamic changes, bacterial endarteritis and degeneration or aneurismal events (2). Moreover which have suitable anatomy for interventional closure (8).

The unique pattern of the presented case was disease manifestation during adulthood, accompanied by another congenital cardiac disease. To the best of our knowledge, this form of fistulous connection, accompanying such a complex congenital cardiac disease like TOF, has not been previously reported. In fact, other types of involvement, besides the previously introduced congenital cardiac diseases, should be noted. Moreover, other patients introduced by other clinicians should be considered as new cases and dependent re-examinations need to be performed.

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