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A Case Report on an Aortic Root Pseudoaneurysm, a Rare Complication of Infective Endocarditis Revealing a Multitude of Aortic Arch Anomalies Including an Aortic Coarctation in a 20 Year Old Patient

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Abstract **Case Report**

We describe a rare combination of aortic arch variant anatomy, seen on trans-thoracic echocardiogram and cardiac CT during the course of investigations for a suspected aortic dissection in a 20-year-old patient with a history of infective endocarditis. This combination included a truncus bicaroticus, an aberrant right subclavian artery, an aortic coarctation, and a bicuspid aortic valve. This patient also presented a rare complication of infective endocarditis namely a pseudoaneurysm of the Valsalva sinus and some abdominal aortic branch variations. These included a right hepatic artery emanating from the superior mesenteric artery, and a left gastric artery emerging directly from the abdominal aorta adjacent to the coeliac trunk. In isolation, these anomalies have been reported with different frequencies, but as far as we know, this particular combination has rarely been reported. We provide an imagistic portrayal of these anomalies on Tran's thoracic echocardiogram and cardiac CT.

Keywords: Aortic coarctation, Pseudoaneurysm, infective endocardtis, bicuspid aortic valve, cardiac CT.

INTRODUCTION

Coarctation of the aorta (CoA) is a congenital heart disease that accounts for 5%–8% of all congenital heart disease (CHD). It occurs in 4 out of 1000 live births with a male predominance and has been linked to several cardiac and vascular anomalies, such as bicuspid aortic valves (BAV), ventricular septal defects, patent ductus arteriosus and aortic arch hypoplasias [1].

We herein report the case of a young male patient with a history of infective endocarditis, who was admitted to our department for the suspicion of an aortic dissection, and in whom we rather discovered an aortic coarctation coupled with a number of other anomalies notably, an aberrant right subclavian artery, a truncus bicaroticus, a bicuspid aortic valve, as well as a pseudoaneurysm of a Valsalva sinus which was attributed to his past infective endocarditis.

CASE REPORT

A 20 year old male patient was admitted to our hospital for a sudden worsening of his long term dyspnea which raised the suspicion of an aortic dissection. His initial transthoracic echocardiogram revealed a dilated and hypertrophied left ventricle which nevertheless maintained its systolic kinetic function, a severe aortic insufficiency and an aneurysmal dilation of the sinus of Valsalva.

Further imaging in the form of a cardiac CT angiography was performed confirming the presence of a saccular paravalvular pseudo aneurysm of the posterior aortic cusp measuring 32 x 15 x 33 mm with a neck size of 11 mm. The right and left coronary arteries both arose from the anterior aortic cusp, the right coronary artery arising 20 mm above the aortic annulus and the left coronary artery arising 17 mm above the aortic annulus. The cardiac CT angiography additionally showed an array of congenital aortic arch anomalies comprising an aberrant right subclavian artery, a truncus bicaroticus, a bicuspid aortic valve and an aortic coarctation. The latter was post ductal, localized at the level of the fifth dorsal vertebra, and measured 7 mm in diameter. It was responsible for a downstream hypoplasia of the aorta measuring 19 mm in maximal anteroposterior diameter after the coarcation and for the development of an important network of collateral circulation involving the internal thoracic arteries, the intercostal arteries, and the thoracic and abdominal wall arteries.

On the abdominal CT sequences, we noticed some anatomical variations of some of the arteries branching off the abdominal aorta. The right hepatic artery, for instance, rose directly from the superior mesenteric artery instead of bifurcating from the proper hepatic artery. Moreover, the left gastric artery rose directly from the aorta adjacent to the celiac trunk instead of arising directly from the celiac trunk.

DISCUSSION

The first imagistic exam performed in the context of our dyspneic patient was a transthoracic echocardiogram which revealed a dilated and hypertrophied left ventricle nevertheless maintaining its systolic kinetic function, a bicuspid aortic valve, a severe aortic insufficiency and a paravalvular aneurysmal formation of the aortic root.

Due to the limited diagnostic abilities of echocardiography and the fact that this imaging modality is operator-dependent, a cardiac CT was also performed to detect additional anomalies and to explain the patient's clinical symptoms.

The cardiac CT performed confirmed the presence of a posterior paravalvular pseudoaneurysm of the sinus of Valsalva which has been attributed to the patient's history of infective endocarditis. Infective endocarditis is a non-contagious infection, usually bacterial or fungal, of the endocardium and heart valves which may lead to numerous complications. The most common complications attributed to infective endocarditis include valvular dysfunction, perivalvular extension of infection, formation of an abscess or fistula, or aortic rupture. A rarer and much less recognised complication of infective endocarditis is a pseudoaneurysmal formation of the Valsalva sinus as was our patient's case. Pseudoaneurysm of the sinus of Valsalva has been reported to occur in up to 28% of patients with infective endocarditis [2-4].

Other anomalies non visualized on the transthoracic echocardiogram, but which were detected on cardiac CT included various aortic arch anomalies such as a truncus bicaroticus, an aberrant right subclavian artery, and a post-ductal coarctation of the aorta.

The bicuspid aortic valve (BAV) noticed on the transthoracic echocardiogram was also confirmed by the cardiac CT. It represents the most common congenital heart abnormality, with a prevalence of 1–2%. Aortic stenosis, regurgitation and infective endocarditis are the most common complications resulting from a bicuspid aortic valve [5].

The association between bicuspid aortic valves and aortic coarctation present in our patient is well known and has been reported in the literature. In fact, bicuspid aortic valves occur in up to three-fourth of patients with coarctation [6].

Variations in the branching pattern of the aortic arch are due to a modified development of the primitive aortic arches and of the aortic sac during embryological development.

When the aortic sac fails to bifurcate for instance, the left common carotid artery directly connects to the aortic sac leading to the development of a bicarotid trunk [7].

Similarly, an aberrant right subclavian artery is believed to result from an interruption of the embryonic right arch proximal to the seventh cervical intersegmental artery [8].

A quite rare association allying a truncus bicaroticus and an aberrant right subclavian artery was found in our patient. Aberrant right subclavian artery can be associated with other congenital anatomical variants such as bicarotid trunk, Kommerell diverticulum, right sided aortic arch, and non recurrent laryngeal nerve. A syste;ic review study by Polguj *et al*. showed that a truncus bicaroticus was concomittent in 19.2 % of patients with an aberrant right subclavian artery [9].

Our patient also presented a couple anatomical variants of the main branches of the abdominal aorta, namely a right hepatic artery emerging directly from the superior mesenteric artery and a left gastric artery that emanates directly from the abdominal aorta instead of the coeliac trunk. Vascular variations stem from defective fusion of the omphalomesenteric arteries during embryonic life [10]. The presence of aortic arch developmental anomalies should entice the radiologist to look for coexisting vascular anomalies namely abdominal or cerebral which may coexist.

CONCLUSION

We present an unusual pattern of aortic arch and abdominal aortic branch variations and a rare complication of infective endocarditis in the same patient. This emphasizes the importance for the radiologist to be aware of complications that can arise actively seek additional anatomical vascular variants in patients already displaying one variant and of knowing the commonly associated variants.

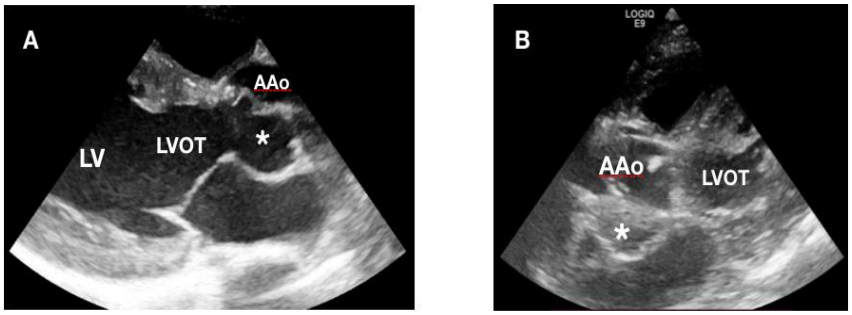

Fig-1: Transthoracic echocardiogram images. A) long axis view of the heart and B) short axis view of the heart showing a paravalvular pseudo aneurysm of the aorta (asterix); LV (left ventricle); LVOT (left ventricular outflow tract); AAo (ascending aorta).

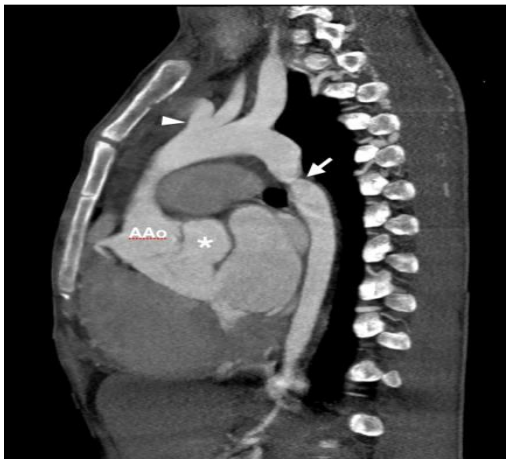

Fig-2: Cardiac multidetector-row computed tomography (sagittal image) showing a truncus bicaroticus (arrowhead); a localized post ductal aortic coarctation (arrow); aortic paravalvular pseudoaneurysm; AAo (ascending aorta).

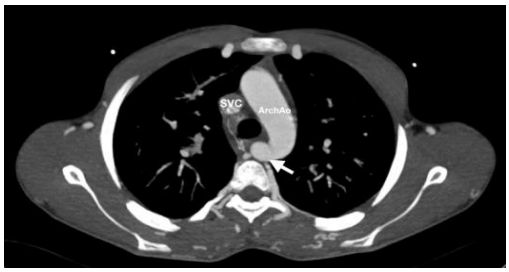

Fig-3: Cardiac multidetector-row computed tomography (axial image) showing an aberrant right subclavian artery (arrow); Aortic arch (ArchAo); Superior Vena Cava (SVC).

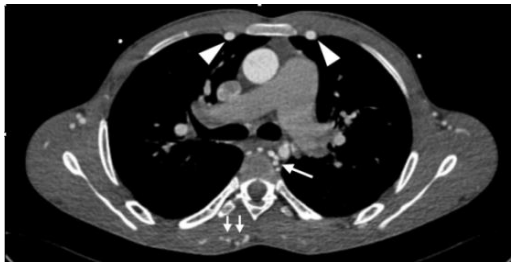

Fig-4: Cardiac multidetector-row computed tomography (axial image) showing a network of collateral circulation involving the internal thoracic arteries (arrowheads), the thoracic cavity wall arteries (double arrows), and a paravertebral collateral circulation (single arrow).

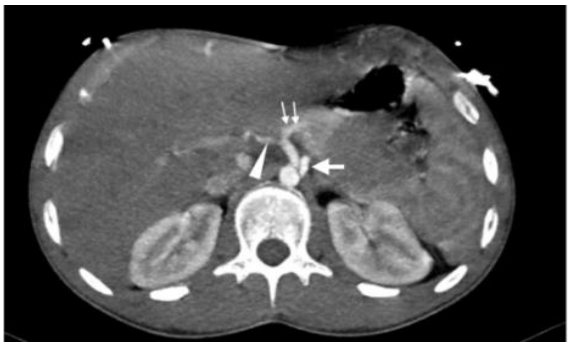

Fig-5: Cardiac multidetector-row computed tomography (axial image) demonstrating an anatomical variation of the coeliac trunk in which the left gastric artery emanates directly from the abdominal aorta (single arrow); proper hepatic artery (arrowhead); splenic artery (double arrow).

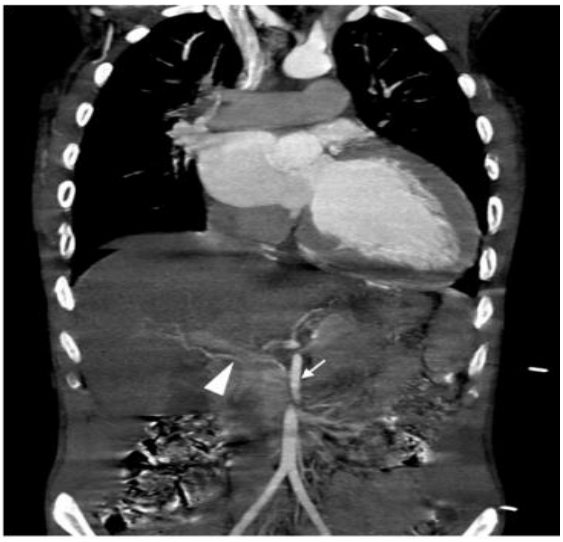

Fig-6: Cardiac multidetector-row computed tomography (coronal image) demonstrating an anatomical variant of the branches of the abdominal aorta consisting of a right hepatic artery (arrow head) emerging directly from the superior mesenteric artery (single arrow).

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