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The First Reported Case of Right Aortic Arch with Left Subclavian Artery Atresia Presenting with Heart Failure with Reduced Ejection Fraction: Exploring an Uncertain Causal Relationship

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

The largest artery in the body is the aorta. The ascending aorta, aortic arch, thoracic aorta, and abdominal aorta are the four sections that make up the aorta. The ascending aorta continues into the aortic arch. It moves inferiorly after arching superiorly, posteriorly, and to the left [1]. The right aortic arch is a rare anatomical anomaly of the aorta [2].

A case of a 43-year-old male was recently diagnosed with heart failure with reduced ejection fraction (HFrEF). Cardiac CT was performed as part of the ischemic heart disease evaluation. The cardiac CT revealed a right-sided aortic arch, along with the absence of the proximal segment of the left subclavian artery, and the perfusion of the left subclavian artery occurring via the left vertebral artery.

This case represents the first documented instance in the literature of heart failure occurring concurrently with these vascular anomalies.

Keywords: Right aortic arch; reduced ejection fraction; congenital heart defect; cardiovascular disease; ejection fraction.

1. INTRODUCTION

The right aortic arch is a rare anatomical anomaly characterized by the aortic arch being positioned on the right side instead of the left, with a reported incidence of approximately 0.1% [2]. “According to Edward's Classification, this condition is categorized into three primary subtypes: Type I, which features a right aortic arch (RAA) with mirror image branching; Type II, which indicates RAA accompanied by an aberrant left subclavian artery; and Type III, which presents RAA with an isolated subclavian artery. Among these, Type II is the most prevalent subtype” [3,4].

“Additional rare subtypes include RAA with isolation of the innominate artery and the right circumflex aorta. Type III, specifically, is a RAA characterized by the presence of an isolated subclavian artery, resulting from the regression of the embryological left arch at two segments adjacent to the left subclavian artery. In some cases, the left subclavian artery may originate from either the vertebral artery or the ductus arteriosus” [5].

“Patients with this condition often exhibit symptoms consistent with subclavian steal syndrome and may report weakness in the left arm” [6]. “If phase-contrast magnetic resonance angiography is conducted, it typically reveals flow reversal in the left vertebral artery. Unlike Type II RAA, Type III is associated with a higher incidence of congenital heart defects, including truncus arteriosus, tricuspid atresia, and tetralogy of Fallot” [7,8]. The recommended treatment approach for this condition typically involves the creation of a bypass graft to connect the common carotid artery to the subclavian artery.

2. CASE PRESENTATION

This report details a 43-year-old male with a medical history significant for bronchial asthma, dyslipidaemia, migraine headaches, and obesity (BMI 30). He was referred to the cardiology department from the family medicine clinic due to complaints of atypical chest pain and occasional palpitations. An electrocardiogram (ECG) revealed a left bundle branch block (LBBB), a finding that was not new for the patient.

Upon referral, a transthoracic echocardiogram (TTE) and a cardiac computed tomography (CT) scan were conducted. The TTE indicated a dilated left ventricle (LV) with a severely reduced LV ejection fraction (EF) of 25% and global hyperkinesia. Consequently, the patient was admitted to the coronary care unit (CCU) for further evaluation and management of newly diagnosed heart failure with reduced ejection fraction (HFrEF).

During the assessment, notable discrepancies in blood pressure readings were observed: 122/99 mmHg in the right arm versus 95/60 mmHg in the left arm. The patient indicated that this difference had been present for a prolonged period but had not been previously addressed.

A chest X-ray revealed the presence of a right-sided aortic arch (Fig. 1). Additionally, cardiac CT demonstrated a calcium score of zero and normal coronary arteries; however, it also confirmed the right-sided aortic arch. To further

evaluate these findings, a chest CT angiogram was recommended.

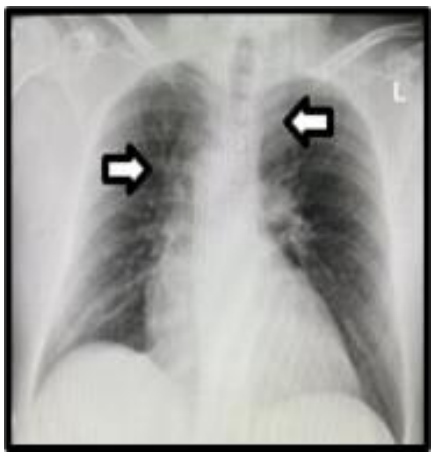

Fig. 1. Chest X- ray

The chest CT angiogram demonstrated a right-sided aortic arch, with the descending thoracic aorta appearing of normal calibre. The branching pattern of the thoracic aorta, from right to left, was identified as follows: right subclavian artery, right common carotid artery, and left common carotid artery. Notably, the proximal segment of the left subclavian artery was absent (atresia), with perfusion of the left subclavian artery occurring via the left vertebral artery, indicative of a steal phenomenon. There was no evidence of aortic aneurysm or dissection.

Mild cardiomegaly with left ventricular dilation was observed. The descending abdominal aorta exhibited a normal course and branching pattern. Examination of the visualized lung parenchyma revealed no abnormal focal or diffuse lesions. Both the pleural and pericardial sacs appeared clear. The remaining branches of the aortic arch were unremarkable, and the left vertebral artery was patent (Figs. 2, 3, 4, and 5).

The patient was diagnosed with non-ischemic cardiomyopathy and commenced on guideline-directed medical therapy (GDMT). Additionally, a second diagnosis of a right-sided aortic arch with atresia of the left subclavian artery was established.

Fig. 2 reveals the presence of a right-sided aortic arch (indicated by the white arrow). The proximal segment of the left subclavian artery is absent (highlighted by the yellow arrow), with perfusion of the left subclavian artery occurring via the left vertebral artery, demonstrating a steal phenomenon.

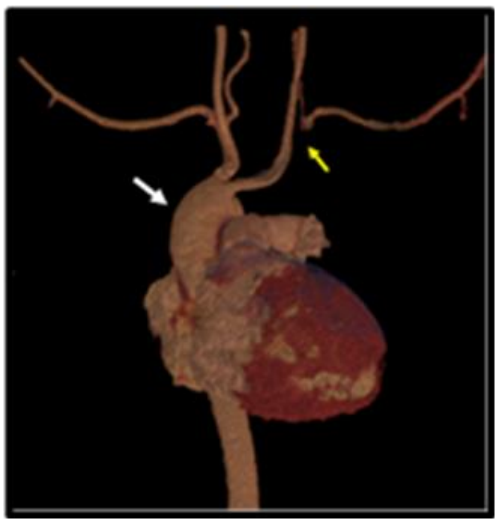

Fig. 2. 3D reconstruction of the heart and thoracic aorta

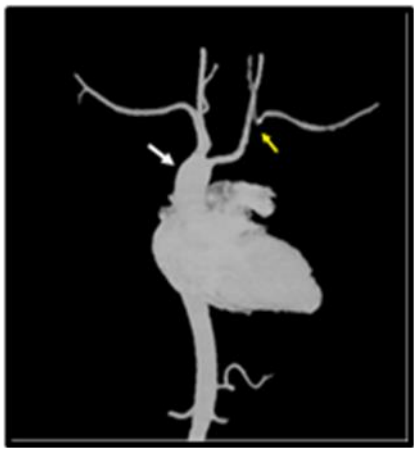

Fig. 3. 3D maximum intensity projection (MIP) angiographic view of the heart and thoracic aorta

Fig. 3 illustrates the presence of a right-sided aortic arch (indicated by the white arrow). The proximal segment of the left subclavian artery is absent (highlighted by the yellow arrow), with perfusion of the left subclavian artery occurring via the left vertebral artery, indicative of a steal phenomenon.

Fig. 4 demonstrates that the left subclavian artery (indicated by the yellow arrow) is receiving its blood supply from the left vertebral artery (highlighted by the white arrow), indicative of a steal phenomenon.

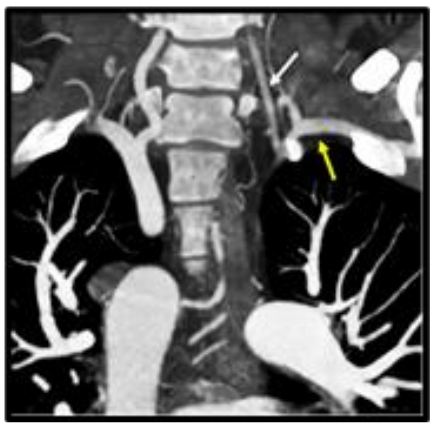

Fig. 4. A coronal maximum intensity projection (MIP) of the chest in the mediastinal window

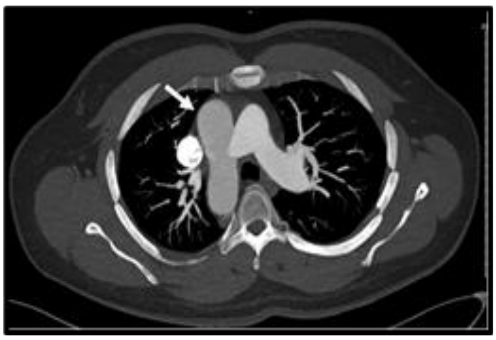

Fig. 5. An axial maximum intensity projection (MIP) of the chest in the mediastinal window at the level of the aortic arch

Fig. 5 reveals the presence of a right-sided aortic arch, as indicated by the white arrow.

3. DISCUSSION

A right-sided aortic arch with an abnormal left subclavian artery is a rare anomaly, occurring in approximately 0.05% to 0.1% of the population. While most cases are asymptomatic, some individuals may present with symptoms related to oesophageal or respiratory compression [9,10]. Steward et al. reported 298 cases of right aortic arch, noting that isolation of the left subclavian artery occurs in only 0.8% of such cases [11].

Despite the rarity of a right aortic arch with an isolated left subclavian artery, “It should be considered in patients exhibiting a right aortic arch on chest X-ray, particularly if accompanied by decreased blood pressure or a delayed and dampened pulse in the left upper extremity during physical examination. This anomaly presents a characteristic angiographic appearance on delayed imaging. Recognizing this condition is clinically significant, as it may lead to vertebrobasilar ischemia or ischemia of the left upper extremity, both of which can be surgically corrected. Furthermore, there is a high incidence of associated congenital heart disease, particularly tetralogy of Fallot. Failure to identify the isolated left subclavian artery may result in unsuccessful attempts to create a left Blalock-Taussig shunt” [12].

4. CONCLUSION

The current case involves an incidentally discovered right-sided aortic arch with atresia of the left subclavian artery, where perfusion of the left subclavian artery occurs via the left vertebral artery. The patient is also diagnosed with heart failure with reduced ejection fraction (HFrEF).

To our knowledge, this is the first documented case in the literature describing heart failure in conjunction with a right aortic arch. This raises important questions regarding the potential relationship between the right aortic arch and the development of heart failure in this patient, including the underlying mechanisms involved, or whether this association is coincidental. While we cannot provide definitive answers at this time, future cases may offer valuable insights. This case may serve to underscore the potential connection between heart failure and right aortic arch anomalies.

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

We hereby declare that NO generative AI technologies, including Large Language Models (such as ChatGPT, COPILOT, etc.) or text-to-image generators, were utilized in the writing or editing of this manuscript.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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