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CASE REPORT
Congenital heart disease

Successful treatment of patent ductus arteriosus accompanying right-sided aortic arch and aberrant left subclavian artery: a case report

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Background The coexistence of a right-sided aortic arch (RAA), an aberrant left subclavian artery (ALSA), and a patent ductus arteriosus (PDA) is a rarely seen vascular ring anomaly. There is currently no general guideline consensus on the management and follow-up of this congenital defect, posing a challenge to the clinicians. At this point, the heart team plays a critical role in the management of the disease.

Case summary In the present case, a 25-year-old male patient was presented to the outpatient clinic with dyspnoea and fatigue. A transthoracic echocardiography revealed PDA with a left-to-right shunt. To evaluate the anatomy thoroughly, a thoracic computed tomographic angiography was performed and showed PDA accompanying ALSA and RAA. The patient was evaluated by the Heart Team, and a percutaneous closure of PDA was recommended due to signs of left ventricular volume overload. The closure was successfully performed with Amplatzer vascular plug II. At follow-up, the patient was free of symptoms.

Discussion Clinicians should be aware of the potential concomitant lesions during the diagnostic work-up. In selected patients, percutaneous closure of PDA may be the first-line therapy in experienced centres.

Keywords Right-sided aortic arch • Aberrant left subclavian artery • Kommerell diverticulum • Vascular ring anomalies • Percutaneous device closure • Congenital heart disease • Case report

ESC Curriculum 2.2 Echocardiography • 2.4 Cardiac computed tomography • 9.7 Adult congenital heart disease • 9.1 Aortic disease

Learning points

- When evaluating a patient with congenital heart disease, the clinician should be alert to the fact that concomitant heart and great vessel anomalies may coexist, and a detailed examination should be performed accordingly.
- A Heart team is essential for determining the treatment of patients with complex congenital heart disease.
- In selected patients, percutaneous closure of PDA may be the first-line therapy in experienced centres.

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Introduction

The patent ductus arteriosus (PDA) is defined as the persistent opening of the ductus arteriosus, which is an essential foetal structure between the proximal descending aorta distal to the left subclavian artery and the proximal left pulmonary artery. Although patients with PDA can be asymptomatic, cardiac function can be affected in some cases.[1] In the current case report, we describe a very rarely seen association of congenital cardiovascular disease, including a right-sided aortic arch (RAA), aberrant left subclavian artery (ALSA) originating from Kommerell's diverticulum and PDA, all of which create a congenital vascular ring formation.

Timeline

Timeline Events	
Day of admission	Patient admitted with dyspnoea and reduced functional effort capacity. The transthoracic echocardiography revealed evidence of PDA with a left-to-right shunt. Computed tomography showed PDA accompanying ALSA and RAA.
24 h after admission	Patient was evaluated by the Heart Team, and the decision was to perform percutaneous closure of the PDA.
48 h after admission	Percutaneous PDA closure was successfully performed with Amplatzer vascular plug II (AVP II).
5th day of hospitalization	Patient discharged.
Follow-up at 6 months	Patient's complaints significantly improved.

Case presentation

A 25-year-old male with a history of PDA accompanied by aberrant left subclavian artery (ALSA) and right-sided aortic arch (RAA) presented to the outpatient clinic complaining of progressive dyspnoea and reduced functional effort capacity with New York Heart Association (NYHA) Class III for an extended period. The patient was diagnosed with an aortic arch anomaly about 3 years ago and was lost to follow-up because of his asymptomatic clinical status. The patient had been suffering from progressive symptoms for the last couple of months, and this brought him to our institution to seek medical advice. He had no past medication history. Physical examination revealed normal heart sounds with a grade 3 continuous murmur over the second intercostal space at the left sternal border. His heart rate was 94 bpm, blood pressure was 120/60 mmHg, oxygen saturation was 98% on ambient air, and peripheral pulsations were normal. The electrocardiogram was in normal sinus rhythm, and a chest X-ray revealed a slightly increased cardiothoracic ratio (*Figure 1*). Initial blood tests were within the normal range, but the brain natriuretic peptide level was slightly elevated at 127.1 pg/mL (normal range: 0,00–86,10 pg/mL). Transthoracic echocardiography showed PDA with a left-to-right shunt between the aorta and pulmonary artery (*Figure 2*; Supplementary material online, *Video S1*). Systolic pulmonary arterial pressure was estimated at 29 mmHg, and right atrial and ventricular sizes were normal. Although left ventricle ejection fraction was normal, mild dilation of the left ventricle and left atrium was observed (left ventricle end-diastolic diameter was 5, 8 cm with a BSA of 3, 3 (cm/m^2); left atrial anteroposterior diameter was 4, 4 cm with a BSA of 2, 5 (cm/m^2)). Finally, the pulmonary-systemic shunt ratio (Qp/Qs) ratio was measured by echocardiography as 1,7:1.

To exclude an additional congenital anomaly that might accompany PDA and obtain more detailed information about the anatomy, the patient underwent thoracic computed tomographic angiography (CTA), which showed PDA accompanied by RAA and ALSA (*Figures 3–5*). CTA revealed a type E PDA with the following diameters: the narrowest was 2 mm at the pulmonary end, and the widest was 6 mm. The left carotid artery, right carotid artery, right subclavian artery, and ALSA were all observed to arise from the RAA (*Figure 4*). The Heart Team held discussions on this patient. Given the lack of symptoms suggestive of tracheal or esophageal compression and with no signs showing on the CTA and any evidence of the left ventricular volume overload, the team pursued the percutaneous strategy for PDA closure. Conservative treatment of abnormal originated left subclavian artery was considered.

Since it was decided to approach the type E PDA retrogradely from the aortic side, a right common femoral arterial puncture was performed. The 6 Fr internal mammary arterial guiding catheter was placed in the aortic side of the PDA. Then, imaging was performed to select an appropriate size for the device and re-evaluate previously taken PDA measurements (Supplementary material online, *Video S2*). To close the PDA at the widest diameter of 6 mm, an 8 × 7 mm Amplatzer vascular plug II (AVP II) (St. Jude Medical, Plymouth, MN, USA) was advanced and deployed. Final angiography showed complete PDA closure without any residual shunt,

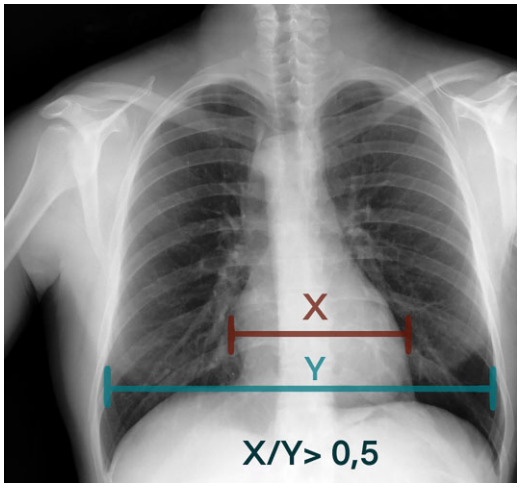

Figure 1 Chest X-ray revealed a slightly increased cardiothoracic ratio.

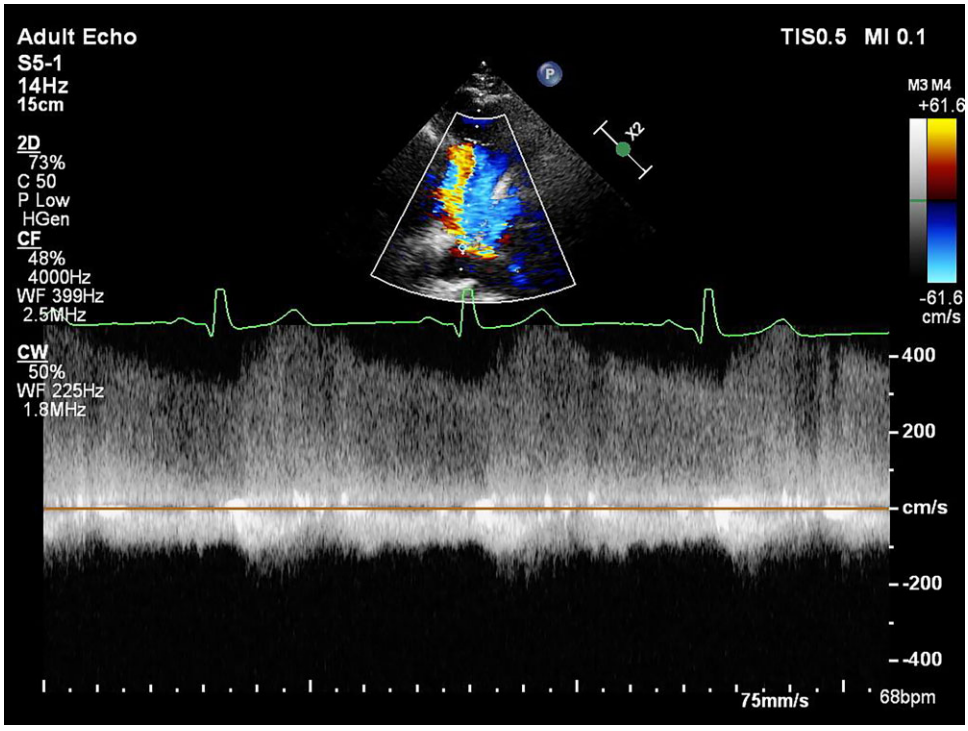

Figure 2 Transthoracic echocardiography in the parasternal short-axis view showing the continuous flow description, which is characteristic of patent ductus arteriosus flow.

and the procedure was completed without any complications (Supplementary material online, *Videos S3–S6*). After 6 months, the patient's effort capacity improved significantly with NYHA Class I, and his complaints subsided completely.

Discussion

The reported incidence of PDA, which accounts for 5–10% of all congenital heart diseases, is predicted to be 1 in 2000 births.[1,2] The direction and the degree of the shunt through the PDA depends on the PDA's diameter and length, the gradient between the systemic and pulmonary artery, and the difference between systemic and pulmonary vascular resistance.[3] According to the European Society of Cardiology guidelines for the management of adult congenital heart disease, PDA closure is recommended in patients with evidence of left ventricle volume overload, such as left ventricle enlargement and increased stroke volume without pulmonary arterial hypertension regardless of symptoms.[4] Given that transcatheter closure of PDA in adults is safe and effective, percutaneous PDA closure has become the first treatment option in selected patients. Considering the preference of patients and recent guidelines, our patient was an appropriate candidate for percutaneous device closure of PDA.[4] However, our patient's coexisting congenital cardiovascular abnormalities necessitated a more detailed approach to ensure the proper treatment strategy.

RAA is an uncommon aortic arch anomaly with an incidence of 0.1–0.5%.[5] The normal aortic arch passes over the left main pulmonary bronchus and descends. However, in RAA anomaly, the ascending aorta passes over the right pulmonary bronchus and descends from the right of the vertebra (*Figure 2*). RAA with ALSA is an

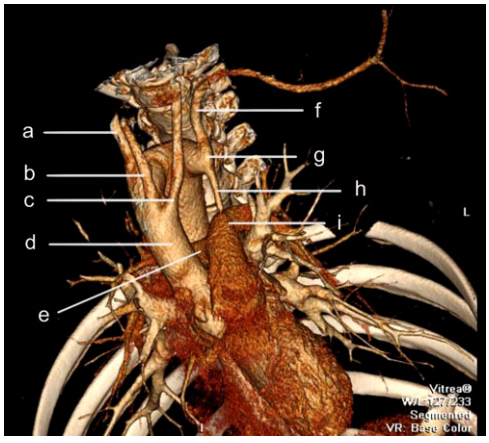

Figure 3 Reconstructed thoracic computed tomographic angiography image demonstrating an anatomical relationship between pulmonary arteries and patent ductus arteriosus, right-sided aortic arch, aberrant left subclavian artery, and Kommerell diverticulum. (*A*) Right subclavian artery, (*B*) right carotid artery, (*C*) left carotid artery, (*D*) right aortic arch, (*E*) right pulmonary artery, (*F*) aberrant left subclavian artery, (*G*) Kommerell diverticulum, (*H*) patent ductus arteriosus, and (*I*) left pulmonary artery.

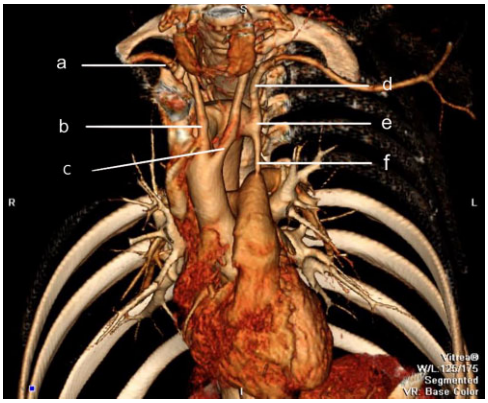

Figure 4 Reconstructed thoracic computed tomographic angiography image demonstrating right-sided aortic arch accompanying anomalies of carotid arteries and right subclavian. (*A*) Right subclavian artery, (*B*) right carotid artery, (*C*) left carotid artery, (*D*) aberrant left subclavian artery, (*E*) Kommerell diverticulum, and (*F*) patent ductus arteriosus.

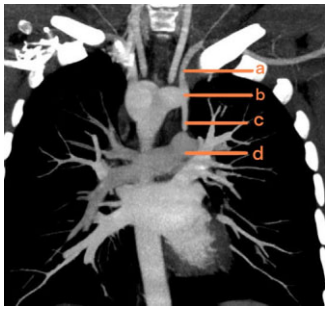

Figure 5 Coronal section of thoracic computed tomographic angiography image showing an anatomical relationship between the left pulmonary artery and aberrant left subclavian artery, patent ductus arteriosus, Kommerell diverticulum. (*A*) Aberrant left subclavian artery, (*B*) Kommerell diverticulum, (*C*) patent ductus arteriosus, and (*D*) left pulmonary artery.

extremely rare condition with an incidence of 0.05%.[6] Kommerell's diverticulum, a bulb-like dilatation of the proximal descending aorta, may coexist as the anomalous origin of the vessel (*Figure 3*).[7] Symptomatic patients and asymptomatic patients whose diameter of diverticulum exceeds 30 mm should be evaluated for surgery.[8] RAA accompanying ALSA and left-sided PDA are the most common subgroups of congenital vascular ring anomalies with an incidence of 0.04%.[9] Since the vascular ring surrounds the trachea and esophagus, it may cause compression symptoms such as dysphagia and dyspnoea at an early age. Intervention should be considered in all patients with symptomatic vascular rings. Clinical findings are related to the intensity of tracheal and esophageal compression. The standard of care for the treatment of a vascular ring is surgical ligation and division of all fibrous bands to mobilize the trachea and esophagus with or without resection of the diverticulum.[10]

Our patient had an association of RAA and ALSA originating from Kommerell's diverticulum that measured 22 mm at the widest diameter and left-sided PDA, all of which created a vascular ring. Due to the very low incidence of this condition and lack of dedicated guidelines, it is recommended to decide on the optimal management strategy depending on the anatomical and functional characteristics of the cardiovascular abnormalities and patient-specific comorbidities. Thus, after discussing the patient with the Heart Team as well as considering the patient's preferences and lack of compression symptoms, we decided to follow the RAA, ALSA, and Kommerell's diverticulum conservatively but to close the left-sided PDA percutaneously.[4] The complete regression of the patient's complaints during the follow-up shows that our treatment was beneficial. Total correction surgery was the other treatment option.

Conclusions

It may be challenging to distinguish whether the symptoms are due to PDA or the vascular ring. This patient, who did not have tracheal or esophageal compression symptoms, underwent the percutaneous PDA closure procedure, considering the symptoms of the left ventricular volume overload. The current case report highlights the importance of being aware of the potential concomitant defects and sheds light on the treatment strategy for rarely encountered vascular ring anomalies. Currently, as there is no general guideline regarding this circumstance, the treatment strategy should be individualized on a case-by-case basis.

Lead author biography

Levent Pay, MD, is a senior resident in cardiology. He has completed his medical degree from the Istanbul Faculty of Medicine. His research interests include electrophysiology and cardiac devices.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

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None declared.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report, including imaging and associated text has been obtained from the patient in line with COPE guidance.

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