Unusual drainage pattern of a supracardiac total anomalous pulmonary venous connection
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DESCRIPTION
An 8-year-old boy presented to us with exertional breathlessness with cyanosis. Cardiovascular examination findings were suggestive of atrial septal defect (ASD). Chest X-ray showed right ventricular hypertrophy and prominent right pulmonary artery with prominent superior vena cava (SVC) shadow (figure 1). A transthoracic echocardiogram confirmed a non-restrictive ASD flowing right to left with dilated right-sided chambers. Echocardiogram examination also revealed absence of pulmonary venous connections to left atrium suggestive of an associated total anomalous pulmonary venous connection (TAPVC; video 1). On suprasternal views, the pulmonary venous confluence was seen draining into the vertical vein and then into the SVC on the right side of the spine (video 2). A contrast CT was carried out with three-dimensional reconstruction, which showed the right and left pulmonary vein united at the vertical vein, which ascended on the right side of the spine and anterior to the pulmonary artery, to drain into the SVC (figures 2 and 3). Cardiac catheterisation was performed with a multipurpose catheter that was taken to the pulmonary venous confluence via SVC and vertical veins to demonstrate the venous drainage pattern (figure 4). Clinical investigations confirmed a non-obstructed supracardiac TAPVC with direct venous connections to left atrium suggestive of an associated total anomalous pulmonary venous connection (TAPVC; video 1). On suprasternal views, the pulmonary venous confluence was seen draining into the vertical vein and then into the SVC on the right side of the spine (video 2). A contrast CT was carried out with three-dimensional reconstruction, which showed the right and left pulmonary vein united at the vertical vein, which ascended on the right side of the spine and anterior to the pulmonary artery, to drain into the SVC (figures 2 and 3). Cardiac catheterisation was performed with a multipurpose catheter that was taken to the pulmonary venous confluence via SVC and vertical veins to demonstrate the venous drainage pattern (figure 4). Clinical investigations confirmed a non-obstructed supracardiac TAPVC with direct venous connections to left atrium suggestive of an associated total anomalous pulmonary venous connection (TAPVC; video 1). On suprasternal views, the pulmonary venous confluence was seen draining into the vertical vein and then into the SVC on the right side of the spine (video 2). A contrast CT was carried out with three-dimensional reconstruction, which showed the right and left pulmonary vein united at the vertical vein, which ascended on the right side of the spine and anterior to the pulmonary artery, to drain into the SVC (figures 2 and 3). Cardiac catheterisation was performed with a multipurpose catheter that was taken to the pulmonary venous confluence via SVC and vertical veins to demonstrate the venous drainage pattern (figure 4). Clinical investigations confirmed a non-obstructed supracardiac TAPVC with direct

Figure 1 Chest X-ray showing cardiomegaly, prominent right descending pulmonary artery with prominent superior vena cava shadow.

Video 1 Two-dimensional echocardiography apical four-chamber view showing right atrial and right ventricular dilation, absent pulmonary venous connections to the left atrium and a common venous chamber posterior to the left atrium.

Video 2 Two-dimensional echocardiography with colour Doppler, suprasternal view, showing a flow in the vertical vein draining directly into the superior vena cava.

Figure 2 CT of the thorax with three-dimensional reconstruction image showing right and left pulmonary veins forming venous confluence, which is draining into the vertical vein.
drainage of vertical veins to the SVC. The patient was sent for surgical correction under cardiopulmonary bypass. Incisions were made on the left atrium and pulmonary venous confluence, and direct anastomosis was performed. The patient postoperatively is acyanotic and functionally doing well.

Classically, in supracardiac TAPVC, the venous confluence drains into the SVC via the vertical and innominate veins. Very rarely, the vertical vein can drain directly into the SVC or azygous system of veins without draining into innominate veins. These drainage patterns are mostly obstructive in nature. In such cases, the classical ‘snowman’ appearance is not present on chest X-rays as the vertical vein traverses across the right side of the spine obscuring its shadow. In such cases, only the SVC shadow can be demonstrated in the X-ray, as seen in our index case (figure 1). Awareness of such uncommon drainage patterns is important as they pose a diagnostic and therapeutic challenge in diagnosis and treatment of such conditions.

Learning points

- Although most of the supracardiac total anomalous pulmonary venous connections (TAPVCs) drain via innominate veins, rare variants such as direct drainage of the vertical vein into the superior vena cava or azygous veins have also been reported.
- Most of these rare variants of supracardiac TAPVC are obstructive in nature.
- These variants are of particular importance during surgical repair because of the chances of injury to the right pulmonary artery and right bronchus because of its proximity.

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