Rev Port Cardiol. 2016;35(12):697.e1–697.e6

Revista Portuguesa de Cardiologia
Portuguese Journal of Cardiology
www.revportcardiol.org

CASE REPORT

Anomalous pulmonary venous connection: An underestimated entity

Sara P. Magalhães[a,*], Nuno Moreno[b], Marília Loureiro[c], Manuela França[a], Fernanda Reis[a], Sílvia Alvares[c], Manuel Ribeiro[a]

[a] *Serviço de Radiologia do Centro Hospitalar do Porto, Porto, Portugal*
[b] *Serviço de Cardiologia do Centro Hospitalar Tâmega e Sousa, Penafiel, Portugal*
[c] *Serviço de Cardiologia Pediátrica do Centro Hospitalar do Porto, Porto, Portugal*

Received 24 November 2015; accepted 16 January 2016
Available online 17 November 2016

KEYWORDS
Anomalous pulmonary venous connection;
Congenital anomaly;
Scimitar syndrome

Abstract Anomalous pulmonary venous connection is an uncommon congenital anomaly in which all (total form) or some (partial form) pulmonary veins drain into a systemic vein or into the right atrium rather than into the left atrium.

The authors present one case of total anomalous pulmonary venous connection and two cases of partial anomalous pulmonary venous connection, one of supracardiac drainage into the brachiocephalic vein, and the other of infracardiac anomalous venous drainage (scimitar syndrome).

Through the presentation of these cases, this article aims to review the main pulmonary venous developmental defects, highlighting the role of imaging techniques in the assessment of these anomalies.

PALAVRAS-CHAVE
Conexão anómala das veias pulmonares;
Anomalia congénita;
Síndrome de cimitarra

Conexão anómala das veias pulmonares, uma entidade subvalorizada

Resumo A conexão anómala das veias pulmonares é uma anomalia congénita rara na qual todas as veias pulmonares (forma total) ou algumas (forma parcial) drenam numa veia sistémica ou na aurícula direita, em vez da aurícula esquerda.

Os autores apresentam um caso de conexão anómala total das veias pulmonares e dois casos de retorno anómalo parcial das veias pulmonares, uma drenagem supracardíaca ao nivel da veia braquiocefálica e uma infracardíaca (síndrome da cimitarra).

* Corresponding author.
E-mail address: sarapintomagalhaes@sapo.pt (S.P. Magalhães).

http://dx.doi.org/10.1016/j.repc.2016.01.009

Através da apresentação destes casos, este artigo pretende fazer uma revisão dos principais defeitos do desenvolvimento venoso pulmonar e realçar a importância das técnicas de imagem na avaliação destas anomalias.

Introduction

Anomalous pulmonary venous connections are a specific group of congenital heart defects caused by the abnormal drainage of a part or the entire lung to a systemic vein or the right atrium. The estimated incidence is 2/100 000 births.[1] Most frequently only a single pulmonary vein is anomalous. However, more than one vein can have abnormal drainage, and rarely all the pulmonary venous vessels can connect to the right side of the heart, a condition known as total anomalous pulmonary venous connection (TAPVC).

We report the cases of three children with anomalous pulmonary venous connections: two of partial anomalous pulmonary venous connection (PAPVC) with an indolent course, and a neonate with TAPVC requiring urgent surgical intervention.

Case reports

Case 1 (partial anomalous venous return with infracardiac drainage)

We describe the case of a 14-year-old girl assessed for an incidental finding on chest X-ray (pulmonary asymmetry). The chest X-ray revealed right pulmonary hypoplasia as well as an anomalous pulmonary vein descending below the diaphragm creating a curved shape on the right side, the scimitar sign. Echocardiography showed dextrocardia with apex on the left, at least two pulmonary veins draining into the left atrium, intact atrial and ventricular septa, no dilation of the cardiac chambers, preserved global biventricular systolic function and no signs of pulmonary hypertension. Computed tomography (CT) confirmed and better characterized these imaging findings (Figure 1A). Magnetic resonance imaging (MRI) (Figure 1B), in addition to angiographic evaluation, was also important for excluding associated congenital heart disease, and for assessment of right ventricular (RV) systolic function and volume, as well as left-to-right shunting (Qp:Qs 1:2). Given that the patient was asymptomatic and there was no evidence of cardiac functional impairment, a conservative strategy was adopted.

Case report 2 (partial anomalous venous connection with supracardiac drainage)

A nine-year-old girl was referred to the pediatric cardiology department for a cardiac murmur and Turner syndrome.

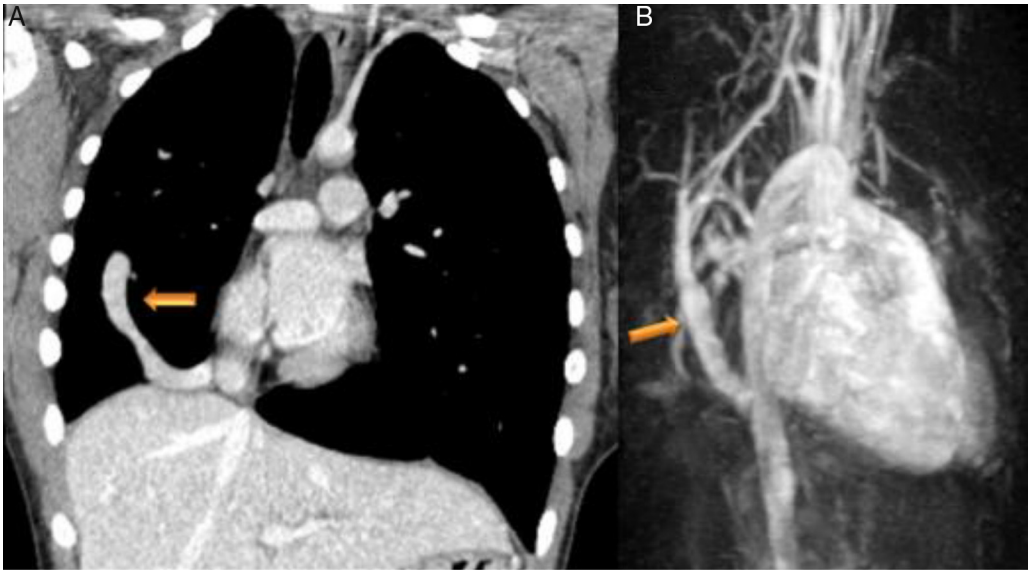

Figure 1 (A) Coronal-reformatted contrast-enhanced computed tomography showing the scimitar vein draining into the inferior vena cava (arrow) as well as right pulmonary hypoplasia with left lung expansion; (B) sagittal four-dimensional magnetic resonance imaging angiographic image also demonstrating the scimitar vein (orange arrow).