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Images of Interest / Imagens de Interesse

The Butterfly Sign in Berry Syndrome

O Sinal da Borboleta na Síndrome de Berry

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Abstract

We report a rare case of a female infant with respiratory distress and growth failure diagnosed with Berry syndrome. CT angiography demonstrated an interrupted aortic arch, distal aortopulmonary window, and the characteristic "butterfly sign." Early recognition of this rare but surgically correctable anomaly is essential for timely management and improved outcomes.

Keywords

Congenital cardiopathy; Berry syndrome; Butterfly sign.

Resumo

Relatamos um caso raro de uma lactente com dificuldade respiratória e má progressão ponderal diagnosticada com síndrome de Berry. A angiotomografia torácica revelou interrupção do arco aórtico, janela aortopulmonar distal e o característico "sinal da borboleta". O reconhecimento precoce desta anomalia rara é essencial para otimizar o prognóstico e orientar a intervenção cirúrgica precoce.

Palavras-chave

Cardiopatia congénita; Síndrome de Berry; Sinal da borboleta.

Case Presentation

A female infant developed progressive respiratory distress, easy fatigability, growth failure and recurrent respiratory infections from the age of six months. Initial evaluation at nine months of age by echocardiography suggested a major aortopulmonary communication.
Thoracic CT angiography (CTA) demonstrated cardiomegaly and a type A interrupted aortic arch. A distal aortopulmonary window (APW) was identified, with the right pulmonary artery arising from the ascending aorta and the left pulmonary artery arising from the main pulmonary artery. A patent ductus arteriosus was also present, continuing as the descending aorta. These findings are shown in figure 1.

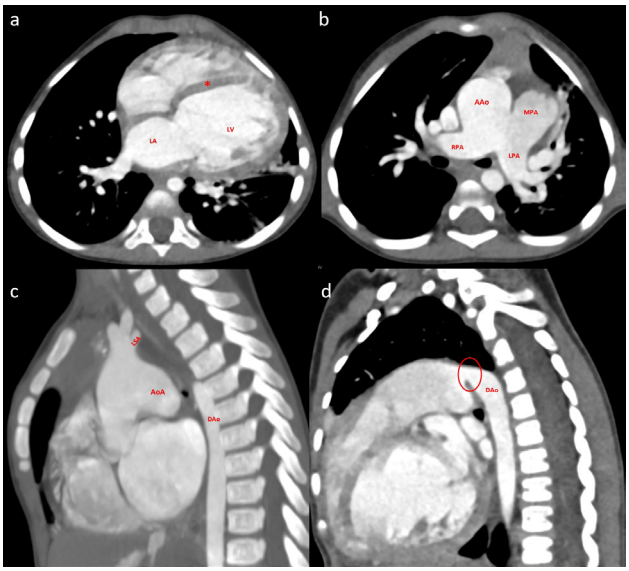

Figure 1 – Axial thoracic CT angiography scan (a) shows cardiomegaly with dilated left heart chambers (LA, left atrium; LV, left ventricle). An intact interventricular septum is present (*).(b) shows an aortopulmonary window with the right pulmonary artery (RPA) originating from the ascending aorta (AAo) and the left pulmonary artery (LPA) originating from the main pulmonary artery (MPA), forming the characteristic "butterfly" sign. Sagittal images demonstrate a type A interrupted aortic arch (AoA), between the left subclavian artery (LSA) and the descending aorta (DAo) (c), and a patent ductus arteriosus (circle, d) continuing as the descending aorta (DAo) (d). These findings were consistent with Berry syndrome.

Discussion

Berry syndrome, first described by Berry et al. in 1982,[1] is a rare congenital anomaly characterized by an aortopulmonary septal defect, anomalous origin of the right pulmonary artery from the aorta, intact ventricular septum, patent ductus arteriosus, and interruption or coarctation of the aortic arch.[1] Only a few cases have been reported since then.[2,3]
Most patients present in the neonatal period with severe pulmonary hypertension, resulting in respiratory symptoms.[2,3] Prenatal diagnosis is possible but challenging.[2] Postnatally, transthoracic echocardiography is the first-line modality and usually identifies the APW. Further imaging with CTA or magnetic resonance imaging is often needed to delineate the origin of the right pulmonary artery and aortic arch anatomy.[2] Berry syndrome comprises an interrupted aortic arch (IAA), APW, and intact ventricular septum.[1,2]

IAA is a complete discontinuity of the aortic arch and is classified as type A (between left subclavian artery and descending aorta), type B (between left common carotid and left subclavian arteries), or type C (between innominate and left common carotid artery).[3]

APW is a communication between the ascending aorta and main pulmonary artery with separate semilunar valves and is classified as type 1 (proximal), type 2 (distal), type 3 (total), or intermediate (like type 3 with well-formed rims).[3]

Our patient presented with a type 2 APW and type A IAA.

The left and right pulmonary arteries often arise separately - one from the main pulmonary artery and the other from the ascending aorta - producing the characteristic "butterfly sign", which has been primarily described on transthoracic echocardiography.[2] This finding should raise suspicion for Berry syndrome.

Surgical repair focuses on correcting the APW and IAA, ensuring unobstructed left ventricular outflow, restoring continuity of the aortic arch, and eliminating residual pulmonary shunting. Early recognition and timely intervention are essential for optimal outcomes.[1,3]

Conclusion

Berry syndrome is rare but surgically correctable. Recognition of the "butterfly sign" on imaging may facilitate early diagnosis and management.

Ethical Disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

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Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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