ACTA RADIOLÓGICA PORTUGUESA
May-August 2025 | Vol 37 nº 2 | 22-23
https://doi.org/10.25748/arp.40284

ARP Case Report nº 33: Pericardial Agenesis

Falta título inglês

Caso Clínico ARP nº 33: Agenesia do Pericárdio

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Received: 11/02/2025
Accepted: 12/02/2025
Published: 31/08/2025

A 57 year-old male patient presented to the Radiology department of our institution for outpatient evaluation of chest pain. He had a previous known history of smoking, peptic ulcer disease, arterial hypertension, type 2 diabetes mellitus, hypercholesterolemia and benign prostatic hyperplasia.
He had undergone surgical resection of a posterior left mediastinal mass, which was confirmed to be a bronchogenic cyst. He was additionally being followed for a bicuspid aortic valve with moderate to severe stenosis.
Chest XR (figure 1) was unremarkable for acute lung opacities or gross mediastinal changes. However, a closer look allows the identification of a hyperlucent band between the left inferior border of the heart and the left hemidiaphragm and a slight leftward shift of the right heart border that becomes obscured by the thoracic spine. The interposition of a hyperlucent area between the aortic knob and the pulmonary artery can also be seen.

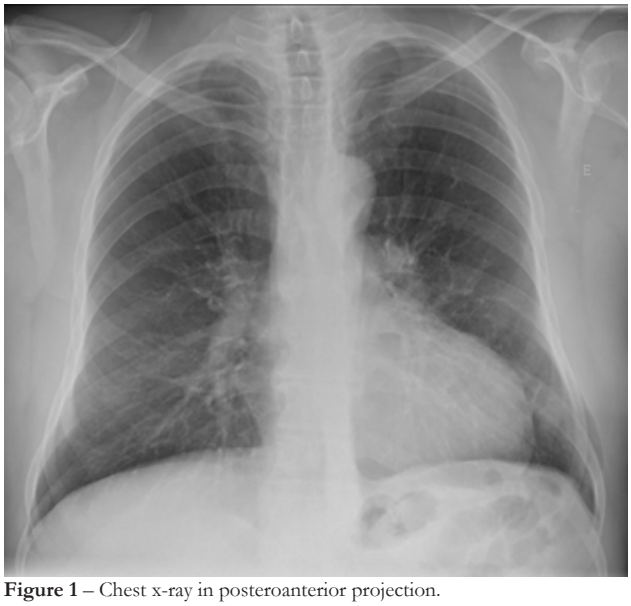
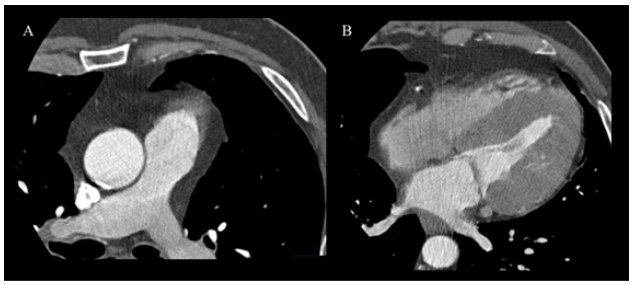


Figure 1 – Chest x-ray in posteroanterior projection.

The CT revealed the almost complete absence of the pericardium (note that in figure 2(b) a thin incomplete pericardial line is seen). Besides this finding, the already known bicuspid aortic valve was noted (not shown in the presented images), as well as left ventricle wall thickening (seen in figure 2(b)) due to left ventricle hypertrophy. There is also a slight leftward shift of the heart and the lung window image reveals interposition of the left lung tissue between the aorta and the main pulmonary artery (figure 2(c)).
Pericardial defects can be congenital or acquired (e.g. after pericardiectomy). Congenital pericardial agenesis (PA) is a rare entity, which can be partial or complete.[1] Its real prevalence is underestimated, since many patients remain asymptomatic, and it is often an incidental imaging finding.[1,2] There is a male predominance (male to female ratio of 3:1).[1,2] Up to 70% of cases involve the absence of the left pericardium.[1,2,3]

Affected patients may be asymptomatic or present with atypical chest pain or other nonspecific symptoms, often resembling other conditions such as acute coronary syndromes, cardiac aneurysms, myocarditis and pericarditis, among others.[2,4]
Complete PA is the least likely to cause complications.[2] Patients with partial PA can be more symptomatic and are at



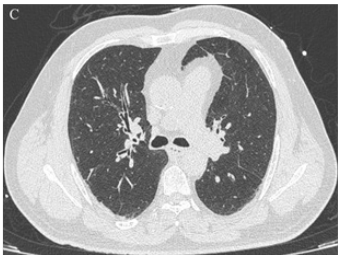


Figure 2 – Contrast-enhanced axial CT images at the level of the main pulmonary artery (a), showing the 4 cardiac chambers (b) and in lung window (c).

higher risk for complications, including herniation of heart chambers or atrial appendages through the pericardial defect, which can lead to cardiac strangulation or compression of the coronary arteries with subsequent ischemia and potential sudden cardiac death.[2,5]

There are known congenital anomalies associated with PA, including cardiac anomalies (e.g. atrial septal defects, patent ductus arteriosus, bicuspid aortic valve, mitral valve disease and tetralogy of Fallot) and non-cardiac anomalies (e.g. bronchogenic cyst, pulmonary sequestration, aberrant lung lobes, pectus excavatum and diaphragmatic hernia).[2,5] In the presented case, the patient had a bicuspid aortic valve and a history of previous surgery for a bronchogenic cyst.(Figure 3)

Typical electrocardiogram findings in PA include right-axis deviation with incomplete right bundle branch block and bradycardia.[2,3] Echocardiography is often non-diagnostic, but certain indirect findings should make clinicians suspect this diagnosis.[1,2,3,4]

On chest XR, the findings of PA are often subtle but may include: leftward shift of the heart with straightening and

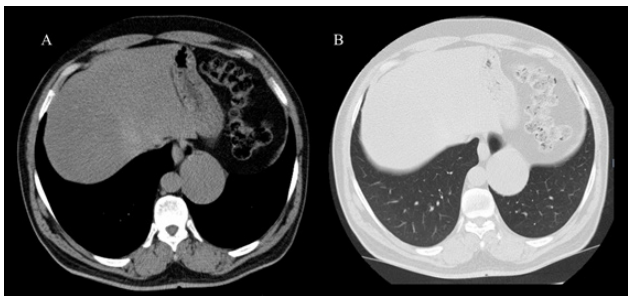


Figure 3 – Previous non-contrast-enhanced axial CT images show a hypodense posterior mediastinal lesion in soft tissue window (a) and in lung window (b), which was confirmed to be a bronchogenic cyst.

elongation of the left heart border ("Snoopy sign"); loss of the right heart border; prominent main pulmonary artery and a band of radiolucency caused by interposing lung tissue between the aorta and the main pulmonary artery; radiolucency separating the left ventricle and the left hemidiaphragm.[2,6] Cardiac CT and magnetic resonance (CMR) can aid in PA diagnosis by revealing the extent of the pericardial defect, the presence of herniated structures, potential concomitant congenital anomalies and additional specific findings such as interposition of lung tissue between the aorta and the main pulmonary artery, as well as interposed lung tissue between the diaphragm and the base of the heart.[2,4,6] CMR is considered the gold standard to identify PA, due to its high spatial resolution and superior tissue characterization. However, lack of visibility of the pericardium on CMR alone does not prove the absence of the pericardium.[2]

Surgery should be performed in symptomatic patients with partial PA or cardiac chamber herniation. For patients with complete PA, no treatment is needed unless mechanical complications occur.[1,2]

In summary, PA is a rare disorder which is often not clinically significant, but can be a cause of chest pain and may be life threatening if complications occur. The Radiologist should be aware of this entity and its findings, in order to provide an early diagnosis.

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.
Financing Support: This work has not received any contribution, grant or scholarship.
Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.
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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