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Case Report

Congenital Pericardial Agenesis presenting as non-specific chest pain: A Case Report[☆]

Sushil Rayamajhi, MBBS^{a,*}, Rekha Shrestha, MBBS^b, Kopila Shahi, MBBS^c,
Bibek Adhikari, MBBS, MD^d, Aditya Mahaseth, MBBS, MD, DM (Cardiology)^e

^a Department of Internal Medicine/Radiology, Swacon International Hospital, Battisputali, Kathmandu, Nepal

^b Department of Medicine, Universal College of Medical Sciences, Bhairahawa, Nepal

^c Department of Medicine, ZH Sikder Women's Medical College, Dhaka, Bangladesh

^d Department of Internal Medicine, Nepal Cancer Hospital and Research Center, Lalitpur, Nepal

^e Department of Cardiology, Shahid Gangalal National Heart Center, Janakpur, Nepal

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ABSTRACT

Congenital absence of pericardium is an exceedingly rare condition with a prevalence of 0.002%–0.004%. Due to its rarity and absence of association with any specific clinical examination finding, the diagnosis may be challenging. Due to the absence of symptoms and clinical awareness, pericardial agenesis is commonly misdiagnosed. It is important to consider this as a differential diagnosis of exertional chest pains. We report the case of a 9-year-old boy who attended our institution for non-specific but frequent symptoms and was diagnosed with congenital complete absence of the pericardium. The rarity of the condition can hinder timely diagnosis, as a physician or radiologist may never encounter a single case in their lifetime.

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Introduction

Congenital pericardial agenesis is an exceedingly rare condition with a prevalence of 0.002%–0.004% [1,2]. The pericardial sac's function is to secure and maintain the heart in the thoracic cavity, while the serous fluid's function is to lessen friction [3].

Congenital pericardial agenesis, which can be full or partial, is a rare and usually asymptomatic disorder. Due to its rarity and absence of association with any specific clinical examination finding, the diagnosis may be challenging [4]. Here, we present a case of a 9-year-old boy who attended our institute for non-specific chest pain and was diagnosed with complete pericardial agenesis.

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* Corresponding author.

E-mail address: dr.sushil.rayamajhi@gmail.com (S. Rayamajhi).

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Case report

A 9-year-old boy with no prior medical history suffered a chest pain for 6 months. He described chest pain as changing in intensity, exacerbated by exertion, and somewhat eased by analgesia. They had no connection to syncope or palpitations. He tried several analgesics with only partial relief of the symptom. He also reported having brief periods of dizziness without a spinning sensation and cold sweats. He never experienced syncope. He had no relevant medical history, cardiovascular risk factors, or family history of heart disease or sudden death.

On the physical examination, the vital signs (heart rate 72 bpm, blood pressure 104/65 mm Hg) were all in the normal range. In addition, femoral and pedal pulses were present, and there was no edema or signs of deep vein thrombosis in the lower limbs. He had several tests, including blood tests, Chest X-ray, and echocardiography. Blood tests were normal.

The electrocardiogram (ECG) revealed a normal sinus rhythm, a heart rate of 70 beats per minute, and diffuse T wave inversion in the anteroseptal leads. Chest X-ray revealed displacement of the cardiac silhouette to the left without tracheal deviation, obliteration of the right cardiac border overlapping the spine and leftward deviation of the heart and apex, and flattening and elongation of the left ventricular contour, that is, Snoopy sign (Fig. 1). The electrocardiogram showed sinus bradycardia, leftward deviation of the heart and apex, and T-wave inversion on the precordial and inferior leads. Echocardiography showed unusual acoustic windows, abnormal ventricular septal motion, exaggerated left ventricular movements, and abnormal swinging motion of the heart (Fig. 2). All these results confirmed the diagnosis of complete pericardial agenesis.

Therefore, we decided to give symptomatic treatment with analgesia. The patient was kept on clinical surveillance in our department. There were no complications, and the patient was discharged 8 days later. Three months later at follow-up, the patient required no analgesia and has had complete resolution of his chest pains. At this time, the patient is symptom free.

Discussion

The pericardium is an avascular sac that consists of 2 layers: an outer fibrous layer and an inner serosal layer. The pericardium stabilizes and maintains the position of the heart in the thorax by its ligamentous attachments.

Congenital absence of pericardium is an exceedingly rare condition with a prevalence of 0.002%–0.004% [1,2]. Left-sided defects are the most prevalent, accounting for 70% of all pericardial abnormalities [3]. Complete bilateral absence of the pericardium accounts for 9% of all abnormalities, while right-sided lesions account for 17% [3,4]. Thirty to 50% of patients with congenital absence of pericardium also have congenital abnormalities like atrial septal defect, patent ductus arteriosus, or tetralogy of Fallot. Patients with aortic connective tissue diseases and Marfan syn-

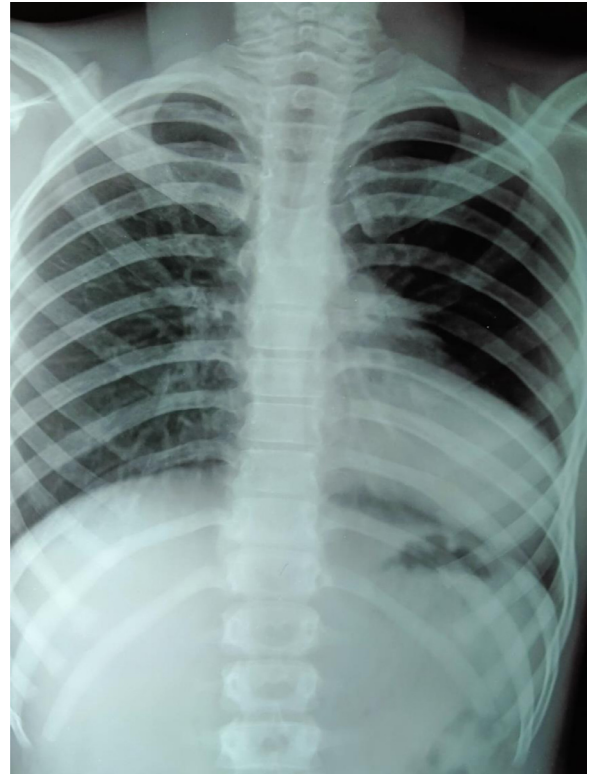


Fig. 1 – Chest X-ray showing displacement of the cardiac silhouette to the left, obliteration of the right cardiac border overlapping the spine, leftward deviation of the heart and apex, and flattening and elongation of the left ventricular contour (Snoopy sign).

drome have also been found to have pericardial anomalies [5,6].

Congenital absence of the pericardium is usually benign; however, it can be mistaken for other pathologic illnesses using standard imaging and screening techniques. Common symptoms include precordial pain of variable intensity, palpitations, dyspnea, syncope, or sudden death. Diagnosis is often difficult because the physical examination is usually non-specific [7].

CXR typically reveals strong levoposition of the cardiac silhouette, loss of the right heart border, a prominent pulmonary artery, and lung tissue between the diaphragm and inferior heart border [8]. Due to the leftward displacement of the heart, there is right ventricular predominance on echocardiography; as a result, the patient may be incorrectly diagnosed with right ventricular dilatation [9]. As in our case, the chest X-ray and echocardiogram were useful for the initial evaluation and confirmation of diagnosis. Cardiac magnetic resonance (CMR) is considered the gold standard imaging technique due to its superior soft tissue definition using spin-echo sequences synchronized with the cardiac cycle and its ability to detect focal myocardial infarctions [10]. In our case, CMR was not ordered as chest X-ray and echocardiography confirmed the diagnosis.

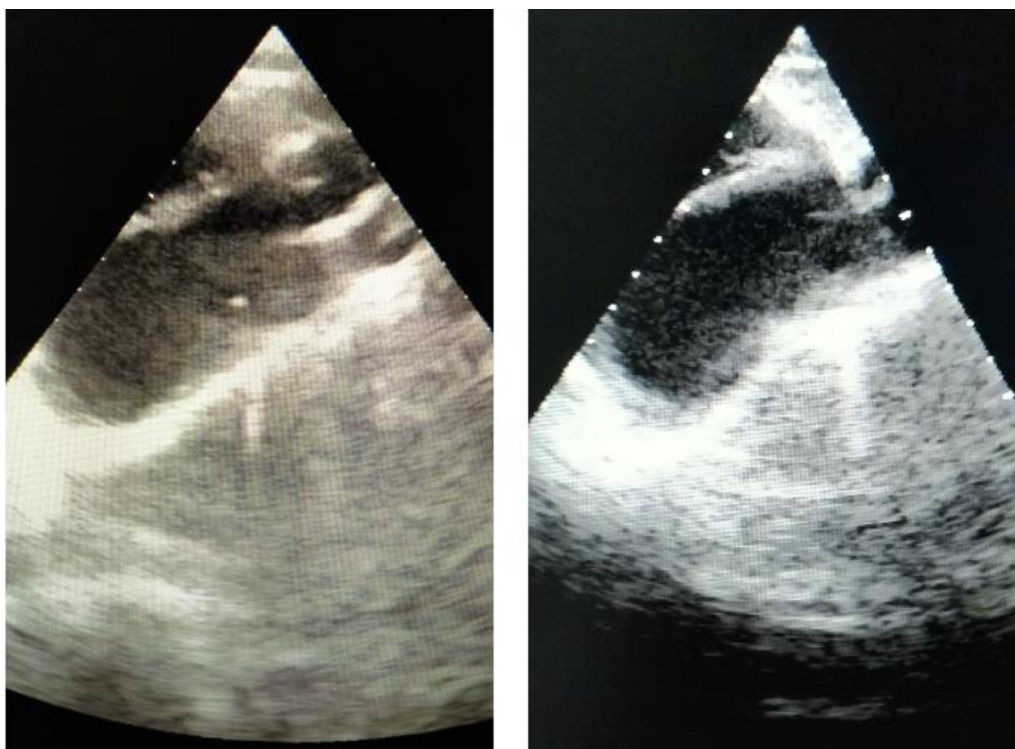


Fig. 2 – Echocardiography showed unusual acoustic windows, abnormal ventricular septal motion, exaggerated left ventricular movement and abnormal swinging motion of the heart.

In cases of total and complete pericardial unilateral agenesis, treatment is not necessary [5]. Symptomatic patients and asymptomatic individuals with evidence of ventricular strangulation on imaging technique should be surgically corrected. In our case of complete pericardial agenesis, we opted for analgesia and clinical monitoring in the cardiology department.

Conclusion

In conclusion, CAP is an uncommon abnormality that is typically asymptomatic. Due to the risk of complications and unexpected mortality in patients with partial abnormalities, it is essential to diagnose and characterize this entity adequately. The rarity of the condition can hinder timely diagnosis, as a physician or radiologist may never encounter a single case in their lifetime.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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