Isolated pericardial agenesis revealed by bradycardia and heart MRI in a healthy 5-year-old child

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SUMMARY: Bersani I, Rigante D, De Nisco A, Natale L, Lanza GA, De Rosa G, Stabile A, Delogu AB. Isolated pericardial agenesis revealed by bradycardia and heart MRI in a healthy 5-year-old child. Turk J Pediatr 2011; 53: 583-585.

We present a five-year-old boy with an unremarkable medical history who was incidentally found to have bradycardia and electrocardiographic signs of right axial deviation. Initial echocardiogram showed left displacement of the cardiac apex with slight enlargement of the right ventricle, while frontal chest radiograph showed a lucent area between the aorta and pulmonary artery. Cardiac magnetic resonance imaging finally revealed a partial left pericardial agenesis and abnormal displacement of the heart into the left hemithorax.

Key words: pericardial agenesis, magnetic resonance imaging, child.

Isolated pericardial agenesis (IPA) represents an uncommon clinical entity occurring in both complete and incomplete forms. Extremely few data are available in the medical literature describing IPA detection among pediatric patients[1].

Case Report

An asymptomatic previously healthy five-year-old boy was referred to our Institution because of colicky abdominal pain without fever, diarrhea, vomiting, or other substantial clinical signs. The medical history of the patient and his family was unremarkable. On the physical examination, palpation of the abdomen and peristalsis were normal, and no visceromegaly was noted. Abdominal pain spontaneously disappeared within 24 hours and no further complications occurred. Nevertheless, a heart rate (HR) of 60 bpm was detected with an innocent systolic heart murmur. The electrocardiogram confirmed marked sinus bradycardia (HR 56 bpm) combined with right QRS axis deviation, incomplete right bundle branch block pattern and mild nonspecific changes of cardiac repolarization. Echocardiography, carried out despite suboptimal windows, highlighted a marked leftward displacement of the heart, a posterior bulging of the cardiac apex, an apparent slight enlargement and hypertrophy of the right ventricle, and mild tricuspid regurgitation. Chest X-ray showed prominent II and III cardiac arches and a lucent area between the aorta and pulmonary artery (Fig. 1). Plasmatic troponin T was negative (<0.01 ng/ml; normal values <0.03 ng/ml), and all routine blood tests were unrevealing. Twenty-four-hour ECG Holter confirmed the prevalence of sinus bradycardia, low resting and mean HR and frequent phases of junctional escape rhythm (mean HR 72 bpm, minimal HR 40 bpm, maximal HR 153 bpm), though with adequate chronotropic response to exertion. Treadmill exercise stress testing showed a good cardiac response to exercise, without disorders of cardiac rhythm or ischemic changes. The presence of left-sided cardiac displacement was finally confirmed by cardiac magnetic resonance imaging (MRI), which revealed the prominence of the pulmonary trunk in the left hemithorax due to its herniation through a pericardial defect (Fig. 2). Considering the complete absence of any symptom, the good cardiac function and the normal results of the cardiovascular risk assessment, the child was treated conservatively with a program of regular cardiac follow-up.

Discussion

Congenital defects of the pericardium are very rare and are often unrecognized until

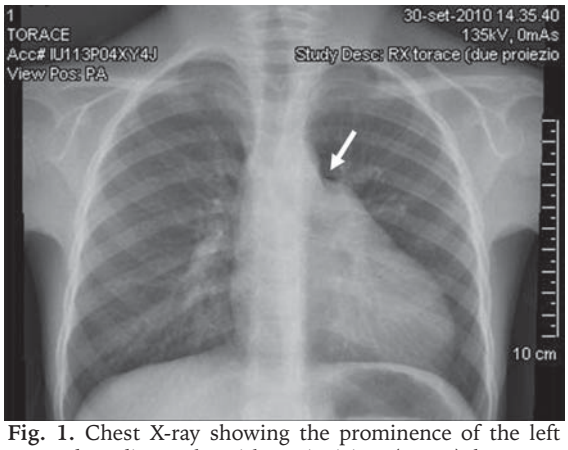

Fig. 1. Chest X-ray showing the prominence of the left second cardiac arch, with an incision (arrow) between the first and second arch, due to the pericardial defect.

adulthood. They include a large spectrum of abnormalities ranging from a small foramen in the pericardium to the complete absence of the entire pericardium[2], showing a male to female ratio of 3:1[3]. IPA mostly represents a completely asymptomatic condition accidentally discovered as an incidental finding on post-mortem examination, during intrathoracic interventions, or from abnormal chest X-ray films[4]. However, it might also be associated with chest pain, palpitations, dyspnea, dizziness, and syncope[5], owing to herniation or incarceration of the left atrial appendage through the defect, torsion of the great artery, or constriction of a coronary artery at the rim of the defect. Moreover, severe complications such as fatal myocardial strangulation, myocardial ischemia and sudden death have also been reported[6].

The pathogenesis of IPA is based on abnormal embryologic development of the pericardium resulting from a premature atrophy of the left duct of Cuvier. This implies a compromised vascular circulation to the left pleuropericardial membrane, which would eventually become the left pericardium. This theory is supported by post-mortem studies showing that congenital pericardial defects almost invariably involve the entire left side of the heart[7]. Detectable anomalies on chest X-rays often include displacement of the cardiac silhouette to the left, a lucent area between the aorta and pulmonary artery secondary to lung interposition, irregular left heart border, loss of the right heart border through its superimposition on the spine, prominence of the main pulmonary artery, and presence of lung tissue between the heart and diaphragm[8]. Common ECG records include right axis deviation, incomplete right bundle branch block, left displacement of the transition zone in the precordial leads, poor R wave progression, and prominent P waves[9]. Concerning echocardiography, the presence of IPA commonly requires a lateral probe position for the apical four-chamber view[10].

Although in our case, no further cardiac malformations were detected, according to the literature, about one-third of all IPA cases can be associated with other cardiac lesions, as patent arterial duct, mitral stenosis or tetralogy of Fallot[11]. Other possible echocardiographic findings include right ventricular dilation and paradoxical septal motion, usually associated with normal ventricular function. Further radiological investigations such as heart MRI are necessary to confirm the suspected diagnosis and determine the extent of the agenesis or to highlight the eventual evidence of herniation of the cardiac chambers through the defect. Typical findings detectable on cardiac MRI include heart levoposition, left atrial appendage/main pulmonary artery beyond the mediastinum, lung between the main pulmonary artery and aorta, lung between the heart and diaphragm, and elevated apex[12].

Most of the described cases of IPA deal with adult patients, and only a few descriptions exist in relationship with childhood. In our pediatric patient, presenting with spontaneously remitting abdominal pain unrelated to the IPA, the diagnosis was established following the investigation of bradycardia, which represented an occasional finding, since no coexisting symptoms were present on the child's arrival

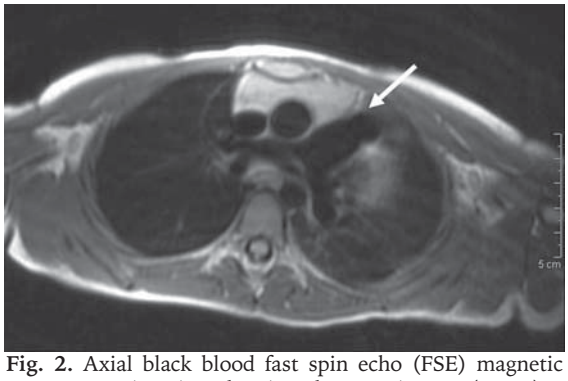

Fig. 2. Axial black blood fast spin echo (FSE) magnetic resonance imaging showing the prominence (arrow) of the pulmonary trunk in the left hemithorax due to herniation through the pericardial defect.

or had ever been detected in the past.

The therapeutic approach of IPA consists of different strategies, which should be adopted according to the large spectrum of clinical presentations. Surgical reconstruction of the absent pericardium may represent a safe intervention in case of debilitating symptoms and lead to symptomatic improvement; on the other hand, a conservative approach including only clinical follow-up is recommended in case of well-compensated clinical pictures and complete absence of symptoms.

In conclusion, our report is aimed both to underline that IPA can be diagnosed early in childhood, even in completely asymptomatic patients, and to familiarize clinicians and radiologists to this uncommon condition. In fact, although IPA may not significantly alter cardiac function, as in our case, the potential development of sudden and severe complications should always be carefully taken into consideration.

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