

Complete Absence of the Left Pericardium

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Abstract

Pericardial defect is a rare congenital abnormality, and most of the presenting cases are reported from intraoperative or post-mortem diagnosis. We report a case (46 yr-old male) with a 2-year history of vague chest pain and dry cough.

Chest roentgenography showed a mass in the supero-medial portion of the left lung without displacement of the heart. Computerized tomography supported the diagnosis of a cystic mass in the medial part at the lingula lobe of the left lung. Echocardiography was normal.

He was operated for symptomatic pulmonary mass and intraoperative findings were complete absence of the left pericardium and a bronchogenic cyst of the lingual (*Iranian Heart Journal* 2007; 8 (2): 56-58).

Key words: complete absence of pericardium ■ bronchogenic cyst ■ chest mass

Congenital pericardial defect is a rare abnormality and is often associated with other congenital heart and lung diseases.

The majority of cases are found either intraoperatively or at postmortem. The pericardial defect may be suspected initially from chest roentgenogram and then confirmed by echocardiography¹, computerized tomography or magnetic resonance imaging. We report a case of congenital pericardial defect accompanying bronchogenic cyst, which was discovered intraoperatively.

Case report

The patient was 46 yr-old male with a history of vague chest pain and dry cough for 2 years. He was a smoker (25 pack/years) as well. He did not give a history of have weight loss, and had no other complaint. His past medical history and familial history was negative.

On physical examination vital signs were stable, heart sounds were normal and there was no cardiac murmur. The apical impulse was palpable at the fifth intercostal space between the mid-clavicular and anterior axillary lines. Blood chemistry was normal. Chest roentgenography demonstrated a cystic mass in the medial part at the lingual lobe of the left lung (Fig. 1) and CT scan demonstrated the same findings (Fig. 2). Echocardiography was entirely normal. Left thoracotomy was done for resection of the lingula of the left lung. We noted a complete defect of the left pericardium and a cystic mass, most probably bronchogenic cyst. The cyst was resected without any intervention on the pericardium, because his symptoms were non-specific and there was no significant complication related to the defect.

We followed the patient postoperatively. Pathologic diagnosis of the mass was bronchogenic cyst.

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Fig. 1. Chest roentgenography demonstrated a cystic mass in the medial part at the lingula lobe of the left lung.

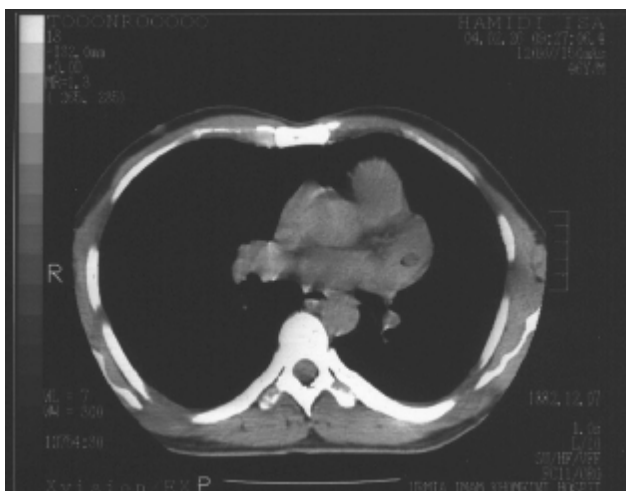
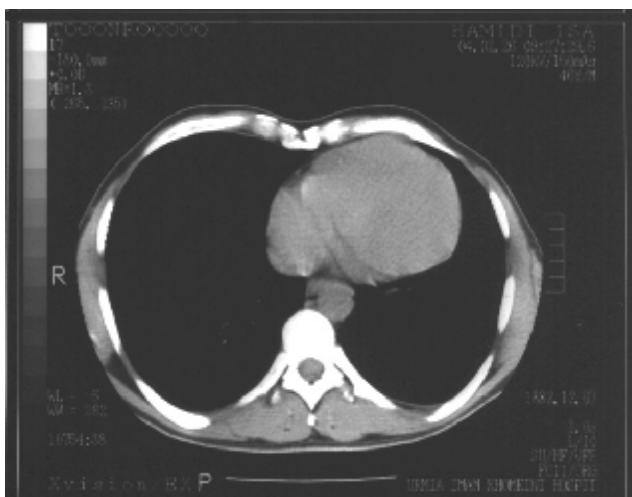


Fig. 2, A, B. CT scan demonstrated the same findings as in the CXR.

Discussion

Congenital defect of the pericardium is a rare abnormality and fewer than 200 cases have been reported in the world literature.²

The majority of presenting cases are reported from intraoperative or post-mortem diagnoses. Thus, it is not surprising that none of 15 pericardial defects seen within a 15 year period at the Mayo Clinic were diagnosed preoperatively.³

The incidence of pericardial defect in the surgical statistics is 1-4/1000. The ratio of incidence in males to females is 3:1 in about 30% of known cases. Other congenital cardiovascular and pulmonary anomalies were also present, such as atrial septal defect, tetralogy of Fallot, patent ductus arteriosus, mitral valve stenosis, tricuspid valve regurgitation, bronchogenic cyst, pulmonary sequestration, pectus excavatum and diaphragmatic hernia.⁴⁻⁶

The most widely accepted theory as to its pathogenesis suggests that premature atrophy of the left duct of curvier (common cardinal vein) leads to loss of blood supply to the left pleuro-pericardial membrane which, in adult life, forms the left pericardium.^{6,7} The right duct of curvier normally persists as the SVC, assuring adequate blood supply to the developing right pericardium. On the left side, complete absence of the pericardium is more common than a partial defect. Right-sided lesions and bilateral complete absence of the pericardium are extremely rare.³ Most patients are asymptomatic, but some may have non-specific chest pain, dyspnea, dizziness and occasionally syncope may occur.

The symptoms could be related either to herniation of a heart structure in the defects such as the atrial appendage or the left ventricle, torsion or strain on the great vessels, compression of the coronary artery or tricuspid valve regurgitation attributable to chordal rupture of the anterior leaflet by complete displacement of the heart into the left pleural space. Complete absence of the

pericardium is a benign condition, quite compatible with a normal life span.³

Physical findings are non-specific, including systolic murmur, lateral displacement of the apex and coexisting pectus excavatum. Electrocardiography may show incomplete right bundle branch block, right axis deviation or complete heart block. In patients with partial left pericardial defect, chest X-ray often shows a prominence of the hilum or the pulmonary artery. This is caused by herniation of the left atrial appendage through the defect. Complete absence of the left pericardium may be characterized by displacement of the heart and aortic knob, long prominence of the pulmonary artery and a flattened left cardiac border.^{1,8}

By leftward displacement, cross-sectional echocardiography can demonstrate right ventricular dilatation, paradoxical anterior motion of the ventricular septum in systole, vigorous left ventricular posterior wall motion and anterior displacement of the left ventricle during systole. CT may show an abnormal position of the heart in the thorax, no visible pericardium or the lung interposed between the main pulmonary artery and the ascending aorta. MRI can demonstrate similar findings.⁹ Moderate-sized defects are the ones most likely to produce symptoms because of their potential for herniation and cardiac strangulation. These complications must be considered absolute indications for surgical treatment with prophylactic closure of the defect.¹⁰

Many surgical procedures have been suggested, including primary closure, partial pericardiectomy, atrial appendectomy and pericardioplasty with partial pleural flaps or patches. Gortex, Dacron, bovine or porcine pericardium and fascia lata have been used as patches.

There is an important point in this case that imaging modalities such as echocardiography or CT didn't demonstrate the defect and it was diagnosed intraoperatively.

References

1. Yung NY, Wei HY, Mason SY. Partial absence of the left pericardium. *Acta Cardiol Sing* 2002; 18: 79.
2. Rusk RA, Kenny A. Congenital pericardial defect presenting as chest pain. *Heart* 1999; 81: 327-328.
3. Van SJ, Danielson GK, Schaff HV. Congenital partial and complete absence of the pericardium. *Mayo Clin Proc* 1993; 68: 743-747.
4. Skalski J, Wites M, Haponiuk I. A congenital defect of the pericardium. *Cardiovasc Sur* 1999; 47: 401-404.
5. Strenson SW. Congenital defects of pericardium. *Arch Intern Med* 1938; 61: 223.
6. Nasser WK. Congenital disease of pericardium. *Cardiovascular Clin* 1976; 7: 271.
7. Chang CH, Leigh T. Congenital partial defect of the pericardium associated with herniation of the left atrial appendage. *Am J Roentgen* 1961; 86: 517-522.
8. Nasser WK, Helmen C, Tavel ME. Congenital absence of the left pericardium. Clinical electrocardiographic, radiographic, hemodynamic and angiographic findings in six cases. *Circulation* 1970; 41: 469-477.
9. Fowler NO. Congenital defect of the pericardium. Its resemblance to pulmonary artery enlargement. *Circulation* 1962; 32: 114-120.
10. Finet G, Bozio A, Frieh JP. Herniation of the left atrial appendage through a congenital partial pericardial defect. *Eur Heart J* 1991; 12: 1148-1149.