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Mini Review

Congenital Pericardial Absence: A Review

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Abstract

Congenital absence of pericardium can vary from partial to complete. While partial absence of pericardium does not usually create much of difficulty, complete absence leads to strikingly mobile and markedly displaced heart. CT and MRI remain the main modalities of diagnosis and treatment basically remains symptomatic.

Keywords: Pericardium; Congenital Anomalies

Introduction

Pericardium is the covering of the heart with 2 distinct layers; outer parietal and inner visceral. The serous ultafiltrate of plasma-pericardial fluid is in between the two layers, about 20-30 ml in adults. Congenital absence of pericardium can vary from localised to complete absence [1-3]. The basic developmental pathology lies in the premature atrophy of duct of Cuvier, resulting in deficiency of blood supply to left pleuropericardial membrane [4,5].

The incidence of congenital absence of pericardium has been found to be 1/14,000 at necropsy [6]. Approximately two-thirds of it is partial absence [6-8]. One third of them are associated with other congenital anomalies [1]. Contact stress is exerted by outer parietal pericardium that contributes to diastolic pressure of ventricle and limits its acute dilatation [9]. The dimensions and pressures of the right sided chambers depends chiefly on pericardial constraint [9]. Hence, absence of parietal pericardium completely will lead to alterations in systemic venous return and increase in right ventricular size [9,10].

This abnormality usually comes to light when an abnormality is seen on routine chest x-ray which is generally taken of an asymptomatic patient or as a part of cardiovascular evaluation of a patient with coexisting heart disease [1]. Most common symptom is the chest pain which can vary from mild to debilitating [1]. It is left sided, stabbing, increased in left lateral decubitus position and relieved on standing [3]. The cause of pain in partial absence of pericardium has been attributed to the herniation of left atrial appendage through pericardial defect whereas in complete absence of pericardium the pain is believed to originate from torsion of thoracic inlet [11]. It can also have the symptoms of syncope, palpitations, dyspnea, dizziness and sudden death. Females are relatively three times protected than males.

Congenital partial absence of pericardium children can have abnormal facies, growth hormone deficiency or associated with VATER defects [12,13]. The arterial pulse and jugular venous pressures are normal in these patients. In complete absence of pericardium the heart is very mobile and this leads to the shifting of the heart to left dramatically along with rotation on its long axis; a feature which is not found in partial absence [3]. Torsion of thoracic inlet and great arteries can generate midsystolic murmurs in left lateral position in complete absence of pericardium.

Electrocardiogram is normal in partial absence of pericardium as there is no displacement of heart. The leftward position of the heart in complete absence of pericardium leads to delayed transition in the precordial leads. Rotation of the heart leads to deviation of the axis. Incomplete RBBB is also seen commonly [1]. Chest x-ray is of much more help than ECG in these cases. Herniation of left atrial appendage which is represented by a convexity immediately below the pulmonary trunk is seen in partial absence of pericardium. The close chest x-ray differentials of this condition are intra-pericardial congenital aneurysms and congenital pericardial cysts.

Complete absence of pericardium has dramatic mobility of heart which leads to leftward and posterior displacement of the heart. It can be associated with hypoplasia of left pulmonary artery and left lung too. Right of the vertebral column will not show any cardiac shadow [1].

Echocardiography will show the leftward position of the heart. The right ventricle and atrium are seen to be enlarged and there is paradoxical motion of the inter ventricular septum. It will help to detect the associated congenital malformations. CT scan and MRI are the main modalities of detection of these congenital pericardial absence conditions [14]. Management is mainly symptomatic.

Conclusion

Congenital absence of the pericardium can be both partial and complete. Congenital partial absence is mostly left sided which is evident on chest x-ray as a convex shadow in the location of left atrial appendage. Normally the patient is symptomatic or can present with chest pain which typically changes with position. Partial absence of the pericardium on the right side is very rare entity on the other hand. The other extreme of the spectrum shows complete absence of pericardium congenitally which presents itself in adulthood. The heart is very mobile and is shipped to the left and posterior in the scenario. Additional symptoms can be in the form of dyspnea, palpitations, syncope and sudden death. Definitive diagnosis is made by thoracic magnetic resonance imaging and computed tomography and the treatment basically is symptomatic.

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