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Images in Congenital Heart Disease

Hydropneumopericardium in endomyocardial fibrosis

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An 8-year-old boy, from Mozambique, was seen for severe endomyocardial fibrosis. Clinical examination revealed extensive ascites, in contrast with the absence of ankle œdema, jugular venous distention, and severe hepatomegaly. He had clear lung fields, rapid irregular muffled heart sounds, and a blood pressure of 90/60, without any paradoxical pulse. The chest x-ray (Fig. 1) showed extreme cardiomegaly, obscuring the lung fields, and echocardiography confirmed the presence of a massive pericardial effusion, as well as endomyocardial fibrosis. The cardiac structures were demonstrated better in the left parasternal, short axis view (Fig. 2) after pericardiocentesis. Endocardial fibrosis was more prominent on the right side, with tissue obliterating all but the infundibulum (inf) of the right ventricle, and non-coaptation of the tricuspid valve (tv), secondary to right ventricular involvement. A giant right atrium (RA), full of signals from spontaneous contrast, compressed the left atrium (LA), and the aorta (Ao) was seen in the centre of the short axis view. Chest radiography following pericardiocentesis (Fig. 3) showed a hydropneumopericardium (PP) as the result of defective aspiration during apical drainage allowed air to replace some of the fluid. The white arrows indicate the edge of the pericardium. This evolution proved favorable in this patient, without the need for additional aspiration. Severe diastolic dysfunction of the left ventricle resulting from extension of the fibrotic process to the left side of the heart, unfortunately, precluded construction of a bidirectional cavo-pulmonary connection.

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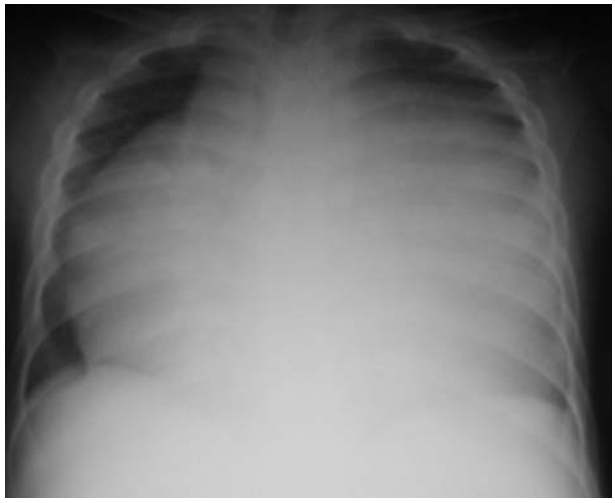

Figure 1.
The chest x-ray showed extreme cardiomegaly, obscuring the lung fields.

Endomyocardial fibrosis is a severe and progressively restrictive form of cardiomyopathy of unknown aetiology. Originally identified in Uganda by Davies in 1948,[1] it is more frequent in certain tropical areas and ethnic groups, where the disease contributes to one-fifth of deaths due to heart failure in the young. The disease is characterized by marked fibrotic endocardial thickening that predominates at the apex and ventricular inflow tracts, with incompetence of the respective atrioventricular valve. In addition, there is almost always a pericardial effusion, which can sometimes be excessive. Half a century after its discovery, aetiologic factors for the disease remain unclear. It seems to be a different entity from the hypereosinophilic syndrome, or Lœffler's disease, which are seen in industrialized countries, although

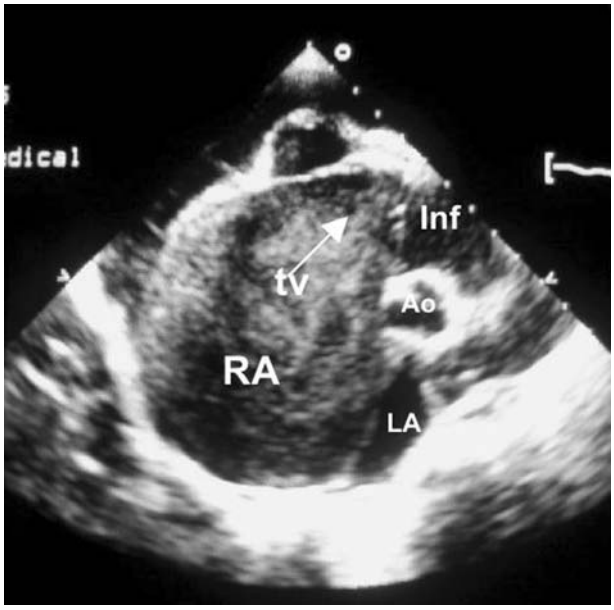

Figure 2.
The echocardiography (short axix view) demonstrated the severe right ventricle endomyocardial fibrosis, with the respect of the infundibulum.

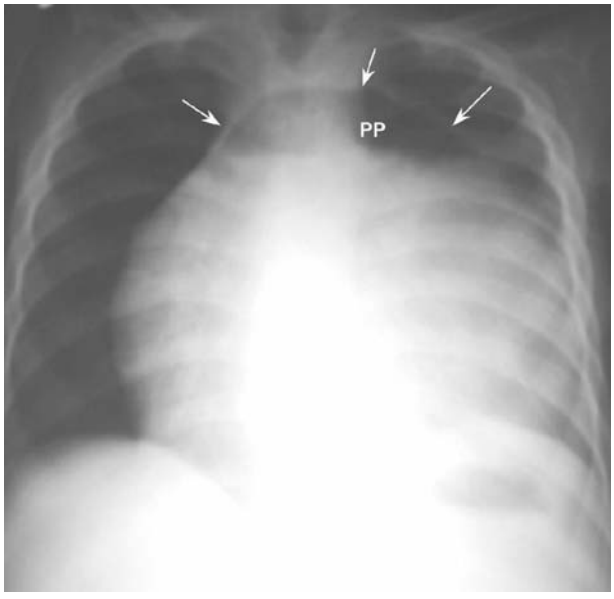

Figure 3.
After pericardiocentesis, a pneumopericardium was seen. The white arrows indicate the edge of pericardium.

toxicity of the eosinophils associated with environmental factors is one hypothesis favoured currently.

Hydropneumopericardium is a very rare condition. It may occur after pericarditis, pulmonary or digestive fistula, or medical procedures, such as pericardiocentesis or drainage. Usually it is well tolerated, but occasionally accompanying haemodynamic compromise necessitates further aspiration.

Reference

1. Davies JNP. Endomyocardial fibrosis. A heart disease of obscure aetiology in Africans. East Afr Med J 1948; 25: 10–14.

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