fluid. There were numerous firm, grey-white nodules 1 to 3 mm in diameter throughout both lungs. Many appeared to be associated with blood vessels. The pulmonary arteries were dilated and diffusely atheromatous. No emboli were noted. The right myocardial ventricle was hypertrophied. The spleen was surgically absent, and the splenorenal shunt was intact. The lobular architecture of the liver was accentuated, and yellow-white firm nodules of 0.6 to 1 cm in diameter were scattered throughout the parenchyma.

On microscopic examination, the nodular areas in the lung consisted of areas of arteritis surrounded by an exuberant fibroblastic reaction (Fig 3). There was extensive fibrinoid necrosis of small pulmonary arterioles, and arteriolar medial hypertrophy was present. Plexiform changes were present in many arterioles and intimal proliferation and fibrosis were prominent findings. Changes of chronic passive congestion were found in the liver, and numerous hepatocellular adenomas were found scattered through the hepatic parenchyma.

**DISCUSSION**

The pathologic alterations in the pulmonary arterial bed of patients with IPH are readily apparent on light microscopy but have not been reported to cause abnormal densities on chest roentgenogram. Pathologic alterations include pulmonary arteriolar medial hypertrophy, intimal proliferation and fibrosis, plexiform lesions, dilatation lesions, and necrotizing arteritis. Although the pathogenesis of these vascular alterations has not been firmly established, Wagenvoort and Wagenvoort\(^4\) have suggested that they occur secondary to intense pulmonary vasoconstriction.

Our patient had all of the pulmonary vascular changes mentioned above, and in addition, had an exuberant fibroblastic reaction around numerous pulmonary arteries. This kind of fibroblastic reaction has not been previously reported in patients with IPH and was probably a focal reaction to her necrotizing pulmonary arteritis. Since no other pathologic changes were found in the lungs, we believe that the nodular pattern of densities on this patient’s chest roentgenogram resulted from these 1 to 3 mm areas of fibroblastic reaction around the pulmonary arteries. Although it has been demonstrated that isolated nodular densities 3 mm in diameter are at the lower limit of size required to cause roentgenographically visible densities, multiple nodular densities as small as 1 mm in diameter may be seen as superimposition shadows.\(^6\) It is likely that most, if not all, of the nodular densities seen on our patient’s chest roentgenogram represent such superimposition shadows.

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**Restrictive Cardiomyopathy in Pseudoxanthoma Elasticum**

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A unique case of severe and diffuse endocardial fibroelastosis covering the entire left ventricle and associated with pseudoxanthoma elasticum is presented. The clinical picture was that of an apparently primary restrictive cardiomyopathy. Specific histologic lesions could be recognized in the endocardium.

Pseudoxanthoma elasticum (PXE) heart disease has not been clearly characterized since autopsy reports are extremely rare and hemodynamic data are not available. Two categories of cardiac involvement have been

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described in the 15 postmortem cases of the literature:1-3 early coronary disease, due to fibrous intimal changes, whose clinical significance has been recently confirmed;4 and fibrous thickening of the endocardium of the atria, the ventricles and/or the atrioventricular valves, usually of limited extension and apparently devoid of clinical relevance, except in four patients suffering from heart failure, which was attributed to mitral stenosis or mitral disease in three and to the endocardial lesions involving localized areas of the left ventricle in the case of Huang et al.3

The following case illustrates that PXE endocardial disease may be most severe and diffuse, covering the whole left ventricular cavity and resulting in a clinical, hemodynamic and anatomic picture entirely similar to the other forms of primary restrictive cardiomyopathies.4

CASE REPORT

A 39-year-old woman was admitted to the hospital because of congestive heart failure and atypical pain of two years' duration. Her mother died of heart disease at the age of 63. On examination, the apical impulse and the first heart sound were normal. There was a grade 2 apical systolic murmur and a soft opening snap followed by a short middiastolic rumble. Blood pressure was 120/80 mm Hg. The chest roentgenogram showed a typical mitral silhouette with a normal left ventricle. The ECG showed atrial fibrillation and a left intraventricular conduction defect. Funduscopic examination revealed a peripapillary zone of choroidal sclerosis with atrophy and pigmentation that was interpreted as residual from an old chorioiditis. Visual acuity was normal in both eyes. The values for blood and urinalysis were within normal limits.

Heart surgery was indicated but the mitral valve was not found stenotic, and there was only moderate regurgitation. Left atrial mean pressure was 36 mm Hg and the left ventricular pressure, 110/6 to 34 mm Hg. A definite middiastolic plateau was apparent in the left ventricular pressure tracing during the longer pauses, and the mitral diastolic gradient was negligible. The patient was controlled periodically in the outpatient clinic, and two years after surgery, slight yellowish reticulated lesions symmetrically distributed on either side of the neck and shoulders were first recognized. The biopsy specimen from this area revealed the typical findings of PXE. A year later she died of heart failure.

Autopsy findings

An extensive porcelain-like thickening of the endocardium covered the small cavity of the left ventricle, burying the trabecular and papillary muscles (Fig 1A and B). The mitral valve appeared diffusely thickened but without fusion of the commissures (Fig 1A). The left atrium was also involved showing a remarkable jet lesion consisting of elevated, whitish plaques heavily calcified and with a roughened surface.

Microscopically, the endocardial thickening was composed of fibrous tissue with a variable pattern of layered elastic proliferation that appeared scattered throughout the thickness of the endocardium or predominantly condensed in the deeper layers. Less frequently, the thickening consisted only of hyaline fibrous tissue. In some sections, the elastic fibers appeared fragmented, curled and with granular degeneration, surrounded by the large multinucleated cells of a granulomatous foreign body reaction (Fig 2). Hyperplasia and hyper-

Fig 1. Macroscopic findings. A (top), opened left-sided heart cavities showing severe endocardial fibrosis covering entire left ventricle. Small nodular fibrous deposits are present in line of closure of mitral valve. B (bottom), section of the left ventricle showing small cavity, thick endocardium, and involvement of papillary muscles.

DISCUSSION

Our case provides anatomic and functional evidence that the heart failure in PXE may be due to a genuine obliterative or restrictive disease, analogous to the more
severe cases of Loeffler's endocarditis or Davies' endomyocardial fibrosis. The marked elevation of the end diastolic pressure in the presence of a small left ventricular cavity reflects the restriction of the filling imposed by the endocardial lining. The mitral regurgitation has been considered a distinctive feature of the endocardial diseases and is accounted for by the fibrosis of the valve or the papillary muscle dysfunction.

It is notable that the cardiac manifestations were the presenting and main aspect of the syndrome, suggesting that the PXE endocardial disease may be easily misinterpreted as a primary cardiomyopathy. It is well known that the cutaneous and eye lesions may not be apparent or specific enough to suggest the diagnosis. The features pointing to the nature of the endocardial fibroelastosis were the degenerative changes of the elastin which have been recognized as characteristic of PXE, although granular degeneration has also been observed in adult cases of apparently primary fibroelastosis. The granulomatous foreign body reaction described in the endocardium has been previously documented in advanced skin lesions. The hypertrophy of the smooth muscle cells is a nonspecific finding.

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