ENDOCARDIAL FIBROELASTOSIS*

Report of a Case

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Endocardial fibroelastosis, a pathologist’s term primarily, was suggested by Weinberg and Himmelfarb in 1943. They described a cardiac condition of obscure aetiology in infancy in which the most striking feature was diffuse left ventricular endocardial thickening. The condition was first recognised by Kreysig in 1826 and designated ‘endocarditis foetalis’. The term endocardial fibroelastosis is largely ambiguous especially because the endocardial thickening may occur in isolated patches alone or may line an entire cardiac chamber. The endocardial fibroelastosis is most often secondary to other cardiac anomalies including aortic stenosis, mitral stenosis, premature closure of the foramen ovale, ventricular septal defect, coarctation of aorta and rudimentary left ventricle (Anderson and Kelly 1956). Far less commonly, the thickening occurs as an isolated pathological finding associated with considerable hypertrophy of the heart. Moller et al. (1964) described 47 cases and called the second group the ‘primary’ type. Manning et al. (1964) described the ‘primary’ type in infants below one year of age. Clinically, these infants presented as feeding problems, failure to thrive and laboured breathing and tachycardia, which resulted from the congestive cardiac failure. They had signs of congestive cardiac failure usually before 6 months, associated with cardiomegaly and an E.C.G. pattern of left ventricular hypertrophy unaccompanied by organic murmurs or cyanosis. The so called ‘primary’ type can most often be seen in newborns especially localised to the left chambers of the heart, but at times, also involving the right side and the various valves. The exact frequency of the ‘primary’ type is not known although such cases are being reported with increasing frequency specially from the United States and Europe. Thomas et al. (1954) described 20 cases of endocardial fibroelastosis in an autopsy series of 10,000 adults and children. Out of these, 8 cases belonged to the ‘infantile’ and ‘childhood’ types with obscure aetiology. Fisher (1962) found an incidence of 1.3 per cent in children below the age of 14 years. Graham (1964) reviewed 14 cases collected in 10 years at the Hospital for Sick Children, Great Ormond Street, London, and out of these only 4 cases were unassociated with other cardiac anomalies i.e., they would fall into the

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primary" type. The rarity of the condition provoked description of a case recently studied in this department.

**Report of a Case**

A one and a half year old female child was admitted for low grade fever, respiratory distress and vomiting accompanied by swelling of the feet of 7 days' duration. At admission she had bilateral pedal oedema and engorged neck veins. She had laboured breathing and central cyanosis. Her pulse was 164 per minute, regular, respiration 45 per minute and temperature 100.2°F. The liver was enlarged 3 cm. below the costal margin in the midclavicular line and was soft. Signs of consolidation of the lower lobe of the left lung were present. An X-ray of the chest taken at this time revealed marked cardiomegaly as the only positive finding. The family history was non-contributory. The natal and prenatal developments were uneventful. She was treated with broad-spectrum antibiotics, oxygen and digitalis. Despite these measures the patient died 2 hours after admission. The final clinical diagnosis was congestive cardiac failure with consolidation of the lower lobe of the left lung.

**Autopsy Findings.** Autopsy revealed a central cyanosis and pedal oedema. The heart, which showed the most significant findings, weighed 100 gm. The left ventricle was dilated and hypertrophied, its wall measuring 0.9 cm. in thickness. The right ventricular wall measured 0.2 cm. in thickness. The endocardium of the left ventricle was markedly thickened, pearly white, smooth and glistening all over, the thickening being pronounced along the outflow tract (Fig. 1). The endocardium in the other chambers appeared grossly normal. Congenital anomalies, mural thrombi or ischemic scars were not seen. Histologically, the changes were confined mainly to the left ventricle. The endocardium was markedly thickened, being readily identifiable as a separate layer (Fig. 2). It was collagenised with extensions into the myocardial sinusoids and at places was seen forming thick 'slings' around the blood vessels. In other places the endocardial extensions were seen enclosing small groups of myocardial bundles. Stains for elastic tissue and collagen revealed many layers of thin, fibrillary elastic fibres running parallel to the surface with varying amounts of collagen in between (Fig. 3). Blood vessels specially near the endocardium were thickened with intimal proliferation and narrowing of the lumina. A small organised thrombus was seen in one section. The subendocardial lymphatics were dilated. The myocardium was hypertrophied and in the sub-endocardial area mild degree of myocardiolysis and fragmentation of the fibres was seen. The interstitium was loose, oedematous and only at places showed collagenisation. There was no evidence of parenchymal or interstitial inflammation. The coronary vessels, the epicardium and the pericardium were unremarkable. The lower lobe of the left lung revealed focal pneumonitis with patchy atelectasis. The only other positive histological finding was a mild fatty metamorphosis of the liver.

**Discussion**

The present case anatomically falls into the group of endocardial fibro-