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IMAGES IN PAEDIATRICS

doi: 10.1136/adc.2006.103093

Idiopathic infantile arterial calcification presenting as fatal hypertensive cardiomyopathy

A 5-month-old girl was admitted to our paediatric intensive care unit with cardiorespiratory failure and hypertension. Chest radiography showed cardiomegaly and pulmonary plethora. Electrocardiography showed left ventricular hypertrophy and strain pattern. Echocardiography showed a dilated and hypertrophied left ventricle, with bright and hyper-reflective myocardium (fig 1). Fractional shortening was 12.5%. The concentration of creatine kinase was 493 U/l (normal range 20–190) and that of troponin I was 3.1 µg/l (normal range <0.03).

The patient initially responded to treatment but collapsed and died 18 h later.

Postmortem examination showed left ventricular hypertrophy with no outflow obstruction or coarctation. The coronary arteries had a normal origin and distribution but were thick walled and diffusely narrowed (fig 2). Cutting into the vessels was difficult. The thoracic and abdominal aorta and great vessels

were also diffusely thickened. Both renal arteries were thickened and completely occluded. There were no thrombi.

Histological examination showed a deposition of calcium in the internal elastic lamina of the coronary and renal vessels, with fibroproliferative changes in the intima, resulting in occlusion (fig 3). Appearances were typical of those seen in idiopathic calcification of infancy, a genetic disorder resulting from homozygotic or compound heterozygotic nonsense mutations in ectonucleotide pyrophosphatase phosphodiesterase-1.¹ This causes calcification of the arterial internal elastic lamina, resulting in occlusive coronary artery disease. Calcification in a peripheral artery with electrocardiographic changes of occlusive coronary artery disease suggests the diagnosis.² Many patients die from myocardial infarction in the first 6 months of life. Hypertension with cardiac failure is a common mode of presentation.³

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Competing interests: None declared.

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Figure 1 Parasternal long-axis echocardiogram showing hypertrophied and dilated left ventricle. Ao, aorta; LA, left atrium; LV, left ventricle.

Figure 2 Gross pathology of the left ventricle (LV), normal aortic outlet (Ao) and thick-walled coronary artery with narrow lumen (arrow).

Figure 3 Microscopic pathology of the left coronary artery showing deposition of calcium in the internal elastic lamina and fibroproliferative changes in the intima.