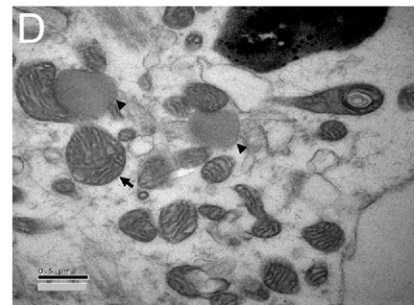
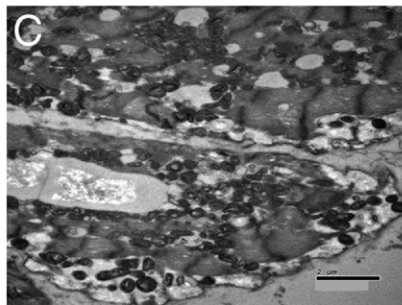
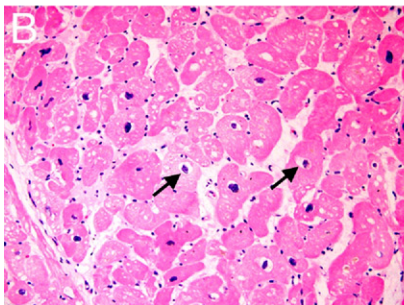
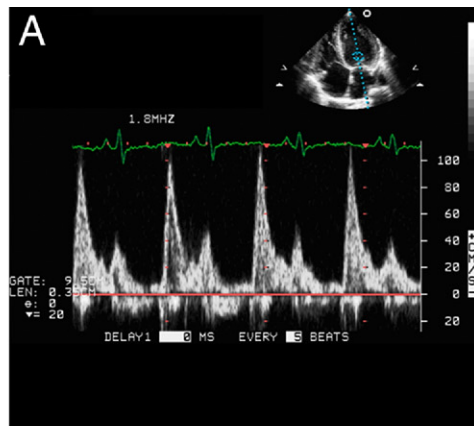


IMAGES IN CARDIOLOGY

Mitochondrial Cardiomyopathy Presenting as Hypertrophic Cardiomyopathy With Advanced Chronic Kidney Disease

Li Ching Lee, MBBS,* Kong Bing Tan, MBBS,† Raymond Ching-Chiew Wong, MBBS*
Singapore



From the *Cardiac Department, National University Heart Centre, National University Hospital, and the †Department of Pathology, National University of Singapore.
Manuscript received September 16, 2009, accepted October 6, 2009.

A 57-year-old man presented with syncope. Clinical examination showed a blood pressure of 145/80 mm Hg and normal neurological functions, but he was in mild heart failure. Laboratory testing revealed marked renal dysfunction: serum creatinine of 262 $\mu\text{mol/l}$ and estimated glomerular filtration rate of 23 ml/min/1.73 m². Echocardiography revealed severe biventricular systolic dysfunction, profound concentric left ventricular hypertrophy, and a restrictive filling pattern (A, Online Videos 1, 2, and 3). Coronary angiography excluded significant epicardial disease. Renal ultrasonography was consistent with chronic parenchymal disease. Bone marrow and abdominal fat pad aspirates were stained negative for myeloma and amyloidosis. Endomyocardial biopsy was later performed. Light microscopy showed cardiac myocytes with marked structural distortion, interstitial fibrosis, and enlarged nuclei with perinuclear cytoplasmic vacuolation (B, arrow). Electron microscopy revealed marked increase of mitochondria that were polymorphic with abnormal cristae pattern (C and D, arrow). Lipid-containing vacuoles were closely associated (arrowheads). The final diagnosis was mitochondrial cardiomyopathy associated with advanced chronic kidney disease. He responded well to antifaailure treatment but declined genetic study.