A 46-year-old man with a clinical diagnosis of dilated phase of hypertrophic cardiomyopathy was followed up at our hospital for 26 years. His refractory congestive heart failure gradually worsened over several years, and he was unresponsive to conventional medications. He then underwent left ventriculoplasty. Biochemical and genetic investigations of the left ventricular myocardium revealed a novel point mutation in the mitochondrial DNA.

Scanning electron microscopy can directly observe both the 3-dimensional structure of a cell with high resolution and the 3-dimensional structure of the membrane system using the osmium-dimethyl sulfoxide-osmium method to remove the cytoplasmic matrix. Conventional transmission electron microscopy detects the fine structure of the membranous components of mitochondria. We used these methods to examine mitochondria in the heart of the present patient. The myocardium excised from the left ventricular anterolateral wall during the operation was processed and observed under a Hitachi S-5000 scanning electron microscope and a Hitachi H-7100 transmission electron microscope.

The endomyocardial biopsy sample with normal histology, obtained from the left ventricle of a patient with suspected idiopathic dilated cardiomyopathy, was used for comparison. Similar-sized normal mitochondria lined up regularly (Figure 1). In contrast, abnormal features of mitochondria were observed in cardiomyocytes of this patient. Several mitochondria were deformed into various shapes, particularly spherical, oval, and enlarged forms, often with concentric circular cristae. Giant mitochondria were intermixed with small mitochondria, which proliferated under the sarcolemma or between

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myofibrils in the cardiomyocytes. Some giant mitochondria appeared to be formed by the fusion of adjacent mitochondria with concentric cristae (Figure 2). Giant mitochondria occasionally showed extensive tubular and lamellar undulating cristae (Figure 3). Transmission electron microscopy also suggested the fusion of enlarged mitochondria with adjacent normal-sized mitochondria (Figure 4). The pathogenic mechanisms of the formation of giant mitochondria remain unclear. However, the giant mitochondria in this patient might reflect mechanisms whereby mitochondria compensate for functional deterioration caused by mutation of mitochondrial DNA.3,4

Disclosures

None.

References


Giant Mitochondria in the Myocardium of a Patient With Mitochondrial Cardiomyopathy: Transmission and 3-Dimensional Scanning Electron Microscopy

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