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Atrio-His (James) accessory pathway and infiltration of $\gamma\delta$ T cells in the heart with MELAS

Gen Matsuzaki,¹ Akifumi Kushiya,² Kenta Uto,^{3,4} Morio Shoda,³ Yoshinori Seko¹

¹Division of Cardiovascular Medicine, Institute for Adult Diseases, Asahi Life Foundation, Tokyo, Japan;

²Division of Diabetes, Institute for Adult Diseases, Asahi Life Foundation, Tokyo, Japan;

³Department of Cardiology, Tokyo Women's Medical University, Tokyo, Japan;

⁴Department of Pathology, Tokyo Women's Medical University, Tokyo, Japan

Correspondence to Yoshinori Seko, sekoyosh-ty@umin.ac.jp

DESCRIPTION

A 40-year-old female patient, who was found to have diabetes mellitus during her first pregnancy, and then developed difficulty in hearing, was diagnosed as suffering from mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes (MELAS) due to mutation of mitochondrial DNA (A3243G). The patient complained of palpitation and faintness; ECG demonstrated short PQ interval with normal QRS width and significant ST-segment depression in II, III, aVF and V4-6 (figure 1). Echocardiogram and coronary CT angiogram showed normal findings. Then, electrophysiological study and myocardial biopsy were performed.

Both A-H and H-V intervals were prolonged from 27 and 16 ms during coronary sinus pacing (basic cycle length S1S1=500 ms) to 92 and 49 ms as a premature stimulus (S1S2=300 ms) was given (figure 2). This strongly suggested that normal AV-junctional conduction was temporarily restored during the refractory period of the atrio-His accessory pathway. The prevalence of WPW syndrome among patients with MELAS was reported to be much higher than in the normal population.¹ However, the atrio-His accessory pathway (James fibre) in a patient with MELAS has never been reported in the literature.

Immunohistochemical analyses of the myocardial specimen were done for CD4 (clone 1F6, Nichirei,

Figure 1 12-Lead ECG demonstrating short PQ intervals with normal QRS width and significant ST-segment depression in II, III, aVF and V4-6.

Figure 2 His bundle electrocardiogram (HBE) 1,2 showed A-H and H-V intervals were prolonged from 27 and 16 ms (basic cycle length S1S1=500 ms) to 92 and 49 ms as a premature stimulus (S1S2=300 ms) was given.

Figure 3 Immunohistochemical analyses of the myocardial tissue. (A) There was mild infiltration of $\gamma\delta$ T cells (immunofluorescent staining). (B) The infiltrating $\gamma\delta$ T cells clearly expressed a cytolytic factor perforin (black arrows) and the cardiac myocytes adjacent to the $\gamma\delta$ T cells (red arrows) appeared to have undergone necrosis. (C) TUNEL staining showed that almost no cardiac myocytes underwent apoptosis.

Tokyo, Japan), CD8 (clone DK25, DAKO), CD14 (clone LeuM3, BD), CD16 (clone 3G8, BD Endogen, Woburn, Massachusetts, USA), T cell receptor $\gamma\delta$ (clone 5A6. E9, Endogen), perforin (clone P1-8)² and TUNEL staining (In situ Apoptosis Detection Kit, TaKaRa Bio Inc., Otsu, Japan). Haematoxylin and eosin staining showed mild degeneration of cardiac myocytes with mild fibrosis. There was mild infiltration of $\gamma\delta$ T cells (figure 3A),

a few macrophages, almost no T helper cells, cytotoxic T lymphocytes and natural killer cells. The infiltrating $\gamma\delta$ T cells clearly expressed a cytolytic factor perforin (figure 3B, black arrows) and the adjacent cardiac myocytes (figure 3B, red arrowheads) appeared to have undergone degeneration. However, TUNEL staining revealed that almost no cardiac myocytes underwent apoptosis (figure 3C), suggesting that myocardial injury was induced

by some autoimmune mechanism rather than the energy production disorder.

Competing interests None.

Patient consent Obtained.

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