CARDIOVASCULAR FLASHLIGHT

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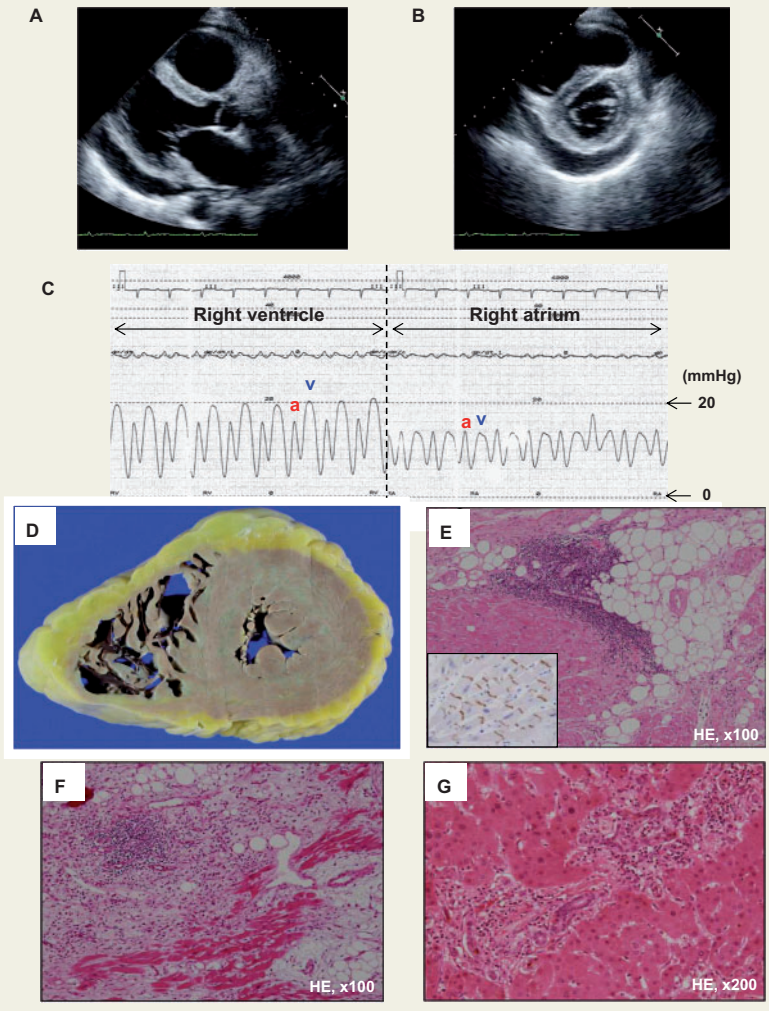
# Lethal heart failure with anti-mitochondrial antibody: an arrhythmogenic right ventricular cardiomyopathy mimetic

**Masayuki Koyama[1], Toshiyuki Yano[2]*, Keisuke Kikuchi[3], Daigo Nagahara[2], Hatsue Ishibashi-Ueda[4], and Tetsuji Miura[2]**

[1]Department of Cardiovascular Medicine, Obihiro Kosei Hospital, West-6 South-8, Obihiro, 080-0016, Japan; [2]Department of Cardiovascular, Renal and Metabolic Medicine, Sapporo Medical University School of Medicine, South-1 West-16, Chuo-ku, Sapporo, 060-8543, Japan; [3]Department of Diagnostic Pathology, Diagnostic Pathology, Obihiro Kosei Hospital, West-6 South-8, Obihiro, 080-0016, Japan; and [4]Department of Pathology, National Cerebral and Cardiovascular Centre, 5-7-1, Fujishiro-dai, Suita, Osaka, 565-8565, Japan
* Corresponding author. Tel: +81-11-611-2111, Fax: +81-11-644-7958, Email: tyano@sapmed.ac.jp

A 60-year-old woman with heart failure and no family history of cardiomyopathy/sudden death was referred to our hospital. Echocardiography revealed large pericardial effusion and thinning of akinetic right ventricular free wall (*Panels A* and *B*). Serum biochemistry showed low level of free thyroxine with elevation of thyroid-stimulating hormone level, elevated creatinine kinase level, and positive anti-mitochondrial antibody (AMA) (M2; 13.4 U/mL). Although supplementation of thyroid hormone and administration of diuretics were started, her condition was progressively deteriorated. Repeated echocardiography revealed reduction of left ventricular ejection fraction (40%), and right heart catheterization showed atrialization of right ventricular pressure pattern (*Panel C*), seen in Uhl's anomaly. Two weeks after her admission, she died from cardiogenic shock despite inotropic/mechanical supports, and an autopsy was performed. Macroscopic examinations showed right ventricle-dominant fibrofatty replacement (*Panel D*), similar to arrhythmogenic right ventricular cardiomyopathy (ARVC). In histology, there was massive infiltration of lymphocytes with various degrees of fibrofatty replacement in right and left ventricles (*Panels E* and *F*). Immunostaining with anti-plakophilin-2 antibody was positive without abnormality (*Panel E*, inset). Primary biliary cirrhosis (PBC) and Hashimoto's thyroiditis were histologically confirmed (*Panel G*). Taken together, the final diagnosis of AMA-positive lymphocytic myocarditis with Hashimoto's thyroiditis and PBC was made.




Cardiac involvement was found in approximately 30% of cases with AMA-positive myopathy, though detailed pathological analysis of the myocardium was not performed. Importantly, cases of right ventricle-dominant myocarditis were misdiagnosed as ARVC have been reported. Here we report a case in which AMA-positive myocarditis was the cause of a right ventricle-dominant cardiomyopathy with fibrofatty degeneration.