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A 19-year-old asymptomatic man was evaluated because of his sister's sudden death due to Naxos disease, a recessive form of arrhythmogenic right ventricular (RV) cardiomyopathy associated with palmoplantar keratoderma and woolly hair. It is caused by a plakoglobin mutation. Twelve-lead ECG showed inverted T waves in V1 through V6, incomplete right bundle branch block, and epsilon waves (Figure 1). Frequent ventricular extrasystoles were recorded on a 24-hour ECG. On echocardiography and angiography, the right ventricle was dilated and diffusely hypokinetic, but the left ventricle appeared normal. During 14 years of follow-up, he had 2 episodes of well-tolerated sustained ventricular tachycardia followed by biventricular progression leading to heart failure (New York Heart Association class III to IV) without arrhythmias. The patient died at 33 years of age while waiting for heart transplantation.

At gross examination and nuclear magnetic resonance (Figures 2 and 3), huge cardiomegaly (heart weight, 630 g) and paper-thin RV walls with transmural myocardial loss were evident, together with subepicardial left ventricular fatty infiltration. A calcified fibrous plaque, 7×9 cm in size, was discovered at the level of the RV outflow tract and confirmed by postmortem x-ray (Figure 4). Histological study showed transmural right and left ventricular myocardial atrophy, with fibrofatty replacement embedding a few surviving myocytes. The peculiar massive calcification of the RV outflow tract, possibly functioning as a ventricular disconnection, might explain the decreased arrhythmogenicity at the end-stage.

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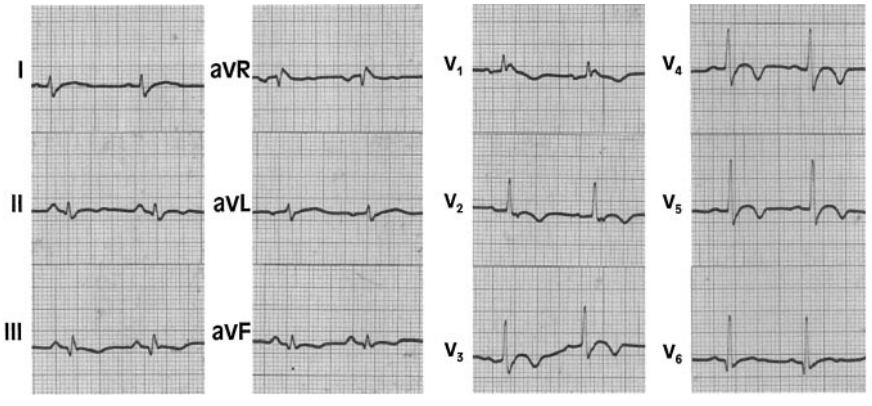

Figure 1. The 12-lead ECG shows inverted T waves (V1 to V6), incomplete right bundle branch block (QRS duration, 140 ms in V1), and epsilon waves.

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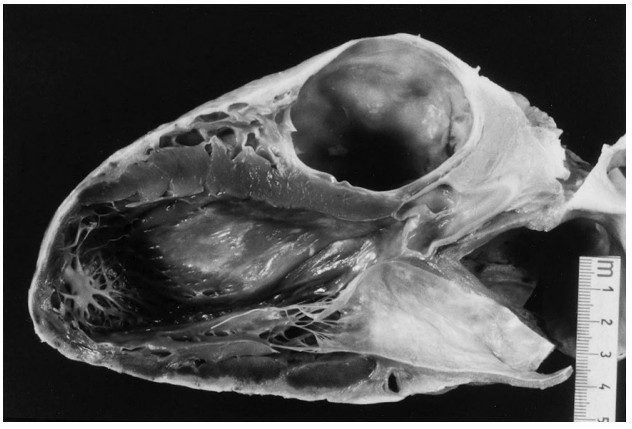

Figure 2. Parasternal long-axis view of the heart shows paper-thin RV walls, transmural myocardial loss, subepicardial left ventricular fatty infiltration, and calcified fibrous plaque at the level of RV outflow tract.

Figure 3. Corresponding long-axis nuclear magnetic resonance scan with diffuse bright signal on anterior RV wall and subepicardial bright signal on the left ventricular wall.

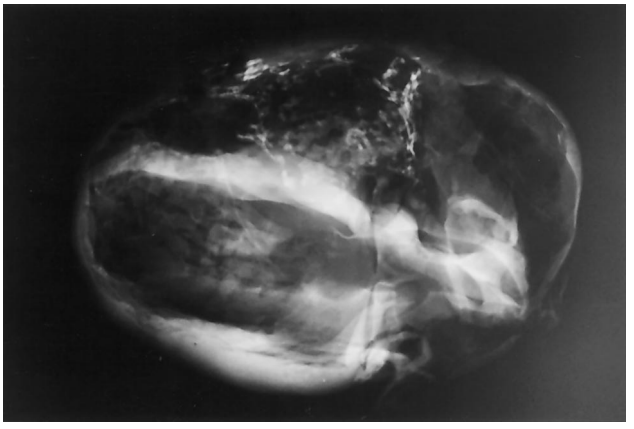

Figure 4. Postmortem x-ray reveals diffuse calcification of the anterior RV wall.