CASE REPORTS

Adult Case of Isolated Ventricular Noncompaction Discovered by Complete Atrioventricular Block

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A 25-year-old male was admitted to hospital with a 3-day history of worsening faintness. The electrocardiogram showed complete atrioventricular block. Echocardiography showed generalized hypokinesis and prominent trabeculations ranged from the apex to the mid-ventricular lateral wall of the left ventricle as well. Furthermore, trabeculations in the left ventricle were seen in his sister and brother. Thus, isolated ventricular noncompaction (IVNC) was diagnosed and a permanent pacemaker was implanted. Common clinical symptoms of IVNC are heart failure, ventricular arrhythmias, and embolic events. This is the first reported adult case of IVNC disclosed by the presence of complete atrioventricular block. (Circ J 2004; 68: 873–875)

Key Words: Cardiomyopathies; Complete atrioventricular block; Echocardiography; Isolated ventricular noncompaction

Isolated ventricular noncompaction (IVNC) is a rare cardiomyopathy that was initially reported in children but recently, adult cases have been reported. Common clinical symptoms of IVNC are heart failure, ventricular arrhythmias, and embolic events.

Case Report

A 25-year-old male was admitted to hospital with a 3-day history of worsening faintness. He had experienced occasional mild dyspnea on exertion since he was 21 years old, but did not have a history of syncope. When he was 16 years old, complete right bundle branch block was noted on the electrocardiogram (ECG). Otherwise his past history was unremarkable. He did not have a family history of sudden cardiac death.

On admission, his blood pressure was 98/64 mmHg with a regular pulse of 48 beats/min. Cardiac auscultation revealed a variable intensity of the first heart sound, but no other abnormal heart sounds or murmurs could be detected. The other examination findings were unremarkable. A 12-lead ECG revealed complete atrioventricular (A-V) block. Cardiomegaly and clear lung fields were present on chest radiography. Laboratory tests showed liver dysfunction, possibly related to alcohol intake. Cardiac enzymes were normal, and the laboratory data did not indicate inflammation. Echocardiography showed an enlarged left atrium and ventricle (left atrium: 51 mm; left ventricular diameters: diastolic 64 mm, systolic 53 mm). The left ventricular wall motion was generalized hypokinesis with wall thinning of the basal ventricular septum. Systolic function was severely depressed (fractional shortening 17%). Prominent trabeculations and deep intertrabecular recesses were observed on the apical and mid-ventricular aspects of the lateral wall of the left ventricle. The ratio of the noncompacted zone to the compacted zone was 2.3. Gene mutations (G4.5, FKBP12, Dystrobrevin, Desmin, Syntrophin) were not detected by genetic analysis.

The coronary arteries appeared normal on coronary angiography. The left ventricular end-diastolic pressure was 18 mmHg, and the cardiac index was 1.82 L·min⁻¹·m⁻² on cardiac catheterization. An endomyocardial biopsy from the left ventricle revealed mild endocardial thickening and interstitial fibrosis. There was no histological evidence of inflammation.

Although we suspected cardiac sarcoidosis as well, the clinical manifestations of sarcoidosis were not seen in the lungs, skin, eye or liver. The concentrations of angiotensin-converting enzyme and gamma globulin in a blood test were normal, and the tuberculin reaction proved positive. Moreover, the gallium-67 scan showed no uptake. Therefore, we diagnosed IVNC, and a permanent pacemaker was implanted. Trabeculations in the left ventricle were seen in his sister and brother as well. The ECG of his sister showed first degree A-V block, and that of his brother showed complete right bundle branch block.

Discussion

According to the previous reports, 88–94% of patients with IVNC show abnormalities on the resting ECG. Oechslin et al reported that abnormal ECGs were seen in 94% of 34 adults, and 56% of them showed ventricular conduction defects; they also found that ventricular conduction defects were more frequently present in nonsurvivors than in survivors. Tachyarrhythmias, such as atrial fibrillation or ventricular tachycardia, are noted in the previous studies, and the life-threatening ventricular arrhythmias needed an implantable defibrillator. The present case showed complete A-V block. Two pediatric cases of IVNC associated with complete A-V block have been reported, and one received an implantable permanent pacemaker, but subsequently died at the age of 13 years. The other patient...
developed A-V block at 4 years. The present patient had maintained sinus rhythm till he was 16 years old and had been working as a manual laborer prior to his admission to hospital. To our knowledge, this is the first adult case of IVNC discovered by the presence of complete A-V block.

The cause of ventricular conduction abnormalities is unclear. Ichida et al suggest that the ventricular conduction abnormalities, such as left bundle branch block, are the result of the progressive endocardial fibrosis in IVNC, and they are observed much more frequently in the adult population because the fibrotic change develops gradually. Because the present patient had complete A-V block, we suspect there is a severe histological change to the conduction system.

Familial occurrence of IVNC has been described. Oechslin et al found it in 18% of their series, and suggested that first-degree relatives should be screened by echocardiography. In a Japanese pediatric series, they found frequent familial occurrence. In the present case, a sister and brother were also diagnosed as IVNC, and the ECG of the sister showed first degree A-V block. She requires special attention, because Ichida et al have reported a case that progressed from second degree A-V block to complete block.

IVNC should be distinguished from prominent normal myocardial trabeculations, hypertrophic cardiomyopathy, left ventricular hypertrophy caused by hypertensive or valvular heart disease, dilated cardiomyopathy, and left ventricular apical thrombus. As in the previously described echocardiographic findings, the present case revealed prominent left ventricular trabeculations with deep intratrabecular recesses on color Doppler imaging. Ventricular noncompaction was located at the apex and the mid-ventricular lateral wall. Oechslin et al found that more than 80% of their IVNC cases showed trabecular formation in the same location as the present case.

To our knowledge, this is the first reported adult case of IVNC discovered by the presence of complete A-V block. The diagnosis of IVNC is often difficult, but accurate diagnosis is needed because of its poor clinical course. We propose that IVNC should also be considered in patients presenting with complete A-V block.

**Acknowledgments**

We thank Fukiko Ichida, MD (Department of Pediatrics, Toyama Medical and Pharmaceutical University) for her skillful assistance with the genetic analysis and Nozomi Watanabe, MD (Department of Cardiology, Kawasaki Medical School) for her help in preparing this manuscript.

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