Atrial Right-to-left Shunt without Pulmonary Hypertension in a Patient with Biventricular Non-compaction Cardiomyopathy Accompanied by Ventricular and Atrial Septal Defects

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Abstract

Echocardiography and magnetic resonance imaging revealed biventricular non-compaction cardiomyopathy with ventricular (VSD) and atrial (ASD) septal defects in an unconscious, 23-year-old hypoxemic man. Doppler echocardiography showed a left-to-right shunt across the VSD and a right-to-left shunt across the ASD. Cardiac catheterization revealed elevated right atrial pressure, although pulmonary pressure was normal. We considered that the atrial right-to-left shunt had induced the hypoxemia, which was related mainly to right ventricular dysfunction in this biventricular non-compaction cardiomyopathy, but it was not related to pulmonary hypertension.

Key words: hypoxemia, hypoplasia, tricuspid valve, right ventricle

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Introduction

Atrial right-to-left shunt is an important cause of hypoxemia that is frequently induced by an interatrial defect with pulmonary hypertension (1). However, even without pulmonary hypertension, this condition rarely occurs in patients with platypnea-orthodeoxia syndrome (2, 3) and with right ventricular dysfunction (4, 5). In contrast, non-compaction cardiomyopathy is a rare congenital disease characterized by prominent ventricular trabeculations and deep intertrabecular recesses (6). It involves the left ventricle (LV) more frequently and rarely the right (RV) or both ventricles (7-10). We describe a rare atrial right-to-left shunt without pulmonary hypertension in biventricular non-compaction cardiomyopathy complicated with ventricular (VSD) and atrial (ASD) septal defects.

Case Report

A 23-year-old man diagnosed with ASD was admitted to our hospital with dyspnea upon exertion followed by a transient loss of consciousness. He had no significant medical history of pathologies such as arrhythmias and cardioembolic events. His family history did not include heart disease, sudden, or premature death. He had a regular heart rate of 80 beats/min, blood pressure of 118/60 mmHg and decreased oxygen saturation of 90% despite oxygen inhalation. Cardiac auscultation revealed no obvious murmur or gallop, and respiratory sounds were normal without rales on pulmonary auscultation. Neurological findings were normal and peripheral edema was absent. Chest radiography and electrocardiography findings appeared normal. Laboratory findings including B-type natriuretic peptide (19.8 pg/mL) were also normal. Transthoracic apical 4-chamber echocar-

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Figure 1. Transthoracic echocardiography shows apical biventricular diffuse hypokinesis with suspected increased trabecularization (Panel A, end-diastole; B, end-systole, arrows) and apical ventricular septal defect (Dotted-line circle).

Figure 2. Color Doppler echocardiography shows left-to-right shunt flow across ventricular septal defect on transthoracic echocardiography (Panel A), and right-to-left shunt flow across atrial septal defect on transesophageal echocardiography (Panel B).

diography revealed apical biventricular diffuse mild hypokinesis with the possibility of increased trabecularization (arrows) and an apical VSD (dotted circle, Fig. 1). The RV longitudinal length and tricuspid annular diameter were 49.2 and 16.5 mm, respectively. Their Z scores, calculated as (observed value-mean normal value) /standard deviation around the mean normal value using normal reference data (11, 12), were -2.02 and -14.5, respectively. These findings suggested hypoplasia of the RV cavity and tricuspid valve (TV). Color Doppler transthoracic and transesophageal

echocardiography revealed left-to-right shunt flow across the VSD (Fig. 2A), and right-to-left shunt flow across the ASD (Fig. 2B), respectively. We further evaluated the apical myocardium using cardiac magnetic resonance cine imaging of the VSD and ASD. The LV excluding the basal septum and the apical RV had hypertrabeculations and deep intertrabecular recesses (Fig. 3A and C) and the calculated ratio of the distance between the epicardial surface and the trough of the recesses (x) to the distance between the epicardial surface and the peak of the trabeculations (y) was 0.30 (Fig. 3B).

Figure 3. Cardiac magnetic resonance cine imaging confirmed hypertrabeculations and deep intertrabecular recesses in left and right ventricles (Panel A and C). Calculated ratio of distance between epicardial surface and trough of recesses (x) to distance between epicardial surface and peak of the trabeculations (y) is 0.30 (Panel B). Ventricular and atrial septal defect is located at apical inferoseptal wall and basal portion of atrial septum near attachment of atrioventricular valves, respectively (Panels C and D).

These findings suggested biventricular non-compaction cardiomyopathy. The hypoplasia of the RV and TV was confirmed. Fat infiltration into the RV suggesting arrhythmogenic right ventricular cardiomyopathy was not found. The VSD and ASD were located at the apical inferoseptal wall and the basal portion of the atrial septum near the attachment of the atrioventricular valves, respectively (Fig. 3C and D). We diagnosed biventricular non-compaction cardiomyopathy complicated with muscular type VSD and secundum type ASD, and considered that the right-to-left shunt across the ASD was the cause of the hypoxemia. We performed cardiac catheterization to clarify the causes of the atrial right-to-left shunt (Fig. 4). Pulmonary artery pressure was normal, but pressure was higher during the diastolic phase in the RV than in the LV, and consistently higher in the right (RA) than in the left (LA) atrium. A pressure gradient in the RV suggesting a double-chambered right ventricle was undetectable. Angiography of the coronary arteries including RV branches revealed no significant stenosis. These findings indicated that the cause of the atrial right-to-left shunt was RA pressure elevation induced by impaired tricuspid inflow from the RA to the RV

due to increased RV diastolic pressure. The elevated RV pressure was not related to pulmonary hypertension but mainly to RV dysfunction in this patient with biventricular non-compaction cardiomyopathy. We are presently considering surgical closure of the atrioventricular defect to improve the symptoms and hemodynamics.

Discussion

We described an atrial right-to-left shunt without pulmonary hypertension in a patient with biventricular non-compaction cardiomyopathy accompanied by ventricular and atrial septal defects.

Atrial right-to-left shunt without pulmonary hypertension is a rare but important cause of hypoxemia. To differentiate between an anatomical phenomenon that does not always require an interatrial pressure gradient and a hemodynamic phenomenon with an interatrial pressure gradient is essential to elucidating the mechanism of right-to-left shunt without pulmonary hypertension (4). The anatomical phenomenon is venous blood streaming from the inferior vena cava to the LA due to an over-developed Eustachian valve, namely

Figure 4. Cardiac catheterization shows significantly higher ventricular pressure during diastolic phase in the right than in left ventricle, and pressure was constantly higher in the right, compared with left atrium, although pulmonary artery pressure was normal.

platypnea-orthodeoxia syndrome (2, 3). Hemodynamically, the higher pressure in the RA than in the LA is closely related to RV dysfunction. Atrial right-to-left shunt due to RV dysfunction can develop in patients with an obstructed right ventricular outflow tract, RV infarction, or arrhythmogenic right ventricular cardiomyopathy (4, 5), but as far as we can ascertain, never in patients with hypoplasia of the RV and TV in biventricular non-compaction cardiomyopathy. The standard treatment for an interatrial defect is surgical closure under extracorporeal circulation. Although transcatheter-closure of an ASD has recently been deemed safe and effective (5), this procedure was considered very dangerous for our patient because of the risk of deteriorating RV dysfunction imposed by increased RV volume.

Biventricular non-compaction cardiomyopathy is rare (7-10), and diagnostic criteria have not been standardized. Non-compaction cardiomyopathy is considerably more difficult to diagnose in the RV than in the LV, because trabeculations are naturally increased in the RV. Burke et al (13) proposed defining RV non-compaction as a non-compact inner RV myocardial layer adjacent to the TV comprising more than 75% of the RV thickness. However, this definition was difficult to apply to the present patient because the non-compact RV layer was distributed in the apex rather than in the base. We thus ruled out RV dysfunction caused by other factors such as RV infarction or ischemia and arrhythmogenic right ventricular cardiomyopathy, and reached conclusions based on other factors in addition to the presence of increased trabeculations in the RV. The clinical manifestations of biventricular non-compaction cardiomyopathy include some symptoms of right heart failure, such as distended neck veins, hepatomegaly and leg edema (8), in addition to the left heart failure, arrhythmia, and thromboembolism associated with LV non-compaction cardiomyopathy. However, symptoms of right and left heart

failure were not remarkable in the present patient, perhaps because venous congestion was unlikely due to 1) the preferential streaming of venous blood from the right to the left atrium through the ASD and 2) the relatively mild severity of the non-compaction.

Conclusion

We described a rare atrial right-to-left shunt without pulmonary hypertension in biventricular non-compaction cardiomyopathy complicated with ventricular and atrial septal defects. Although this condition is extremely rare, we believe that it should be considered when an atrial right-to-left shunt exists in the absence of pulmonary hypertension.

The authors state that they have no Conflict of Interest (COI).

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