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Brief Report

Complete atrioventricular block and reversible systolic dysfunction in left ventricular hypertrabeculation/ non-compaction with metabolic myopathy

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Abstract A 32-year-old female patient presented with cardiac failure because of systolic dysfunction. Five years before, a DDD pacemaker had been implanted because of complete atrioventricular block. Echocardiographic examination disclosed left ventricular hypertrabeculation/non-compaction. Because of sinus tachycardia, ivabradine was started and the patient's left ventricular function returned to normal within 4 months. Recurrent creatine-kinase elevation and reduced nicotinamide adenine dinucleotide staining on muscle biopsy suggested metabolic myopathy.

Keywords: Cardiac failure; cardiomyopathy; pacemaker

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Left ventricular hypertrabeculation/non-compaction is a cardiac abnormality of unknown aetiology, frequently associated with left ventricular dysfunction. Electrocardiographic abnormalities are found in up to 90% of left ventricular hypertrabeculation/non-compaction cases and comprise tall QRS complexes, ST-abnormalities, left bundle branch block, and atrial fibrillation.[1,2] Occasionally, complete atrioventricular block, either congenital or acquired, may be associated with left ventricular hypertrabeculation/non-compaction (Table 1).[2–14]

Case report

In a 32-year-old female patient, complete atrioventricular block with a ventricular rate of 38 per minute was first recorded at the age of 20 years. Her pulse had been low since childhood; however, no electrocardiogram had been performed before the age of 20 years. She was without any cardiac symptoms. She was born as the third child to healthy non-consanguineous parents. Echocardiography at the age of 20 years revealed an enlarged left ventricle (left ventricular end-diastolic diameter 64 millimetres) with moderate systolic dysfunction and mild mitral regurgitation. At the age of 27 years, the patient complained of vertigo and fatigue. Since the atrioventricular block with bradycardia was considered the cause of her symptoms, a DDD pacemaker was implanted. At the age of 32 years, she was hospitalised because of cardiac failure.

Clinical examination revealed leg oedema, resting dyspnoea, jugular vein distension, and pulmonary rales. Blood pressure was 130/100 millimetres of mercury. The electrocardiogram showed sinus tachycardia of 120 per minute and paced ventricular rhythm of left bundle branch block morphology. Pacemaker interrogation showed that a heart rate greater than 100 beats per minute had been present for 40% of the preceding year. Blood chemistry investigations revealed moderate renal insufficiency (creatinine 1.38, normal: less than 1.11 milligrams per decilitre) elevated glutamate-oxalate-transaminase

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Table 1. Cases with left ventricular hypertrabeculation/non-compaction and complete atrioventricular block.

Author	Age/sex	Atrioventricular block	Symptoms	Comorbidities	Therapy, outcome
3	56 years/female	NI	Syncope	Hyperkalaemia, systolic dysfunction	Medication, progressive cardiac failure
4	18 years/female	NI	Pre-syncope	Anterior mitral leaflet cleft	DDD pacemaker, after 12 months symptom-free
5	3 days/male	C	Bradycardia	QT prolongation	DDD pacemaker, cardiac failure after 4 months, improvement after biventricular pacing
6	47 years/male	A	Dyspnoea, syncope	Nail–patella syndrome, mitochondrial myopathy, renal failure	DDD pacemaker, dialysis
7	77 years/male	A	Cardiac failure	Coronary cardiac disease, metabolic myopathy	Died due to cardiac failure
8	3 years/female	NI	Arrhythmia	NI	Pacemaker
	5 years/male	NI	Cardiac murmur	Systolic dysfunction	Died after 13 years due to pulmonary embolism
9	10 years/male	NI	Cardiac failure	Systolic dysfunction	Medication, pacemaker
10	24 years/male	NI	Syncope	Atrial fibrillation	NI
11	11 years/male	A	None	Double-orifice mitral valve	No medication
12	83 years/male	A	Dyspnoea	Systolic dysfunction	Pacemaker, medication, improvement
13	25 years/male	A	Faintness	Systolic dysfunction	Pacemaker
14	70 years/male	NI	Dyspnoea, dizziness	Valvular insufficiency	Medication, refused pacemaker
2	n = 2	NI	NI	NI	NI

A = acquired; C = congenital; NI = not indicated

(213, normal: less than 31 units per litre), glutamate-pyruvate-transaminase (335, normal: less than 34 units per litre), gamma-glutamyl-transpeptidase (81, normal: less than 38 units per litre), creatine kinase (242, normal: less than 145 units per litre), and total bilirubin (1.3, normal: less than 1.11 milligrams per decilitre), positive T-troponin, a reduced serum iron (22, normal: 40–150 micrograms per decilitre) and hypothyroidism (TSH 8.880, normal: less than 3.1 microIU/ml). Plasma pro-brain natriuretic peptide level was 6941 picograms per millilitre (normal: less than 132). Transthoracic and transesophageal echocardiography revealed dilated cardiac cavities, hypertrabeculation of the apex and lateral wall, severely reduced systolic function, prolapse of both mitral leaflets, moderate mitral and tricuspid valve insufficiency, and an estimated pulmonary artery pressure of 45 millimetres of mercury. Coronary angiography showed normal coronary arteries and ventriculography and hypertrabeculated left ventricle with an ejection fraction of 26% (Fig 1). Due to associated neurological abnormalities found with left ventricular hypertrabeculation/non-compaction, she was referred to a neurologist who found weakness of eyelid closure, bilateral distal weakness (M5-) of the upper limbs, diffuse bilateral weakness of the lower limbs (M5-), and generally reduced tendon reflexes, suggesting a myopathy.

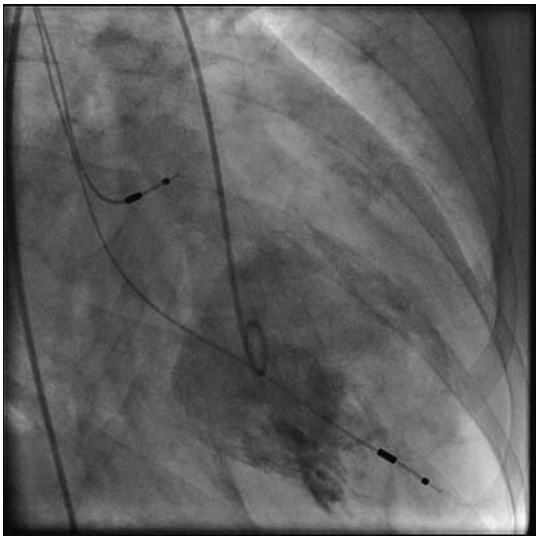

Figure 1.
Left ventriculography in 30 degrees left anterior oblique projection showing the dilated left ventricle with hypertrabeculation/non-compaction of the apical region.

She was categorised in New York Heart Association class II with lisinopril, bisoprolol, spironolactone, and ivabradine, but without any anti-thrombotic therapy. She was re-admitted 3 days later because of a right-sided hemiparesis due to occlusion of the left internal

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carotid artery, which was attributed to cardioembolism. Phenprocoumon was started. Cardiac symptoms improved considerably: the heart rate decreased to 60 beats per minute and pro-brain natriuretic peptide level to decreased to 700 picograms per millilitre. Echocardiography, however, still showed a severely depressed systolic function, which is why implantation of an implantable cardioverter-defibrillator and cardiac resynchronisation therapy was planned. However, echocardiographic examination in February 2009 surprisingly showed normal-sized cardiac cavities and a left ventricular ejection fraction of 55%. The patient did not complain of symptoms of cardiac failure and the pro-brain natriuretic peptide level had further decreased to 200 picograms per millilitre, which is why implantable cardioverter-defibrillator and cardiac resynchronisation was cancelled. At present, the patient is under a pharmacotherapy with phenprocoumon, bisoprolol, ivabradine, and lisinopril. Muscle biopsy revealed reduced nicotinamide adenine dinucleotide staining and muscle fibre type-II predominance.

Discussion

Complete atrioventricular block may be congenital or acquired. The aetiology of atrioventricular block may be due to affection of the cardiac conduction system by coronary cardiac disease, infiltrative cardiac disease, infectious disease, collagen vascular disease, trauma, post-surgery, tumours, and neuromuscular disorders. Neuromuscular disorders in which a complete atrioventricular block has been frequently reported include myotonic dystrophy, Emery–Dreifuss muscular dystrophy, Duchenne and Becker muscular dystrophy, and mitochondrial myopathy. Congenital atrioventricular block is frequently associated with maternal autoimmune disorders.[15] Whether atrioventricular block in our patient was congenital or acquired remains uncertain, like in most of the reported cases with atrioventricular block and left ventricular hypertrabeculation/non-compaction (Table 1). In our case, atrioventricular block was most probably caused by cardiac involvement of the suspected myopathy. The coexistence of mitral valve abnormalities and atrioventricular block in left ventricular hypertrabeculation/non-compaction has been described previously.[4,11] Most probably, these abnormalities were congenital since both of these previously described cases are children or adolescents, like our patient.

Systolic dysfunction in our patient may be due to the insufficiently dosed pharmacotherapy, inadequate rhythm control and coexisting myopathy with cardiac involvement. Development of systolic dysfunction due to dyssynchrony of ventricular contractions after right ventricular pacing is a further possibility.[16] A cohort study in children with congenital complete atrioventricular block found an incidence of left ventricular systolic dysfunction of 6% during a follow-up period of 15 years.[17] In patients with left ventricular hypertrabeculation/non-compaction, deterioration of systolic function after pacemaker implantation has been so far only reported in a baby with congenital atrioventricular block (Table 1).[5] In our patient, however, left ventricular enlargement and systolic dysfunction had been documented already before pacemaker implantation and regressed considerably after rhythm control had been achieved; thus, right ventricular pacing is an unlikely cause of deterioration of systolic function. Improvement of systolic function after rate control with ivabradine may also indicate that our patient suffered from tachycardia-induced cardiomyopathy; however, that mechanism does not explain the left ventricular dysfunction at the age of 20 years.

Whether left ventricular hypertrabeculation/non-compaction was congenital or acquired in our patient cannot be definitively assessed. Left ventricular hypertrabeculation/non-compaction is not mentioned in the echocardiographic report when she was investigated at the age of 20 years; however, left ventricular hypertrabeculation/non-compaction might have been overlooked.[1] The atrioventricular block is likely to be congenital since her heart rate has been low since childhood.

We conclude that patients with left ventricular hypertrabeculation/non-compaction and an atrioventricular block require close clinical and echocardiographic follow-up. In cases with left ventricular systolic dysfunction, adequately dosed neurohumoral pharmacotherapy and oral anti-coagulation should be prescribed. Neurohumoral pharmacotherapy and rhythm control may lead to considerable clinical and echocardiographic improvement in patients with left ventricular hypertrabeculation/non-compaction. If cardiac failure persists despite pharmacotherapy, implantation of a cardioverter-defibrillator and cardiac resynchronisation should be considered. Whether patients with left ventricular hypertrabeculation/non-compaction and myopathy deserve a closer follow-up than those with left ventricular hypertrabeculation/non-compaction patients without myopathy, is at present unknown.

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