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Kill Two Birds with One Stone: Special Ventricular Arrhythmias and Complex Cardiac Malformation

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1. Abstract

We are reporting a case of special ventricular arrhythmias from the aortomitral continuity. However, it is difficult to treat with radiofrequency ablation, because the patient also suffered from the complex cardiac malformation (Shone's complex and double-chambered left ventricle). Staged operations were performed to treat this patient, and the follow-up results showed that the treatments were satisfactory

2. Clinical Presentation

A 22-year-old Chinese female without previous family history of cardiovascular disease was admitted to our hospital due to several episodes of syncope in the past few years. Of note, she reported a recurrent palpitation in the past few years associated with progressive dyspnea reaching New York Heart Association Class II. The initial clinical examination revealed frequent premature heart beat with a heart rate of 84bpm and an apical grade 3/6 blowing murmur.

What should we do next? We were ready to conduct the inspections, including the electrocardiogram, Holter monitoring and the catheter electrophysiological examination, to make a definite diagnosis.

3. Supplementary Examination

3.1. Thoracic X-ray

Thoracic x-ray (TOSHIBA Japan E7252X) showed a double hump aspect for the left middle arch and a protrusion of the right lower arch and an S-shaped lateral bending of the thoracolumbar segment.

3.2. Electrocardiogram (ECG) Characteristics

Sinus rate: incomplete right bundle branch block. Ventricular arrhythmias (right bundle branch block type): premature ventricular beat, sustained ventricular tachycardia (123bpm, Figure 1A a) and accelerated idioventricular rhythm (94bpm, Figure 1A b). The main wave orientations of lead V1-V6, II, III and aVF were positive (R/S＞1). The main wave orientations of lead I and aVL were negative. The origin of the ventricular arrhythmias rhythm located in the region of aortomitral continuity (AMC, Figure 1A c, d, e) 1, according to the ECG.

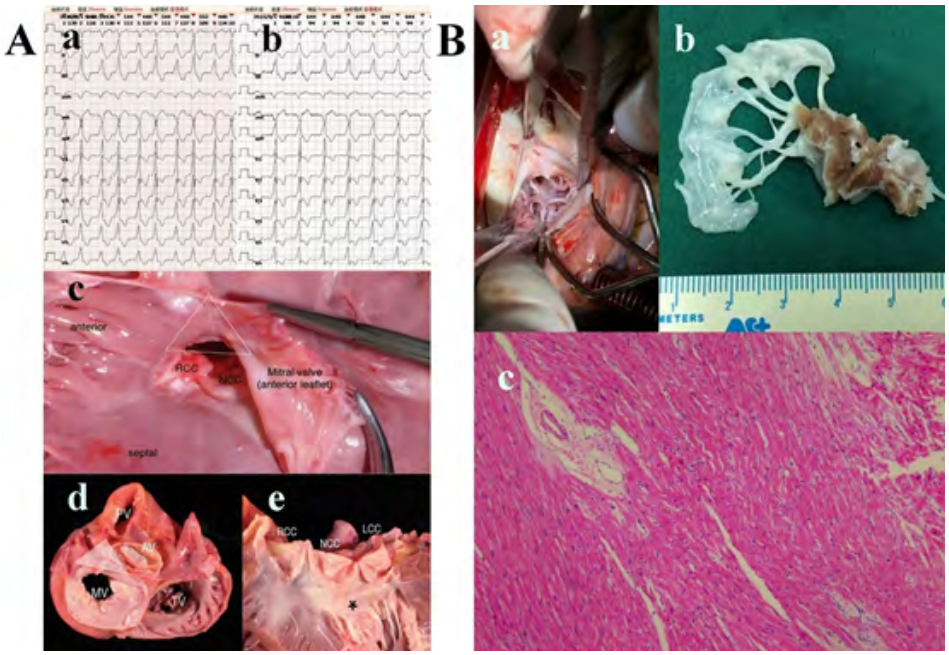

Figure 1: The electrocardiogram and the anatomy of aortomitral continuity and the pathological images of the parachute mitral valve and the abnormal muscular septum

A The electrocardiogram and the anatomy of aortomitral continuity

a) Ventricular tachycardia, 123bpm, 25mm/s 5mm/mV.

b) Accelerated idioventricular rhythm, 94bpm, 25mm/s 5mm/mV.

c) Left ventricular outflow tract (left ventricular view): The triangle region (△) represents the AMC, which is between LCC and MV (anterior leaflet).

d) The anatomic relationship among AV, PV, MV and TV (atria view): The triangle region (△) represents the AMC.

e) The anatomic relationship between AV and MV (left ventricular view): The * represents the anterior leaflet.

B The pathological images of the parachute mitral valve and the abnormal muscular septum

a) The annular mitral valve.

b) The removed mitral valve with all chordae inserted into a single papillary muscle and the abnormal muscular septum.

c) The HE staining of the abnormal muscular septum.

AV: aortic valve, PV: pulmonary valve, MV: mitral valve, TV: tricuspid valve, LCC: left coronary cusp, RCC: right coronary cusp, NCC: Non-coronary cusp, AMC: aortomitral continuity, HE: hematoxylin-eosin

3.3. Holter Monitoring

Holter monitoring showed sinus with heart rate of 38bpm to 105bpm. Total 20484 ventricular premature contractions and 113 ventricular runs, the longest of which was 210 ventricular beats, were detected during 24 hours (Table 1).

We need to clear the heart structure of this patient before the catheter electrophysiological examination. Therefore, we arranged the echocardiography and computed tomography angiography.

Table 1: The results of the transthoracic echocardiography and the Holter monitoring

	Before treatments	After the first operation	After the second operation	Follow-up
the results of the transthoracic echocardiography				
LA (mm)	43	43	39	39
LV (mm)	46	42	39	43
EF (%)	60	51	55	60
Mitral valve				
Morphology	mitral valve with all chordae inserted into a single papillary muscle	Artificial valve	Artificial valve	Artificial valve
MV-E (m/s)	1.51	2.63	1.71	1.79
Aortic valve				
Morphology	bicuspid aortic valve	bicuspid aortic valve	bicuspid aortic valve	bicuspid aortic valve

AV (m/s)	2.62	2.1	1.47	1.83
Abnormal structure in left ventricular Muscular septum	Abnormal muscular septum in the middle of left ventricular	surgical resection	surgical resection	surgical resection
Blood flow velocity in left ventricular (m/s)	1.9	Normal	Normal	Normal
Aorta coarctation Width of the orifice (mm) Velocity (m/s)	4.64 4.04	4.04 3.5	release of aorta coarctation 1.56	release of aorta coarctation 1.85
the results of the holter monitoring				
General				
QRS complexes	83542	96145	124410	108240
Ventricular	23048	1	0	1
Supraventricular	439	39	41	155
Heart rates				
Minimum	38	62	84	59
Average	56	71	91	76
Maximum	105	91	98	107
Max R-R (s)	1.86	1.41	0.8	1.39
Ventriculars				
Isolated	20484	1	0	1
Couplets	362	0	0	0
Bigeminal cycles	12157	0	0	0
Runs	113runs 1840beats	0	0	0
Longest run	210beats 89bpm	0	0	0
Fastest run	27beats 104bpm	0	0	0

LA: left atrium LV: left ventricular EF: ejection fraction MV-E: mitral valve flow velocity-E AV: aortic valve flow velocity

3.4. 2- and 3-Dimensional Transthoracic Echocardiography (TTE)

The TTE (Philips Nederland iE Elite) showed that (Figure 2A) 1) Left Atrium (LA): 43mm left ventricular (LV): 46mm Ejection Fraction (EF): 60%; 2) the stenosis mitral valve with all chordae inserted into a single papillary muscle (parachute mitral valve, the orifice area:1.68cm2); 3) abnormal muscular septum in the middle of left ventricular, which originated in the middle of posterior-inferior wall and posterior ventricular septum extending to the left ventricular, separated it into two chambers; 4) bicuspid aortic valve without stenosis or regurgitation; 5) a stenosis of the junction area between the aortic arch and descending aorta (width of the orifice: 4.64mm, velocity: 4.04m/s, mean pressure gradient: 31mmHg, max pressure gradient: 65mmHg).

3.5. Heart and Aorta Computed Tomography (CT) Angiography

Heart and aorta CT (SIEMENS Germany somatom sensation 64) angiography showed a double-chambered left ventricle with almost the equate size of each cavity due to an abnormal muscle septum extending out from the middle of posterior-inferior wall and posterior interventricular septum (Figure 2C a). Other anomalies included a significantly enlarged left atrium, pulmonary congestion and spinal curvature. 3-dimensional reconstruction of the CT scan revealed a stenosis of the junction area between the aortic arch and descending aorta (the minimum diameter =7.3mm, Figure 2C b), constriction on the initial segment of the descending aorta with expansion of the distal part (maximum diameter = 32.2mm)

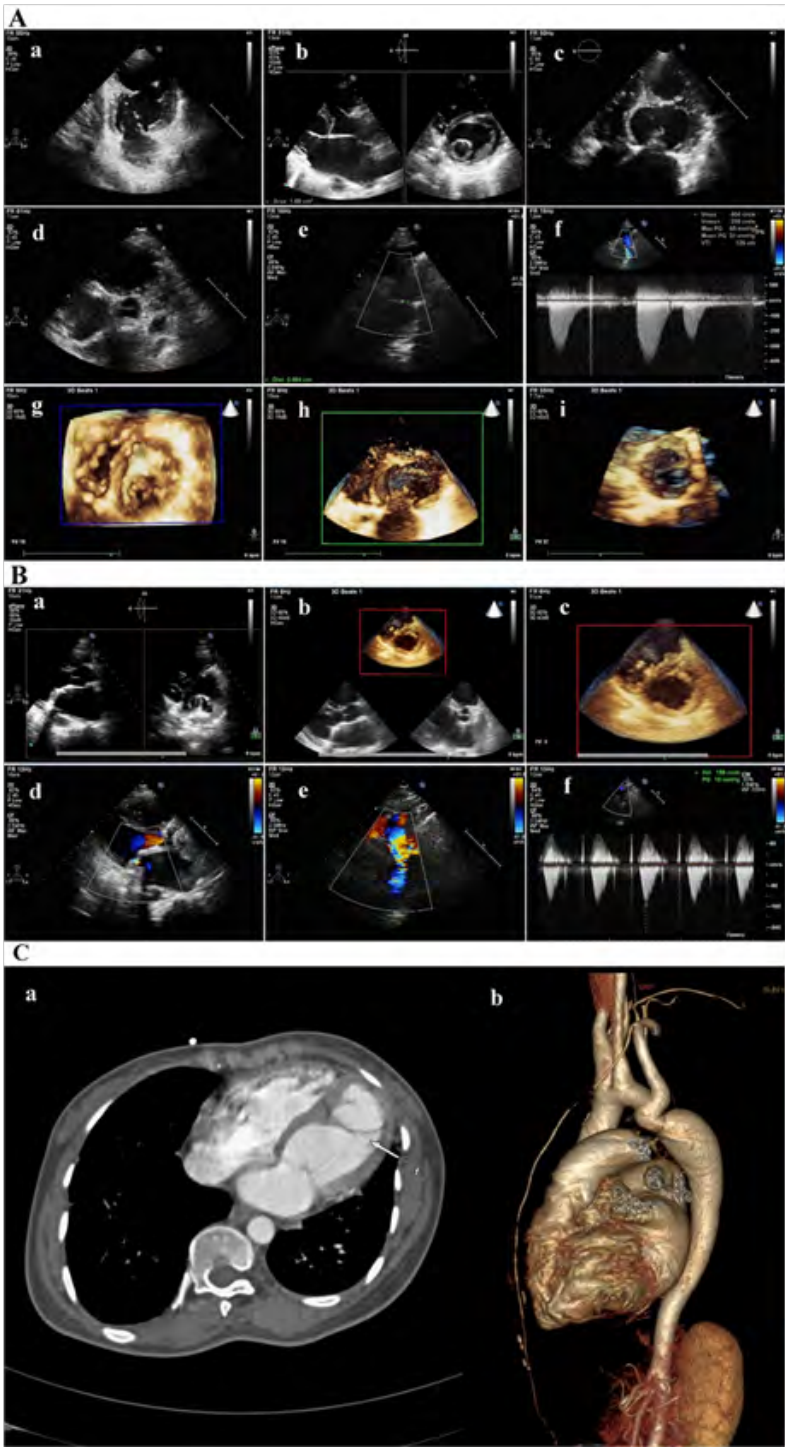

Figure 2: 2-, 3-dimensional transthoracic echocardiography and computed tomography angiography

A transthoracic echocardiography before treatments

a) 2-dimensional mode shown the parachute mitral valve (mitral valve with all chordae inserted into a single papillary muscle, apical 2-chamber view).

b) X-Plane mode shown the parachute mitral valve. In parasternal long axis view, all chordae insert into a single (posterior) papillary muscle. In parasternal short axis view mitral valve level, the orifice of the parachute mitral valve is eccentric and the area is 1.68cm^2.

c) 2-dimensional mode shown the muscular septum in the middle of left ventricular and the double-chambered left ventricular (apical 4-chamber view).

d) 2-dimensional mode shown the bicuspid aortic valves (parasternal short axis view).

e) 2-dimensional mode shown the region of descending aortic arch coarctation (suprasternal fossae view, width of the orifice: 4.64mm)

f) Doppler spectrum mode shown the aortic blood flow velocity accelerated (suprasternal fossae view, Vmax: 4.04m/s, max pressure gradient: 65mmHg, mean pressure gradient: 31mmHg).

g) 3-dimensional mode shown the parachute mitral valve opening restriction.

h) 3-dimensional mode shown the muscular septum in the middle of left ventricular, the double-chambered left ventricular and the parachute mitral valve (mitral valve with all chordae inserted into a single papillary muscle).

i) 3-dimensional mode shown the bicuspid aortic valves.

B transthoracic echocardiography after two operations

a) X-Plane mode shown the artificial valve (parasternal long axis view and parasternal short axis view, mitral valve level at the same time) after the first operation.

b) 3-dimensional mode shown the left ventricular after the first operation, in which the muscular septum was removed to treat the double-chambered left ventricular.

c) 3-dimensional mode shown the middle left ventricular (parasternal short axis view, papillary muscle level) after the first operation, in which the muscular septum was removed to treat the double-chambered left ventricular.

d) 2-dimensional mode shown the middle left ventricular (parasternal long axis view) after the first operation, in which the muscular septum was removed to treat the double-chambered left ventricular.

e) 2-dimensional mode shown the region of descending aortic arch (suprasternal fossae view) after the second operation, which released of obstruction in the descending aortic arch area.

f) Doppler spectrum mode shown the aortic blood flow velocity return to normal (suprasternal fossae view, Vmax: 1.56m/s) after the second operation.

C heart and aorta computed tomography angiography

a) The abnormal muscular septum in the middle of left ventricle.

b) 3-dimensional reconstruction of the aorta coarctation.

4. Diagnosis

After the above preoperative examinations, the patient was diagnosed as ventricular arrhythmias including premature ventricular beat, sustained ventricular tachycardia and accelerated idioventricular rhythm, from the AMC and Shone's complex (parachute mitral valve, bicuspid aortic valve and aortic coarctation) combined with double-chambered left ventricle.

5. Clinical Treatments

What should we do next? How to treat the ventricular arrhythmias, radiofrequency ablation or other treatments? Did the Shone's complex and double-chambered left ventricle need to be treated?

Considering the patient's situation, staged operations were recommended as follow: 1 mitral valve replacement and resection of the abnormal muscular septum, 2 releases of aorta coarctation, 3 catheter electrophysiological examination and radiofrequency ablation for the ventricular arrhythmias.

In the first stage, the patient received the operation of mitral valve replacement and resection of the abnormal muscular septum in the left ventricular (on 2015.09.14). During the first surgical operation, the deformities were confirmed that the stenosis annular mitral valve with all chordae inserting into a single hypertrophic papillary muscle (Figure1B a, b) and abnormal muscular septum (Figure1B b, c) separated the left ventricle into two chambers. After the operation, the patient had an uneventful recovery and reexamination of the Holter monitoring and TTE. Something unexpected happened: the Holter monitoring showed sinus rhythm with heart rate of 62bpm to 91bpm; there were a total of 96145 heart beats during 24 hours and only one ventricular premature beat without particular anti-arrhythmias therapeutic options (Table 1). The TTE showed that (Figure2B a, b, c, d) 1) LA: 43mm LV: 42mm EF: 51%; 2) the parachute mitral valve was removed and replaced by an artificial valve, which function was normal; 3) the abnormal muscular septum in the middle of left ventricular was removed, and the left ventricular retunes to one chamber; 4) bicuspid aortic valve without stenosis or regurgitation; 5) a stenosis of the junction area between the aortic arch and descending aorta (width of the orifice: 4.04mm, velocity: 3.50m/s, mean pressure gradient: 24.6mmHg, max pressure gradient: 49mmHg) (Table 1).

The second-stage plastic operation of aortic coarctation was performed on 2015.11.04. The constrictive segment was broadened with a piece of vascular prosthesis patch after removing a septum in the descending aorta. The patient recovered successfully and reexamination of the Holter monitoring and TTE after the operation. The Holter monitoring showed sinus rhythm with heart rate of 84bpm to 98bpm. There was a total of 124410 heart beats during 24 hours and without ventricular arrhythmias (Table 1). The TTE showed that (Figure 2B e, f) 1) LA: 39mm LV: 39mm EF: 55%; 2) the parachute mitral valve was removed and replaced by an artificial valve, which function was normal; 3) the abnormal muscular septum in the middle of left ventricular was removed, and the left ventricular returns to one chamber; 4) bicuspid aortic valve without stenosis or regurgitation; 5) release of aorta coarctation (velocity: 1.56m/s) (Table 1).

6. Follow-up

During the follow-up (on 2016.01.20), the patients did not suffer syncope, pre-syncope, palpitation or dyspnea. The Holter monitoring showed that the ventricular arrhythmias almost vanished (Table 1). Moreover, the TTE showed that 1) LA: 39mm LV: 43mm EF: 60%; 2) the parachute mitral valve was removed and replaced by an artificial valve, which function is normal; 3) the abnormal muscular septum in the middle of left ventricular was removed, and the left ventricular returns to one chamber; 4) bicuspid aortic valve without stenosis or regurgitation; 5) release of aorta coarctation (velocity: 1.85m/s) (Table 1).

7. Discussion

The patient suffered from AMC ventricular arrhythmias, Shone's

complex and double-chambered left ventricle. To the best of our knowledge, it is the first case reported in the literature so far.

7.1. Treatments for the AMC ventricular arrhythmias

We were concerned about the AMC ventricular arrhythmias in this case and hoping to treat it. The radiofrequency ablation for the AMC ventricular arrhythmias is the first consideration [1]. However, there were lots of obstacles for this procedure. First of all, the aorta coarctation was impeding the ablation catheter to reach the AMC region along the aorta retrogradely. Secondly, the orifice area of parachute mitral valve is only 1.68cm^2, which also was obstructing the ablation catheter to reach the origin of the ventricular arrhythmias anterogradely. Finally, the abnormal muscular septum in the middle of left ventricle would hinder the manipulation of the catheter by the operator. Therefore, we concluded that radiofrequency ablation was not very feasible before correcting the heart malformation.

Meanwhile, the AMC region, located between the left coronary cusp and mitral valve (anterior leaflet), might be surgically removed during the artificial valve implantation. In fact, the AMC ventricular arrhythmias were eliminated along with the removal of the parachute mitral valve after the first operation. That was an operation to kill two birds with one stone. If the AMC ventricular arrhythmias were not eliminated after the first operation, we will consider radiofrequency ablation after release of aorta coarctation.

7.2. Complex Cardiac Malformation: Shone's Complex and Double-Chambered Left Ventricle

Shone's complex is a rare and severe type of congenital heart disease. It is a series of four obstructive or potentially obstructive left-sided cardiac lesions (supravalvular mitral ring, parachute mitral valve, subaortic stenosis, and coarctation of the aorta), which includes the complete form (all four lesions) and incomplete forms (2-3 lesions) [2]. From the literature reports [3-5], no other levels of left-sided cardiac obstruction were found in the Shone's complex. Our case demonstrated an incomplete Shone's complex, including parachute mitral valve, bicuspid aortic valve and the aorta coarctation, accompanied by double-chambered left ventricle in a female adult. Shone's complex and double-chambered left ventricle are both rare cardiac anomalies in clinical practice, with the former often being diagnosed in neonatal period or childhood [6] and the latter being characterized by the separation of the left ventricle into two chambers by abnormal septum or extra muscle bundle [7]. It is the first time to find these two anomalies coexist in an adult patient. Our results of TTE showed that the abnormal muscular septum located in the middle of left ventricle, originated in the middle of posterior-inferior wall and posterior ventricular septum, extending to the left ventricular chamber and dividing it into two chambers. Moreover, we noticed the blood flow accelerates in left ventricle when it is passing through the abnormal muscular septum defect. Therefore, from another point of view, the double-chambered left ventricle may also represent as another level of obstructive left-sided cardiac lesion (Left ventricle obstruction). And this case may possibly expand the definition of Shone's complex (a five-leveled left-sided cardiac obstruction: supravalvular mitral ring, parachute mitral valve, double-chambered left ventricle, subaortic stenosis and aorta coarctation).

Surgery is currently the first line treatment for the Shone's complex. Indications for mitral valve repair are abnormal mitral valve (parachute mitral valve) with less than 2cm^2 orifice area, mean resting end-diastolic gradient of >5mmHg, presence of supravalvular ring of the left atrium and associated left ventricular outflow obstructive lesions [8]. Meanwhile, indications for intervention for the aorta coarctation are peaktopeak gradient≥20mmHg, significant anatomic evidence of narrowing on imaging with extensive collateral flow, presence of systemic hypertension, additional cardiac defects and/or single ventricular physiology, left ventricular hypertrophy and elevated left ventricular end diastolic pressure [9]. In this case, the abnormal mitral valve (parachute mitral valve) was less than 2cm2 orifice area, and the aorta coarctation peaktopeak gradient was more than 20mmHg. Therefore, both of these two lesions should be treated. Due to the high risk of long-time surgery on general anesthesia and cardiopulmonary bypass, we decided to perform a staged operation. As we know, the degree of left ventricular inflow tract obstruction is the predominant factor determining outcome in Shone's complex [10]. Therefore, in the first operation, we confirmed and addressed the parachute mitral valve and the abnormal muscular septum in the left ventricle. And in the second one, interventions to the aorta coarctation were performed.

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