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Case Report

Multimodality imaging of cor triatriatum dexter complicated with hypertrophic cardiomyopathy of restrictive phenotype ☆,☆☆

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ABSTRACT

Cor triatriatum dexter (CTD) is an extremely rare congenital cardiac malformation in which a membrane divides the right atrium into 2 chambers. Hypertrophic cardiomyopathy (HCM) with restrictive phenotype is also a rare cardiomyopathy. We report a case with an 18-year history of chest discomfort, fatigue and syncope following intense physical activity was finally diagnosed with CTD complicated with HCM, and the HCM is a special type, restrictive phenotype. Multimodal imaging was used to diagnose this complex disease and analyzed the main cause of her heart failure, which provided accurate evidence for clinical treatment and prognosis.

Introduction

Cor triatriatum dexter (CTD) is an extremely rare congenital cardiac malformation in which a membrane divides the right atrium into 2 chambers [1]. It is frequently associated with right-sided cardiac abnormalities, such as pulmonary artery stenosis or atresia, hypoplastic right ventricle, Ebstein anomaly, and atrial septal defect (ASD) [2]. CTD has varying clinical manifestations, depending on the degree of partitioning or septation of the RA, from completely asymptomatic to severe right-sided heart failure.

Hypertrophic cardiomyopathy (HCM) is the most common genetic cardiac disease with an estimated prevalence of 0.2% in the general population [3,4]. But HCM with restrictive phenotype is a rare variant, which performance for enlarged atria with mild to moderate LV hypertrophy, moderate myocardial fibrosis, and usually with mild to moderate pericardial and/or

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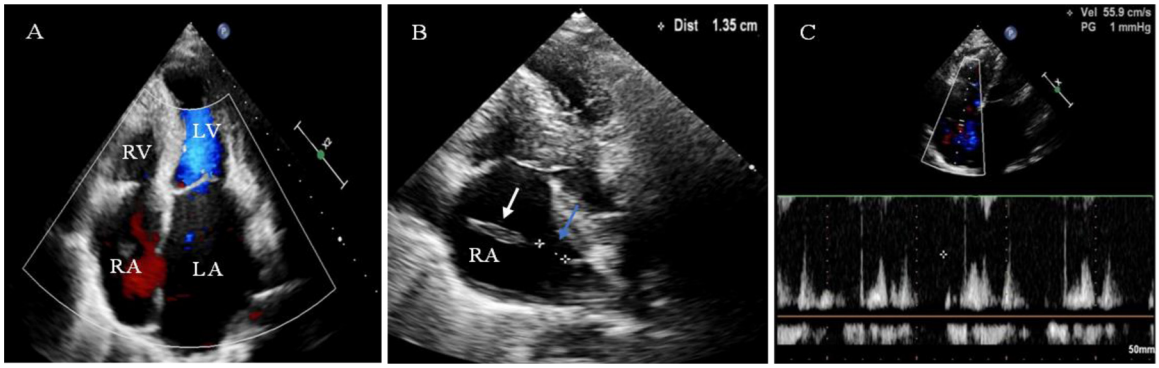

Fig. 1 – picture A showing both right and left atria (RA and LA) were enlarged, and asymmetric left ventricular hypertrophy, picture B showing a membranous partition(White arrow) in the right atrium with the defect(Blue arrow), picture C showing the blood flow through the defect.

pleural effusion. It has not only severe clinical symptoms but also a poor prognosis [5].

In this report, we present the case of a young woman with heart failure who suffering from both CTD and HCM for decades. To the best of our knowledge, the combination of cor triatriatum dexter and HCM is reported for the first time. Non-invasive imaging multimodalities are extremely crucial and allow the best guarantee for the accurate diagnosis and tissue characteristics of such complex heart diseases, particular in such rare disease. And it is particularly important to analyze the main causes of heart failure for clinical treatment decisions.

Case report

A 25-year-old female with an 18-year history of chest discomfort, fatigue and syncope following intense physical activity was initially diagnosed with hypertrophic cardiomyopathy (HCM) by transthoracic echocardiography (TTE) (no image was available). For further assessment, she was referred to our hospital. The ECG demonstrated sinus rhythm with P-wave abnormalities, incomplete right bundle branch block and right ventricular hypertrophy. Evaluation with TTE showed both the right and left atria were enlarged (left atrium, 55mm × 64 mm; right atrium, 47mm × 71mm; left ventricle,30 mm; right ventricle,31 mm), there was also asymmetric left ventricular hypertrophy (interventricular septal,16-17mm) without obstruction of biventricular outflow tracts (Fig. 1). Interestingly, a membranous partition measured 28mm was detected in the right atrium (RA), which divided RA into 2 chambers connected by defect with a diameter of 13.5mm (Fig. 1), and the forward flow velocity through the defect on the membrane was 55.9cm/s (Fig. 1). Cardiac computed tomography angiography (CCTA) confirmed the presence of membrane partition in RA (Fig. 2) and ruled out other complex cardiac anomalies. Further cardiac magnetic resonance imaging (CMR) cines and 3D magnetic resonance angiography (MRA) indicated the detailed morphologic information of this squashed, hourglass-shaped membrane across RA extending from upper part of the atrial septum to the lower lateral wall of the right atrium. The superior margin of membrane was located behind the right auricle and adjacent to the superior vena cava orifice, the lower margin was located in front of the coronary sinus extending to the right atrial free wall (Fig. 3). However, the 2D velocity-coded flow imaging indicated that there was no hemodynamic obstruction caused by membranous structure (Fig. 3). In addition, as suggested by TTE, multiple view cines also showed asymmetric hypertrophy of the ventricular septum and significant enlargement of atria (Fig. 3). Further first-pass perfusion at rest showed there was no sign of ischemia in the left ventricular myocardium (Fig. 3), patchy late gadolinium enhancement was visualized in the septum and inferior insertion (Fig. 3), and myocardial T1 and ECV increased significantly. The fluorodeoxyglucose (FDG)-positron emission tomography (PET) indicated an abnormal myocardial perfusion/metabolism of LV with suspicion of cardiomyopathy.

Discussion

Based on the above examinations, the patient was finally diagnosed with cor triatriatum dexter complicated with hypertrophic cardiomyopathy which was confirmed by myocardial biopsy.

Cor triatriatum (or triatrial heart) is rare congenital heart anomaly with a prevalence of 0.1%-0.4% in all congenital heart diseases [1,2]. Left atrium is predominantly involved, which is also called cor triatriatum sinistrum (CTS). Cor triatriatum dexter (CTD) is rarer than the CTS with only about 100 cases have been reported in the Pubmed so far, which the right atrium is divided by persistence of the right valve of the sinus venosus into 2 chambers, with the caval veins and coronary sinus on one side, and the tricuspid valve and right atrial appendage on the other [6]. Abnormal right valve are divided into 2 categories: one is the right valve remnant with filamentous or reticular connections (called Chiari net); the other is a smooth vein separated from the trabecular part of the right atrium (CTD), this case belongs to the latter.

Due to the rarity of CTD, no definite classification has been found. The clinical manifestations of the CTD depend on the presence of perforations in the fibromuscular mem-

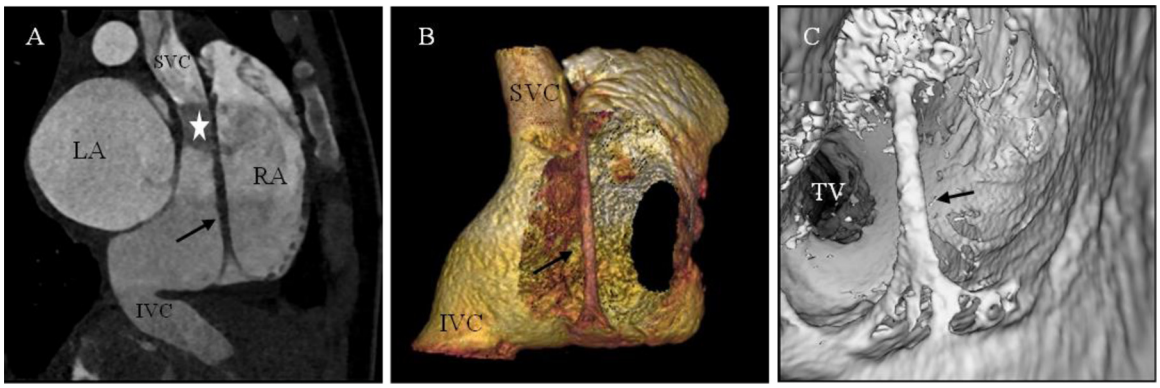

Fig. 2 – The multiple planar reformation(A), volume rendering(B) and CT virtual endoscopy(C) showed the membrane partition (Black arrow) in RA and slow blood flow (White star) in superior vena cava (SVC). inferior vena cava; TV, tricuspid valve.

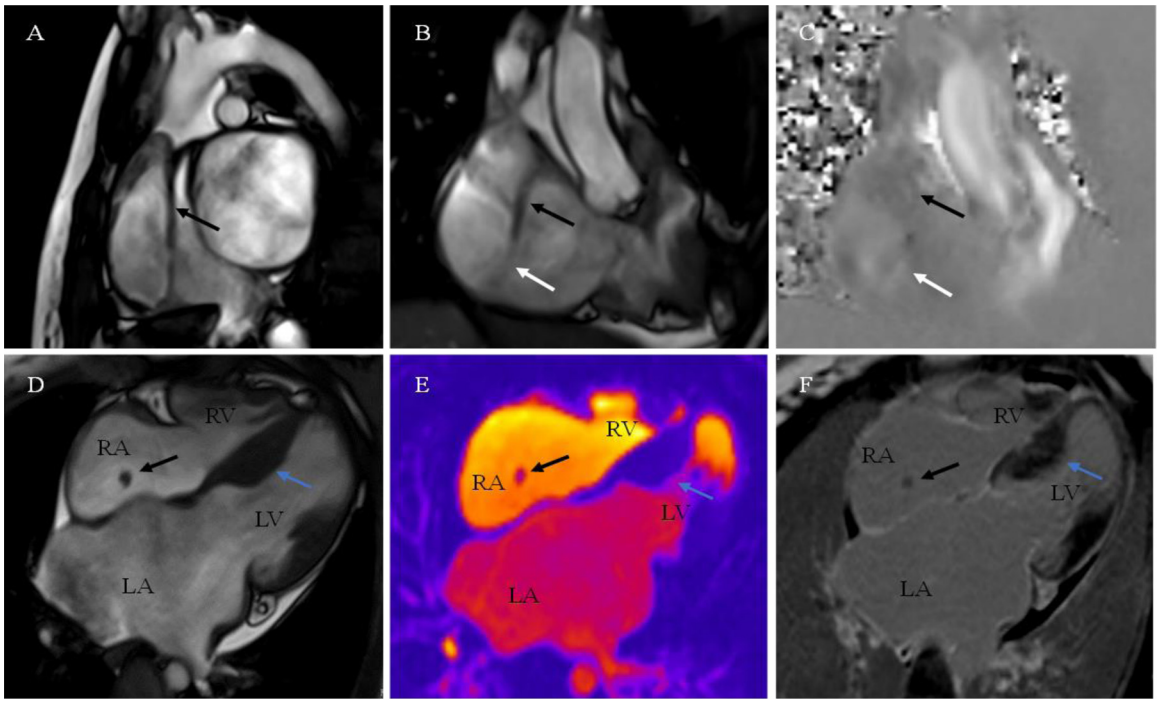

Fig. 3 – picture A, B and C showing the membrane (Black arrow) and no hemodynamic obstruction (White arrow). picture D showing asymmetric symmetric hypertrophy of the ventricular septum and significant enlargement of atria. First-pass perfusion at rest(E) showed there was no sign of ischemia in the left ventricular myocardium, patchy late gadolinium enhancement(F) was visualized in the septum and inferior insertion.

brane, the size of the perforations, right-to-left shunt, and associated malformations [7,8]. When the septation is mild, it often is asymptomatic and is discovered by chance during surgery to correct other cardiac abnormalities, cardiac examinations, or postmortem examinations [9]. But more-severe septation can cause right sided heart failure and elevated central venous pressure, secondary to obstruction of the tricuspid valve, the right ventricular outflow tract, or the inferior vena cava [10].

There are several diagnostic modalities have been found to be useful in diagnosing CTD, such as transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), cardiac computed tomography angiography (CCTA), catheter angiography (CAG), and cardiac magnetic resonance imaging

(CMR). TTE is very helpful in making a definitive diagnosis and the procedure of choice to detect CTD and associated cardiac malformations [11]. It further helps in excluding the lesions leading to right ventricular inflow obstruction. Color flow Doppler helps in the location and estimation of the size of communication between the proximal and the distal chambers. Pulsed wave Doppler is used to estimate the maximum/mean pressure gradient and a velocity across the orifice [12]. TEE helps in confirmation of membrane in patients with poor transthoracic window [13]. 3D echocardiography with matrix array probe clearly delineates the orifices 'shape and size [14]. CAG is now rarely used to diagnose intracardiac structural abnormalities, but it could be useful in diagnosing artery pressure and vascular resistance. CCTA and CMR can effectively establish the presence of interatrial membrane and associated malformations [15]. CCTA provides better spatial resolution and multiplanar image reconstructions, allowing improved delineation of cardiac structures and cardiac masses [16]. CMR provides superior tissue contrast and associated blood flow turbulence across the membrane.

Hypertrophic cardiomyopathy (HCM) is the most common genetic cardiac disease, which occurs in 1 of 500 adults and is inherited as an autosomal dominant, is the most common cause of sudden cardiac death (SCD) in children and adolescents [17]. The clinical manifestations are heterogeneous but are a result of the following: (1) left ventricular outflow tract (LVOT) obstruction, (2) diastolic dysfunction, (3) myocardial ischemia, and (4) supraventricular and ventricular arrhythmias [18].

Diastolic dysfunction is considered as one of the most important pathophysiologic consequences of HCM because of disturbed calcium kinetics, ischemia, LV hypertrophy, and fibrosis [19]. If the MRI features only include mild to moderate LV hypertrophy, nonobstruction of LV outflow tract with enlarged atria, and ventricles of normal or small size with moderate myocardial fibrosis, we consider it to be HCM with restrictive phenotype [5]. According to Kubo's research, this particular HCM with restrictive phenotype accounts for about 1.1% of all HCM [20]. In the current study, the diameters of both atria in restrictive phenotype were markedly larger than those in typical patients with HCM, which may reflect the manifestation of heart failure with preserved ejection fraction. These patients often have severe clinical symptoms of impaired cardiac function and a higher incidence of atrial fibrillation, which may result in a poor prognosis.

At present, the diagnosis of HCM mainly relies on echocardiography and CMR. TTE and TEE as initial screening methods for HCM, they can not only observe the degree of cardiac hypertrophy, but also allows estimation of the subaortic pressure gradient and often reveals mild to moderate mitral regurgitation. CMR is a more reliable imaging modality compared with echocardiography to detect myocardial hypertrophy and aneurysm formation, which not only provides structural and functional information but also can provide histological features, such as LGE, corresponding to myocardial fibrosis [21].

In general, for complex heart diseases, especially if it's so rare, multimodality imaging are very necessary. And with these tests we know that, there was no obstruction of inferior vena cava due to the large diaphragm orifice, the membrane partition in this case had no hemodynamic significance. Therefore, we can assume that the chest discomfort, syncope and arrhythmia were mainly caused by HCM.

Availability of data and materials

Data has been presented in the text.

Author Contributions

Li Liang contributed to the conception and design and acquisition of data, as well as participate in drafting and revision of the manuscript., Min-Jie contributed to idea design and critically revised the manuscript and as well as approved the final submission of manuscript.

Patient consent

Patient has given both verbal and written informed consent to publish the case including publication of images.

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