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Congenital Gerbode Defects Complicated by Infective Endocarditis: 2 Case Reports

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ABSTRACT

Gerbode defect is a rare shunt between the left ventricule and right atrium. The etiology is typically congenital. The infravalvular type is the most common. The congenital defects are believed to close by forming an aneurysmal pouch through incorporating adjacent tricuspid valve tissue. Endocarditis is responsible for this shunt by re-opening the defect. Diagnosis is based on the transesophageal echocardiography. Surgical closure demonstrated an excellent outcome. We present 2 cases with this uncommon congenital shunt complicated by infective endocarditis and septic embolism.

KEYWORDS: Gerbode Defect, Infective Endocarditis**,** Transesophageal Echocardiography

I. INTRODUCTION

The Gerbode defect is a communication between the left ventricle and right atrium. It is usually congenital but can be acquired, secondary to infectious endocarditis, myocardial infarction, trauma or after cardiac surgery. This shunt is associated with high risk of infective endocarditis.

CASE 1

A 46-year-old man with a history of unoperated congenital heart disease, who presented to hospital with a three-month history of intermittent fever and night sweats. 2 weeks before admission, he described a sensation of heavy legs associated with dorso-lumbar pain. On physical examination, a loud ejection murmur was heard most prominently at the left sternal edge. At our hospital, transthoracic echocardiogram demonstrated a perimembranous ventricular septal defect with a shunt from the left to the right ventricle and a communication between the left ventricle and right atrium (Gerbode defect), and showed a mobile vegetation attached close to the septal leaflet of the tricuspid valve with a severe tricuspid regurgitation and laminar flow. Transesophageal echocardiogram was performed and confirmed the diagnosis (Figure1). Blood cultures returned positive for streptococcus constellatus and antibiotic therapy was initiated. Magnetic resonance imaging of the dorso-lumbar spine was performed and revealed a L4-L5 spondylodiscitis (Figure2).

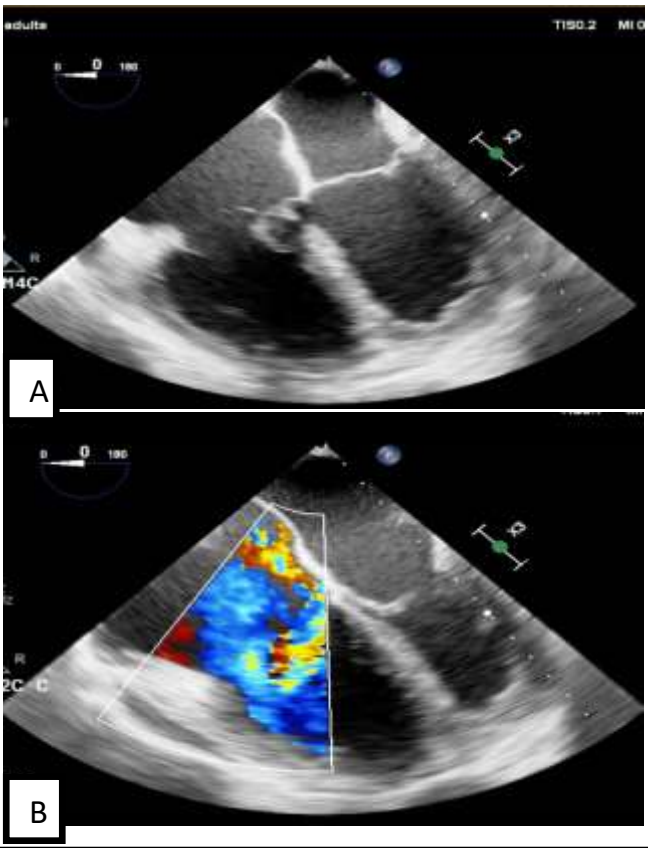

Figure 1: **A**: Transesophageal echocardiogram image showing a comminucation between the left and the rignt ventricle with a mobile vegetation attached close to the septal leaflet of the tricuspid valve with a severe tricuspid regurgitation. **B** : TOE in 4-chamber view showing a shunt between the LV and RA (Gerbode Defect) and a shunt between the LV and RV (case 1).

We noted a clinical and biological improvement by week 6 of intravenous antibiotic therapy (vancomycin with gentamicin).

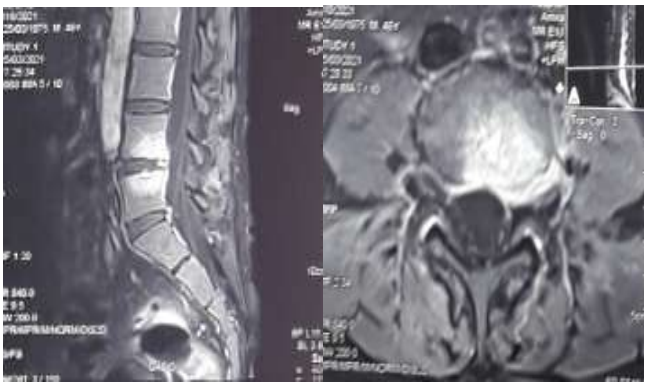

Figure 2: Bone MRI image showing pathological signal between L4 and L5 (case 1).

CASE 2

A second case involved a 23-year-old man who presented to hospital with a 2 months history of fever, dyspnea and palpitations. In his clinical history, he reports a notion of unoperated congenital heart disease. He was recently diagnosed with left knee septic arthritis and during his hospitalization in rheumatology department; we discovered a systolic heart murmur heard at the left sternal edge with murmurs include aortic, mitral and tricuspid regurgitation.

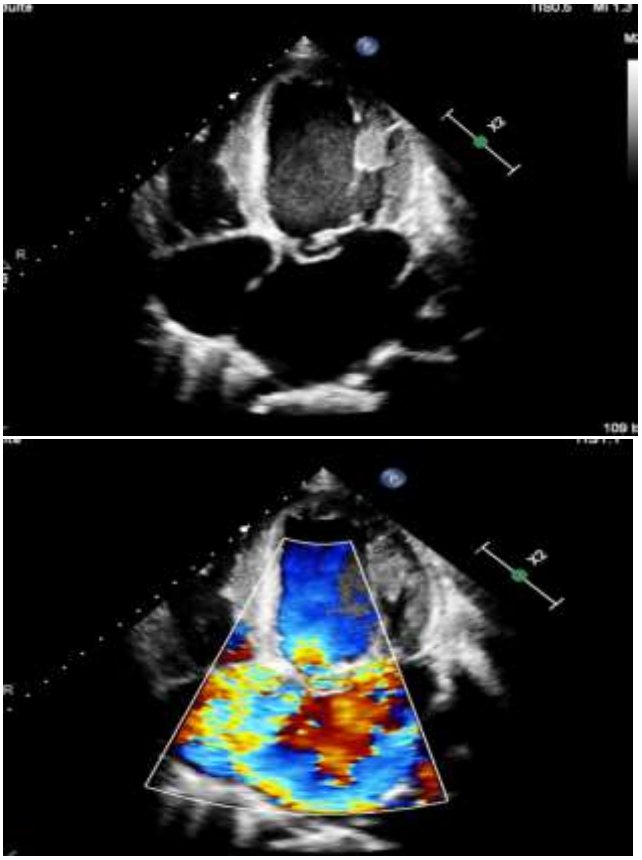

Figure 3: TTE in 4-chamber view showing ruptured anterior mitral valve aneurysm with a mild to severe regurgitation (case 2).

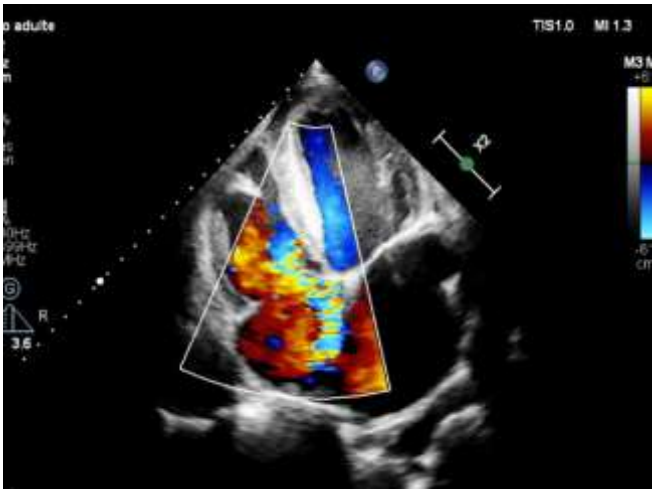

Figure 4: TTE in 4-chamber view showing the shunt between the LV and RA (case 2).

A transthoracic echocardiogram was done on admission at our hospital showing a shunt between the left ventricle and right atrium (Gerbode defect) with several vegetations on aortic valve complicated by a severe aortic regurgitation characterized by an eccentric jet directed toward the anterior mitral leaflet (Figure4). A ruptured anterior mitral valve aneurysm with a mild to severe regurgitation was also detected with a large mass measuring 18 mm (figure3). Another mobile vegetation was seen attached to the septal leaflet of the tricuspid valve with severe tricuspid regurgitation. The likelihood of IE was strengthened by 2 positive blood cultures for *Staphylococcus epidermidis*. Targeted antibiotic therapy (vancomycin and gentamicin) was started. The body CT scan was performed showing a bilateral septic pulmonary embolism. After one month of antibiotic therapy, the patient presented with global heart failure. Unfortunately, the patient died intraoperatively.

II. DISCUSSION

The congenital LV-RA shunt was first mentioned in an autopsy report in 1838 [1]. In 1958, five patients with this anomaly were successfully operated by Gerbode et al. [2] and named it 'Gerbode defect'. The frequency of this anomaly is only 0.08% [3] of intracardiac shunts and less than 1% of all congenital cardiac defects [4].

The defects occur in the AV septum and are known as supravalvular defects in approximately 1/3 of patients [5]. 2/3 of this anomaly occur between the ventricles and are known as infravalvular defects. This classification was elaborated by Sakakibara and Konno [6], and they include a third type with both supravalvular and infravalvular components, referred to as intermediate defects. The incidences of the three types accounted for 76%, 16%, and 8% of the total, respectively [7].

Taskesen et al. [3] and Sinisalo et al. [8] categorized the supravalvular defects as type 1 and the infravalvular defects as types 2 and 3 [9]. The types 2 and 3 are the most common

forms, which have many variants to septal leaflet anomalies of the TV [10]. These variants include a cleft, widened commissural space, abnormal chordae, perforation [8]. The defects are believed to close by forming an aneurysmal pouch (aneurysmal transformation) through incorporating adjacent TV tissue.

Endocarditis is responsible for LV-RA shunt by re-opening a congenital defect, widening a small, insignificant shunt or by destructive perforation of the septum [11]. For endocarditis patients with fever and septicemia, these general symptoms may mask a new shunt, making it easy to miss [8]. The staphylococcus aureus (41%) and Streptococcus species (17%) are the most common bacteria [3]. The LV-RA shunts in association with VSDs increases the risk of endocarditis (58 per 10,000 patient-years) in comparison to typical VSDs or mitral regurgitation (5.2 per 10,000) [3].

Depending on the volume and duration of the LV-RA shunt, the Gerbode defect may be asymptomatic or manifested by signs of severe heart failure, [8]. The small shunts are usually asymptomatic [3]. The physical examination finds a characteristic murmur similar to that of a VSD: loud, harsh pansystolic, grade III to VI, unvarying with respiration and often associated with a thrill along the left sternal border [8]. The two-dimensional transesophageal echocardiography (TEE) is the procedure of choice for the diagnostic of the Gerbode defect [12]. It is the most sensitive method for detecting LV-RA shunts [7], especially in patients with prosthetic heart valves [3]. This shunt is highly likely when echocardiographic interrogation, in addition to the physical examination, reveals an unusually dilated right atrium [13]. The color flow Doppler reveals usually a characteristic systolic flow disturbance within the right atrium [14]. This flow originates from the membranous septum, in contrast to tricuspid regurgitation, which originates from the valve. An important clue to their presence, however, is a systolic fluttering of the tricuspid valve best seen with M-mode echocardiography [15]. Continuous wave Doppler interrogation through this jet typically demonstrates an unusually high systolic velocity (>4 m/sec) originating from the upper membranous septum and directed toward the RA reflecting the gradient between the high pressure left ventricle and the low pressure right atrium [16]. This characteristic is highly suggestive of a LV-RA shunt and must be distinguished from other conditions such as ruptured sinus of Valsalva aneurysms, endocardial cushion defects, VSD, and TR [3]. Silbiger et al. [5] specified several key echocardiographic clues suggesting Gerbode defect, including: (1) atypical jet direction, (2) persistent shunt flow into diastole, (3) lack of ventricular septal flattening, (4) no right ventricular hypertrophy, and (5) normal diastolic pulmonary artery pressure as estimated from the pulmonic regurgitant velocity.

The locus of the high systolic flow from the membranous septum helps to distinguish Gerbode defect from TR. Severe pulmonary arterial hypertension (PAH) will be erroneously diagnosed if this jet is mistaken for tricuspid regurgitation [5]. The diastolic pulmonary arterial pressure identified from the pulmonic regurgitation jet is helpful in diagnosis of true PAH from the high velocity jet in the right atrium caused by Gerbode defect [12]. Gerbode defects typically produce a shunt from the LV to RA in systole, while ruptured sinus of Valsalva aneurysms will, in addition, cause diastolic shunting because of the diastolic gradient that exists between the aorta and right atrium [17].

The treatment of Gerbode defect depends on severity of symptoms [3,7]. Asymptomatic, small or chronic defects can be managed conservatively [13]. Long-term follow-up results concluded that a small fraction of the LV- RA shunts close spontaneously, while a few develop infective endocarditis [7].

Toprak et al. [18] proposed suggested that asymptomatic patients with insignificant intracardiac shunt with no right ventricular volume or pressure overload due to a small LV-RA shunt be kept under close follow-up rather than undergo surgery [7]. Conversely, Yacoub et al. [19] suggested that all Gerbode defects be repaired, regardless of their size to preclude infective endocarditis. Surgical closure demonstrated an excellent outcome and recommended for closure of all direct Gerbode defects [13]

III. CONCLUSION

We have presented two cases of congenital Gerbode defect remained asymptomatic and complicated by infective endocarditis. The gerbode defect is a rare congenital shunt. The TTE remain the procedure of choice for the diagnostic. Surgical treatment is always feasible with excellent outcome in order to avoid complications especially infective endocarditis.

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