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Acquired Gerbode Defect Secondary to Severe Bicuspid Aortic Valve Endocarditis

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Abstract

An annular aortic abscess is a fatal complication of infective endocarditis. Echocardiography is the initial imaging modality to confirm the diagnosis in suspected infective endocarditis. Here, we present a case of a bicuspid aortic valve infective endocarditis caused by *Staphylococcus aureus* and complicated with annular aortic abscess resulting in acquired Gerbode defect (type II) with tricuspid valve vegetation which was undiagnosed preoperatively. The intraoperative transoesophageal echocardiography yields a new finding prior to the surgical incision, which impacted the clinical decision-making and increased the burden of the procedure.

Keywords

Acquired Gerbode Defect, Infective Endocarditis, *Staphylococcus aureus*, Bicuspid Aortic Valve

1. Background

Gerbode defects are rare congenital cardiac anomalies that account for less than 1% of all congenital cardiac abnormalities and only 0.08% of intracardiac shunts [1]. It is defined as abnormal shunting between the left ventricle and right atrium resulting from either a congenital defect or prior cardiac insults (**Figure 1**). The pathophysiology underlying the development of Gerbode defect is a disease process that injuries the atrioventricular septum and leads to the abnormal shunting of blood. Although the most common cause of Gerbode defect has historically been congenital, an increasing trend towards acquired cases has recently been reported in the literature.

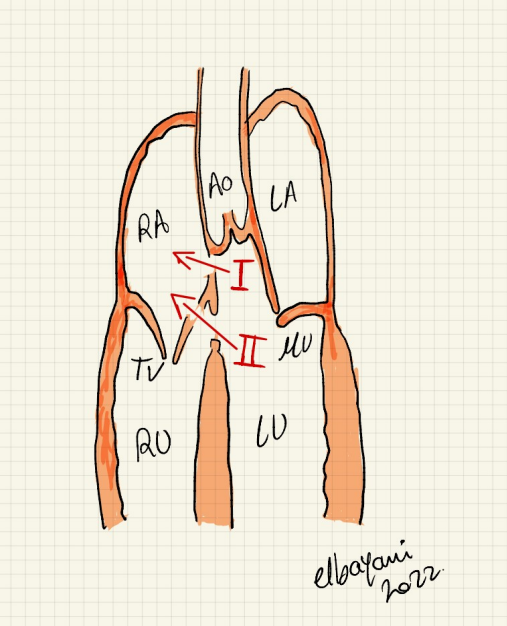

Figure 1. Schematic representation of Gerbode defect types. Defect number one (I) is the supravalvular (direct) type. Defect number two (II) is the infravalvular (indirect) type. LA—left atrium, LV—left ventricle, RA—right atrium, RV—right ventricle, TV—tricuspid valve, MV—mitral valve, AO—aorta.

2. Case Presentation

A 38-year-old white, non-smoker male (1.83 m, 75 kg, BSA 1.9 m^2, BMI 22) presented to the Emergency Department (ED) in secondary healthcare facility complaining of fever, cephalgia, and delirium. He denied intravenous illicit recreational drug use. He had no history of known congenital heart disease. Prior to his current illness, there was no administration of any immunosuppressive medications. He had no physical limitations and regularly engaged in sports activities.

On presentation, blood pressure was 120/90 mmHg, heart rate 120 beats per minute, respiratory rate of 21 per minute, temperature 38,9 degrees Celsius using ear thermometer, and oxygen saturation of 94% on ambient air. The arterial blood gas analysis was unremarkable except for hypokalaemia (K * 3 mmol/L). Physical examination was remarkable for a soft diastolic murmur; Lung auscultation revealed bibasilar crackles. Neck veins were non-distended, and lower extremity edema was not detected. Due to the febrile infection, the patient was admitted to the COVID ward for isolation. SARS-COV-2 infection could not be seen in the PCR tests performed so that the patient could be a few hours later de-isolated.

2.1. Investigations

Initial laboratory test results were remarkable for white blood cell count of 11.5 × 10^3/ul (4 - 10 × 10^3/ul) with elevated neutrophils, platelets of 64 × 10^3/ul (150 - 400 × 10^3/ul), C-reactive Protein of 26.7 mg/dl (0.0 to 0.8 mg/dl), procalcitonin

of 9.8 ng/ml (0 - 0.5 ng/ml) and abnormal liver function with elevated total Bilirubin 3.1 mg/dl (<1.23 mg/dl). Renal function tests were within normal limits. Four sets of blood cultures were initially positive for gram-positive round-shaped cocci, which were identified as *Staphylococcus aureus* later (Methicillin-sensible, MSSA). Abdominal sonography showed mild splenomegaly. An electrocardiogram showed normal sinus rhythm.

Because of the medical health providers' arousal towards the pandemic and the patient's vaccine opponent attitude, a low dose CT scan was performed to exclude pneumonia, which revealed no typical COVID-19 changes in the lungs.

Transthoracic echocardiography revealed a sclerosed aortic valve with bicuspid morphology, severe aortic valve insufficiency with no evidence of vegetation, and slightly impaired systolic Left ventricular function with an estimated left ventricular (LV) ejection fraction of 45%.

Subsequently, trans-oesophageal echocardiography (TEE) was performed, which confirmed the aortic valve of bicuspid nature with progredient insufficiency. However, there were no endocarditis-susceptible vegetations on any of the heart valves.

Ultimately, definite acute endocarditis was diagnosed according to the Duke criteria, including two blood culture results of *Staphylococcus aureus*; the severe progredient regurgitation of the aortic valve revealed by an echocardiogram; and such clinical and laboratory features as fever, splenomegaly, and heart murmur.

2.2. Management

Initially, the patient was treated empirically for infective endocarditis with intravenous antibiotics. The patient was then transferred to our heart center for surgical intervention. The cardiac catheterization showed no evidence of obstructive coronary artery disease.

Intraoperatively and prior to the surgical incision, the trans-oesophageal probe was introduced, and TEE was performed, which confirmed aortic valve of bicuspid nature, and aortic annular abscess with left ventricular outflow tract defect to the right atrium (LVOT-RA) consistent with Gerbode defect (**Figure 2**).

After median sternotomy, the patient was heparinized with checking activated clotting time. Aortic and bicaval cannulation was installed, and cardiopulmonary bypass was initiated for 141 minutes. The aortic cross-clamp was then applied for 96 minutes. A normothermic blood cardioplegia was administrated through the aortic root, directly through the coronary Ostia, and retrograde through the coronary sinus, which achieved a satisfactory diastolic arrest. Aortotomy was performed, and stay sutures were placed. The aortic valve was inspected, and it was heavily calcified and bicuspid. Cusps were resected, and the annulus was debrided. After resection of the abscessed tissue, a shunt connection from the Left ventricle to the right ventricle entering the right atrium was revealed (**Figure 3**). After right atriotomy, involvement of the tricuspid valve was confirmed. An aneurysm with vegetations was seen at the septal leaflet of the

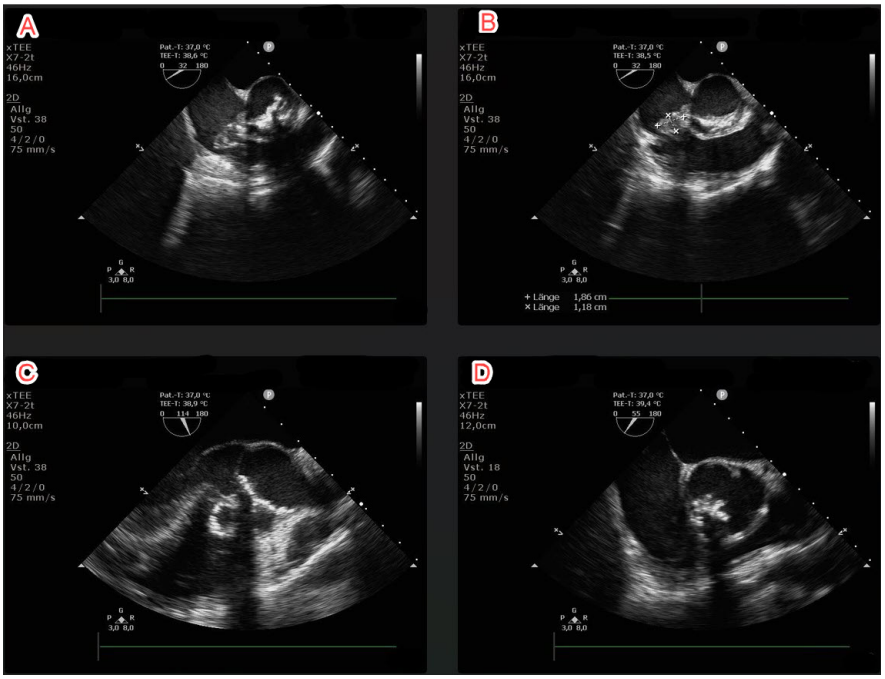

Figure 2. intraoperative trans-oesophageal echocardiography. (A) mid-oesophageal short-axis view demonstrating large vegetation in RA and ruptured/flail leaflet of the bicuspid aortic valve; (B) mid-oesophageal short-axis view shows the dimensions of the vegetation (1.86 cm × 1.18 cm); (C) mid-oesophageal long-axis view demonstrates Gerbode ventricular septal defect; (D) mid-oesophageal right ventricular inflow-outflow view demonstrating the hyperdense vegetation.

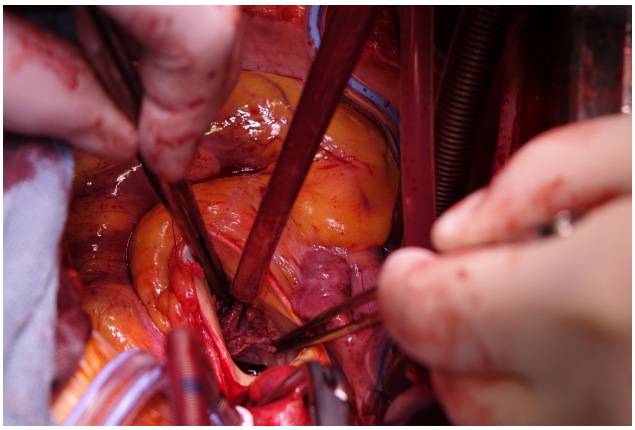

Figure 3. Surgical aortic valve replacement to a mechanical valve prosthesis after debridement of the aortic annulus a septal defect could be detected.

tricuspid valve. Below the tricuspid annulus to the ventricle, a shunt leading to the LVOT could be seen.

Vegetations on the tricuspid valve could be surgically resected, and the integrity of the tricuspid valve could be preserved (**Figure 4**). The decision was already made to proceed with aortic valve replacement with a 23-mm Regent mechanical valve (St. Jude Medical, St. Paul, MN) and primary closure of a septal defect using pledgeted sutures. The right coronary ostium was confirmed to be patent

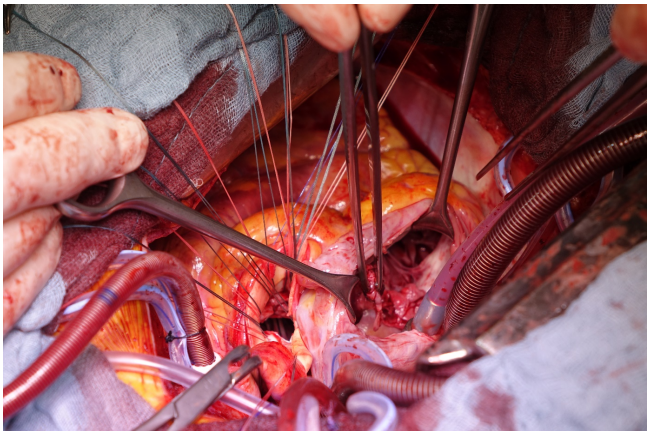

Figure 4. Intraoperative picture where the right atrium was opened, and the vegetations on the septal leaflet were grasped and gently pulled with forceps, preserving the tricuspid valve's integrity.

multiple times during the procedure. The aortotomy was then closed in layers, and the root was vented. The heart was de-aired, and the aortic cross-clamp was removed. The weaning off the cardiopulmonary bypass was performed as usual after the spontaneous return of the heartbeats. Protaminized decannulation was done, and hemostasis was secured. The chest was closed in layers. The patient tolerated all the mentioned procedures well and was monitored in an intensive care unit for 24 hours and then shifted to the surgical ward later for further postoperative care, which was uneventful. The patient was isolated through the whole period of hospital admission is due to the detection of vancomycin-resistant enterococci (VRE) in the rectal and mouth, nose, and throat smear.

The intraoperatively resected valve and vegetation were sent for histopathological and microbiological studies to isolate and identify the pathogen. Histopathological examination of the excised aortic valvular tissue showed fibrous, dystrophic calcification, inflammatory cell infiltration, and bulky vegetation comprised of fibrinous exudative material with areas of neovascularization and areas of granulation tissue. Findings were suggestive of subacute bacterial endocarditis. And the DNA samples from tissues of the aortic valve and the vegetation of the tricuspid valve were both positive for *Staphylococcus aureus* in the Polymerase chain reaction (PCR) test.

The patient had a stable functioning prosthesis with preserved ejection fraction in the follow-up transthoracic echocardiography. He was then discharged resilient and asymptomatic. He successfully recovered post-operatively with inpatient rehabilitation and continuation of intravenous antibiotic therapy.

3. Discussion

Staphylococcus aureus bacteremia (SAB) is an urgent medical problem due to its growing frequency and its poor associated outcome. As healthcare delivery increasingly involves invasive procedures and implantable devices, the number of patients at risk for SAB and its complications is likely to grow. The mechanisms

leading to SAB involve host factors and environmental factors predisposing to infection, whereas the impact of genotypic features on the ability of different strains to cause infection is still controversial [2]. Unfortunately, it is the leading cause of infective endocarditis, and its mortality has remained high despite better diagnostic and therapeutic procedures over time. However, recent studies suggest that valve replacement improves outcome [3]. Transoesophageal echocardiography (TEE) is essential in intraoperatively monitoring adult and congenital heart surgery. It gives us real-time data about the heart during the whole procedure that the adequacy of the repair can be ensured immediately through a review of TEE images directly after surgery; in our case, the intraoperative TEE yield a new finding of the left ventricular outflow tract defect to the right atrium (LVOT-RA), which was not known at the time of the surgery. Therefore, we believe the intraoperative TEE increased the burden of that procedure but helped us make the right decision to deal with the case.

The pathophysiological mechanisms of Gerbode defect usually are reopening of a congenital defect, widening of a small defect, or destruction of the membranous ventricular septum [1]. The Gerbode defect observed in our patient is an acquired defect rather than congenital given the subvalvular location and proximity to the ruptured aortic valve abscess.

The current approach in managing acquired Gerbode defect secondary to IE is surgical repair of the defect as a transcatheter device is not an option due to the presence of infection [4].

The current American Heart Association/American College of Cardiology (AHA/ACC) guidelines do not include a bicuspid aortic valve in a high-risk group category for pre-operative IE antibiotic prophylaxis [5]. However, in a recent study, the bicuspid aortic valve was found to carry a substantially increased risk of IE and intracardiac complications [6].

4. Conclusion

This case highlights the potential of the intraoperative TEE to influence clinical decision-making for cardiac surgical patients. It also illustrates that the use of TEE in high-risk populations can yield a higher incidence of new findings. Furthermore, it emphasizes the need to revisit the infective endocarditis prophylaxis guidelines in the future to include antibiotic prophylaxis in patients with the bicuspid aortic valve.

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The authors would like to acknowledge all who contributed to this case diagnosis and decision-making.

Data Availability Statement

The raw data supporting the conclusions of this article will be made available by the authors without undue reservation to any qualified researcher.

Ethics Statement

The patient signed informed consent related to the clinical course; therefore, the Institutional Review Board was waived due to the retrospective nature of the educational case report.

Authors Contributions

All authors contributed to the patient care, diagnosis, treatment, and authoring this article.

Conflicts of Interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Abbreviation and Acronyms

- TEE Transoesophageal echocardiography
- TV Tricuspid valve
- CT Computer tomography
- LVOT Left ventricular outflow tract
- IE Infective endocarditis
- BSA Body surface area
- BMI Body mass index
- CPB Cardiopulmonary bypass
- RA Right atrium
- DNA Deoxyribonucleic acid