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RESEARCH ARTICLE

GERBODE DEFECT: A VERY RARE COMPLICATION OF INFECTIVE ENDOCARDITIS

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Abstract

Gerbode defects are a very rare anomalies, characterized by direct communication between left ventricle (LV) and right atrium (RA). Most defects are congenital, but they can also be acquired following destructive heart diseases, such as infective endocarditis, myocardial infarction, trauma, or cardiac surgery. In this paper, we report the case of a 35 years-old patient hospitalized for management of an infective endocarditis complicated by a Gerbode defect.

Introduction:-

AGerbode defect (GD) is a very rare type of ventricular septal defect (VSD), described as a direct communication between left ventricle (LV) and right atrium (RA) [1]. Its etiology is complex; most defects are congenital, as a consequence of structural abnormality of the central fibrous body in combination with arrested maturation of the membranous ventricular septum, but there have been reports of acquired defects, following infective endocarditis (IE), myocardial infarction, trauma, or cardiac surgery [2-5].

In this paper, we report the case of a 35 years-old patient hospitalized for management of an IE complicated by a GVSD.

Patient And Observation:-

A35 years-old male patient with a history of poorly managed and undocumented interventricular septal defect, was admitted to the cardiology department. His main complaint was asthenia and anorexia beginning fifteen days ago, without fever.

Physical examination found that his heart rate was 92 beats per minute and blood pressure 126/57 mmHg. His respiratory rate was16 breaths per minute at rest, and O_2 saturation at 100%. During auscultation, a loud holosystolic murmur was heard in all cardiac focal points. There were no signs of heart failure. There were no neurologic or cutaneous findings.Electrocardiogram at admission was normal.

On the electrocardiogram, there were signs in favor of left ventricular hypertrophy. Cardiomegaly was found on the chest X-ray. Biological findings included elevated inflammatory markers (C-reactive protein level at 72 mg/L and ferritin level at 430ng/mL) and hyperleukocytosis (22,640 cells/mm^3), alongside severe inflammatory anemia (hemoglobin level at 3.6 g/dL). Electrolytes and renal function were. Blood cultures found an unspecified strain of Enterococcus.

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A transthoracic echocardiogram (TTE) found a restrictiveGD, 9 mm in length, partially closed by a membranous aneurysm, with a LV-to-RA shunt. There were many vegetations, the largest being 27 mm in length, on the left side of the defect (Figure 1).

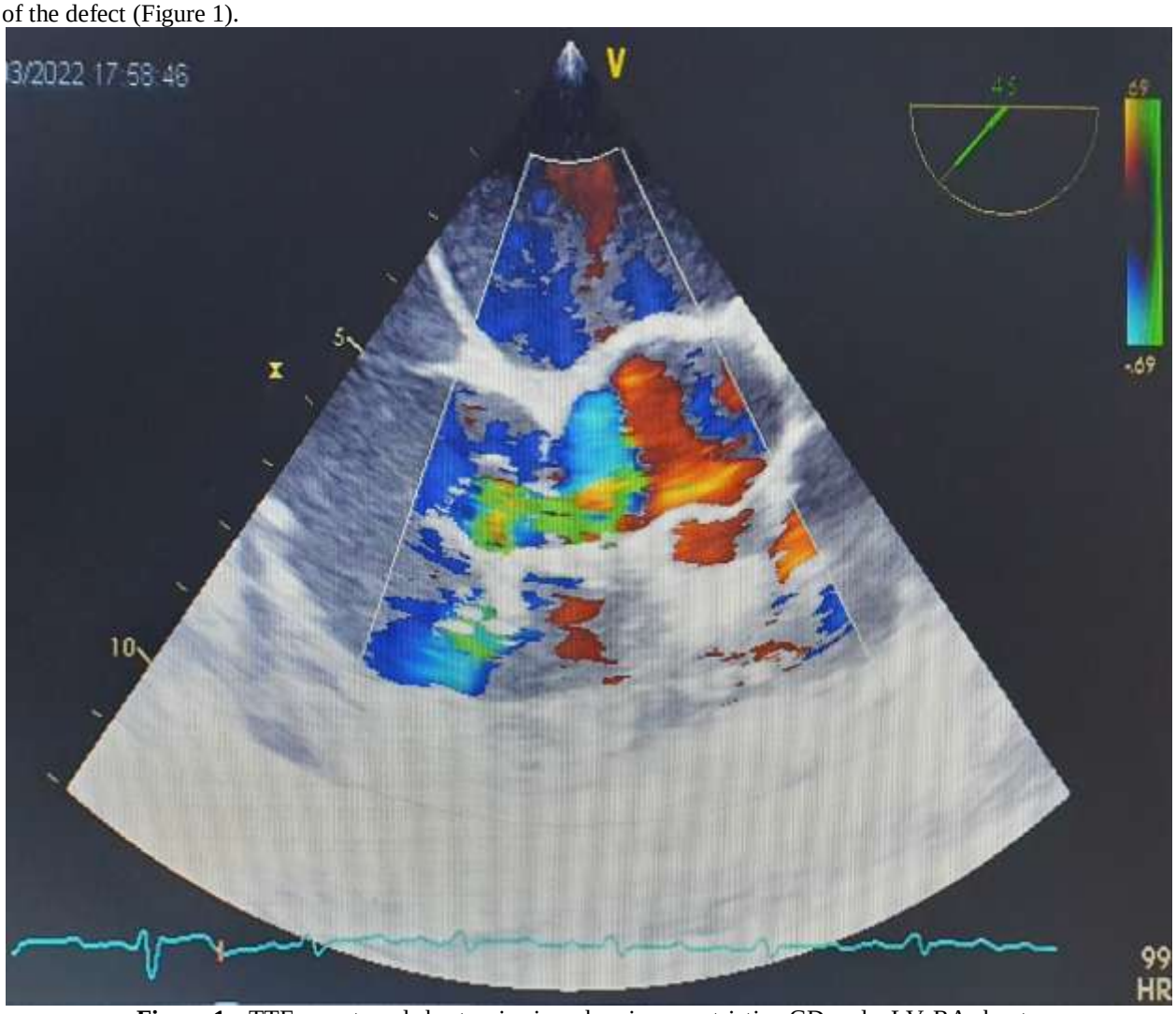

Figure 1:- TTE parasternal short-axis view showing a restrictive GD and a LV-RA shunt.

A transesophageal echocardiogram was then performed, demonstratingthe presence of many vegetations on an aneurysmal pocket of the ruptured membranous interventricular septum, the largest measuring 17 mm, creating a septal defect with a left-to-right shunt. A large element, 25 mm in length, compatible with a vegetation, was found on the tricuspid valve, alongside mild regurgitation (Figure 2).Abdominal and cranial CT scans were performed to look for embolic complications and both returned normal.

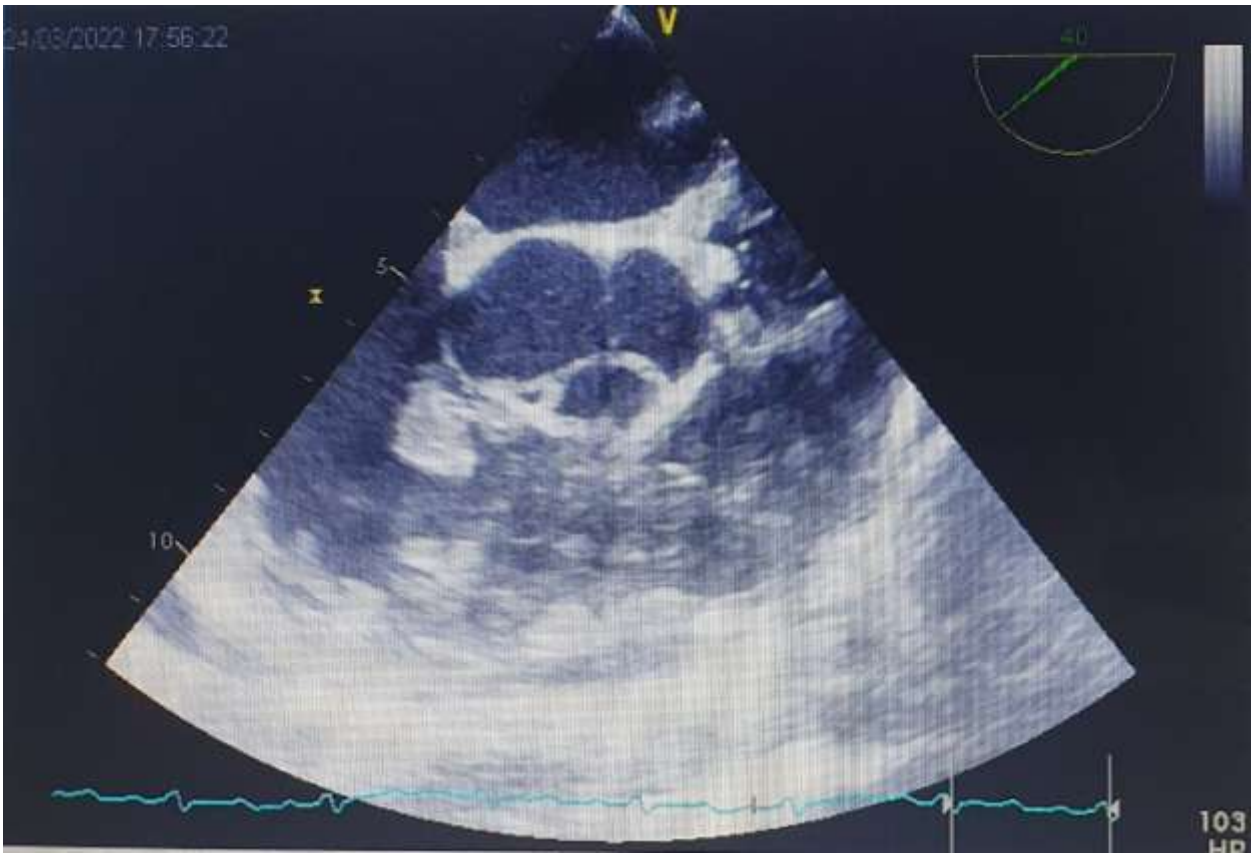

Figure 2:- Transesophageal echocardiogram short-axis view showing many vegetations on an aneurysmal pocket of the ruptured membranous interventricular septum creating a defect with a left-to-right shunt.

The patient received parenteral bi-antibiotherapy, consisting of ceftriaxone for one month and gentamycin for fifteen days. The patient also benefitted from a transfusion of six packed red blood cells.He felt much better after a week of treatment. Biologically, the patient evolved favourably, with C-reactive protein levels and a white blood count reaching 22 mg/L and 8200 cells/mm^3, respectively. Hemoglobin levels also stabilized at 9.6 g/dL.However, a control TTE demonstrated the persistence of vegetations after a month of antibiotherapy. The patient was addressed to cardiovascular surgeons and is scheduled for surgery.

Discussion:-

The LV-RA shunt was first mentioned in an autopsy report in 1838. In 1958, five patients with this anomaly were successfully operated byGerbode et al. [1]. Congenital GDs are very rare, representing 0.08% of intracardiac shunts and less than 1% of all congenital cardiac defects [5, 6].

A classification was proposed by Sakakibara and Konno. In most patients,the defect is infravalvular, between the ventricles. In some cases, the defect is supravalvular, in the AV septum. Intermediate defects include both supravalvular and infravalvular components [8]. The incidences of the three types are 76%, 16%, and 8%, respectively [9].Taskesen et al. [6] and Sinisalo et al. [10] categorized the supravalvular defects as type 1 and the infravalvular defects as types 2 and 3 [11]. Types 2 and 3 are the most common, with many anatomical variants [12].

Endocarditis can cause GDs by re-opening a congenital defect, widening a small, insignificant shunt or by destructive perforation of the septum [13]. For endocarditis patients with fever and septicemia, these general symptoms may mask a new shunt, making it easy to miss [10]. Staphylococcus aureus (41%) and Streptococcus species (17%) are the most common bacteria. GD 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) [6].

Depending on the volume and duration of the LV-RA shunt, the GD may be asymptomatic or manifested by signs of severe heart failure [9,11]. Small shunts are usually asymptomatic [6]. Physical examination finds a characteristic VSD murmur: loud, harsh, pansystolic, and often associated with a thrill along the left sternal border [10].

TEE is the procedure of choice for the diagnostic of GDs. It is the most sensitive method for detecting LV-RA shunts, especially in patients with prosthetic valves [6,9]. A dilated RA is the first indirect sign one should look for [15].Doppler color flowshows a characteristic systolic flow disturbance within the RA, originating from the membranous septum, in contrast to tricuspid regurgitation (TR), which originates from the valve [5]. Systolic fluttering of the tricuspid valve, best seen with M-mode echocardiography, is a common sign [16]. Continuous wave Doppler interrogation through this jet typically demonstrates an unusually high systolic velocity (>4 m/s) originating from the upper membranous septum and directed toward the RA reflecting the gradient between the high-pressureLV and the low-pressure RA [17]. 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 [6].Silbiger et al. specified several key echocardiographic clues suggesting Gerbodedefect, 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 [7].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 RA [18].

Management of GDs depends on severity of symptoms [6,9]. Asymptomatic, small, or chronic defects can be managed conservatively [15].Long-term follow-up results concluded that a small fraction of the LV-RA shunts closes spontaneously, while a few develop IE [9].Toprak et al. suggested that asymptomatic patients with insignificant shunt and no right ventricular volume or pressure overload due to a small LV-RA shunt be kept under close follow-up rather than undergo surgery [19].Conversely, Yacoub et al. suggested that all GDs be repaired, regardless of their size to preventIE [20]. Surgical closure demonstrated an excellent outcome and recommended for closure of all direct Gerbode defects [15].

Conclusion:-

GDs are very rarely encountered in clinical practice. TTE remains the diagnostic method of choice. Small, uncomplicated congenital defects can be managed conservatively, but surgical treatment is an option with excellent outcomes, especially in the case of acquired defects.

Competing interests

The authors declare no competing interest.

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