Spinal abscess in a patient with undiagnosed Gerbode defect: a case report

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Background

Gerbode defect (GD) is a rare cardiac defect in which an abnormal communication occurs between the left ventricle and right atrium. The aetiology is usually congenital but acquired defects can occur.

Case summary

We report on a 47-year-old male with atrioventricular block prior to decompression of an epidural abscess extending from the skull base to the 7th thoracic vertebrae. Following positive blood cultures for Staphylococcus aureus, a transoesophageal echocardiogram performed revealed a small GD with associated endocarditis. In our case, the defect was small and there was no evidence of heart failure, there was little guidance or literature available on how to best manage our patient. A multidisciplinary decision was taken to treat the endocarditis medically and to not close the defect in the acute setting. He recovered well and did not suffer any further cardiac complications. A repeat transthoracic echocardiogram did not reveal any evidence of endocarditis.

Conclusion

Gerbode defects are rare but have been known to increase the risk of developing endocarditis. It is important to have a high clinical suspicion of endocarditis in patients with evidence of conduction disorders and systemic infection.

Keywords

Gerbode defect, Infective endocarditis, Complete heart block, Epidural abscess, Inherited cardiac conditions, Case report

Introduction

A Gerbode defect (GD) is an abnormal communication between the left ventricle and right atrium. Gerbode defect was first described in an autopsy report in 1838 and further investigated by Thurnam in 1838, Buhl in 1857, and Hillier in 1859. It was eventually named after the surgeon Frank Gerbode who published a case series of successful surgical repairs in five patients. Congenital GD are extremely rare and only account for <1% of all congenital heart disorders. Acquired GD can be present following

Learning points

• Have high index of suspicion for endocarditis in patients with evidence of conduction disorders with systemic infection.
• Important to look for abnormal left ventricle to right atrium communications on echocardiography in patients with suspected endocarditis.
• Patients should be followed up regularly to monitor the defect and to assess for complications.
prior cardiac surgery, myocardial infarction, trauma, or associated with endocarditis. Due to the advancement of imaging modalities and increase in awareness amongst clinicians, more acquired GD are being identified in patients.

Here, we describe an atypical presentation of GD associated with infective endocarditis in a patient with conduction abnormalities. Following a comprehensive review of the literature, we have not been able to identify any cases published in which GD was detected in this context.

**Timeline**

| Time | Event |
|------|-------|
| Day 0 | Presentation to emergency department with acute onset lower limb weakness, pyrexia, and lower back pain. Magnetic resonance imaging of spine: collection in anterior and posterior epidural space causing compression of thecal sac. Electrocardiogram (ECG) prior to surgery revealed complete atrioventricular (AV) dissociation prior to surgery however ventricular rate was not impaired. Urgent cervicotomochord decomposition and washout performed. | Patient was noted to be bradycardic post-surgery with ECG again revealing complete AV dissociation and started on noradrenaline. |
| Day 1 | Admission to the intensive care unit (ICU). Blood cultures taken on admission and pus obtained during surgery both positive for *Staphylococcus aureus* and patient started on extended course of antibiotics. | Transoesophageal echocardiogram revealed an abnormal communication between the left ventricle and the right atrium with an associated small vegetation. |
| Day 17 | Transthoracic echocardiogram repeated to assess for endocarditis. Previous vegetation was not appreciated on the scan. | An infective haematogenous spread was suspected as *Staphylococcus aureus* was grown from peripheral blood cultures and pus obtained during spinal surgery. There was no obvious bacterial entry point identified on inspection of the patient, however a formal dental check was not performed. Given the evidence of heart block pre- and post-surgery, the source of infection was suspected to be from the heart. A transthoesophageal echocardiogram performed revealed evidence of GD associated with small vegetation measuring 4.0 × 7.0 mm (Figure 3, Videos 1 and 2) Both atria were mildly dilated but bi-ventricular size and function were normal. There were no additional valve lesions observed and no evidence of an aortic root abscess. |
| Day 3 | Blood cultures taken on admission and pus obtained during surgery both positive for *Staphylococcus aureus* and patient started on extended course of antibiotics. | Discharged from hospital to spinal rehab facility. |
| Day 49 | Completion of antibiotic therapy. No further positive blood cultures. | An infective haematogenous spread was suspected as *Staphylococcus aureus* was grown from peripheral blood cultures and pus obtained during spinal surgery. There was no obvious bacterial entry point identified on inspection of the patient, however a formal dental check was not performed. Given the evidence of heart block pre- and post-surgery, the source of infection was suspected to be from the heart. A transthoesophageal echocardiogram performed revealed evidence of GD associated with small vegetation measuring 4.0 × 7.0 mm (Figure 3, Videos 1 and 2) Both atria were mildly dilated but bi-ventricular size and function were normal. There were no additional valve lesions observed and no evidence of an aortic root abscess. |
| Day 55 | Discharge from ICU and transferred to the orthopaedic rehab ward. | Discharged from hospital to spinal rehab facility. |
| Day 83 | No further episodes of cardiac rhythm abnormality. | Discharged from hospital to spinal rehab facility. |

**Case presentation**

A 47-year-old male with no cardiovascular history presented to the emergency department with acute onset lower limb weakness. He had a 3-day history of being unwell with pyrexia, rigours, lethargy, supra-pubic discomfort, and bilateral flank pain. Upon arrival, he had a blood pressure of 147/68 mmHg and tachycardic at 125 beats per minute (b.p.m.). His was tachypnoeic at 24 breaths a minute and pyrexial with a temperature of 39.4°C. On examination, he had tenderness over the lumbar spine with reduction of power (2/5) in the proximal lower limbs. There was absence of anal tone on digital rectal examination and he failed a tug test when a catheter was inserted due to urinary retention. His heart sounds were normal with no murmurs appreciated. There was no evidence of splinter haemorrhages, Janeway lesions, or Osler nodes.

Bloods on admission showed raised inflammatory markers, hence a set of blood cultures were obtained. The electrocardiogram (ECG) obtained on admission (Figure 1A) shows sinus tachycardia with a ventricular rate of 125 b.p.m. with a normal cardiac axis, PR interval, QT and QTc intervals, and QRS duration. Given the neurological findings, a magnetic resonance imaging spine (Figure 2) was performed for suspected cord compression. This revealed a collection in both the anterior and posterior epidural space causing compression of the thecal sac. Post-contrast imaging performed confirmed an enhancing epidural abscess extending from the skull base to the 7th thoracic vertebrae. The brain parenchyma was unremarkable with no identifiable abnormality.

An ECG performed prior to surgery (Figure 1B) revealed complete atrioventricular (AV) dissociation (3rd degree AV block). However, the resulting ventricular rate was 75 b.p.m. and the QRS was narrow (<100 ms) in duration. As the ventricular rate was normal, no specific intra-operative precautions were taken at this point. The patient underwent urgent cervicotomochord decompression and washout of cervical vertebrae 6 to thoracic vertebrae 5. Pus obtained during surgery was sent for microscopy, culture, and sensitivity. 

Post-spinal surgery, he was still in complete AV dissociation but now bradycardic (Figure 1C) in the intensive care unit (ICU) and was started on noradrenaline. Due to this change, the cardiology team was contacted to review the patient. Our choice for chronotropic support would have been isoprenaline. However, our colleagues in the intensive care setting were not as familiar with this and felt more comfortable with noradrenaline titration. As the heart rate improved with low-dose noradrenaline and the blood pressure was not compromised, the cardiology and ICU team felt that a temporary pacing wire was not required at that point. The patient remained on a cardiac monitor in ICU with close monitoring from the cardiology team. Given the lack of cardiac history, the initial working diagnosis was autonomic dysreflexia (AD).

An infective haematogenous spread was suspected as *Staphylococcus aureus* was grown from peripheral blood cultures and pus obtained during spinal surgery. There was no obvious bacterial entry point identified on inspection of the patient, however a formal dental check was not performed. Given the evidence of heart block pre- and post-surgery, the source of infection was suspected to be from the heart. A transthoesophageal echocardiogram performed revealed evidence of GD associated with small vegetation measuring 4.0 × 7.0 mm (Figure 3, Videos 1 and 2) Both atria were mildly dilated but bi-ventricular size and function were normal. There were no additional valve lesions observed and no evidence of an aortic root abscess.

The patient was discussed at the infective endocarditis multidisciplinary meeting, the decision was made to treat with IV antibiotics and not close the GD in the acute setting. At the time of discussion, the
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Figure 1 Electrocardiogram. (A) Electrocardiogram on admission. (B) Pre-surgery electrocardiogram—complete atrioventricular dissociation (arrow demonstrating atrial activity within the QRS complex). (C) Post-surgery electrocardiogram—3rd degree atrioventricular block, bradycardia and narrow QRS duration.
The patient was recovering from surgery in ICU with significant paralysis of the lower limbs. Furthermore, the shunt size was small with no ventricular chamber enlargement or reduction in systolic function. A positron emission tomography–computed tomography was not performed as the patient had clear vegetations associated with the GD and no pacing wires inserted. However, it would have been beneficial if the source of infection was unclear. He received IV Vancomycin for 46 days while recovering from spinal surgery.

The vegetation was not seen on transthoracic echocardiogram (Figure 4, Video 3) performed 35 days post-antibiotic initiation. There were no further positive blood cultures and no murmur was heard. Intermittent periods of sinus rhythm with normal AV conduction were noted whilst being monitored in ICU and this was confirmed and sustained during recovery. Electrocardiogram 2 months´ post-surgery demonstrated a sinus tachycardia with no further episodes of complete heart block noted.
The patient was discharged from hospital after 83 days and is currently receiving rehabilitation in the local spinal rehab unit due to persistent lower limb weakness.

Our patient remains severely debilitated and disabled. He has been referred to the regional adult congenital heart disease (ACHD) team for long-term follow-up to monitor the defect and if he develops any symptoms of heart failure. We have deferred the decision to close his defect pending his neurological recovery. If surgery were to be performed, he would almost certainly require permanent pacing as the conduction defect would likely be exacerbated by surgery.

**Discussion**

Gerbode defects can be classified depending on the position of the defect in relation to the tricuspid valve (TV) (*Figure 5*). Type I defects...
are defects located above the TV in the AV component of the membranous septum. Type II defects are located below the TV and occurs via a perimembranous ventricular septal defect. This is almost always associated with a malformed TV. In rare cases, there can be both supravalvular and infravalvular defects are present, this is a type III defect.

A review in 2015 found that acquired GD are more common in males (68%) with an average age of diagnosis is 49 years. Furthermore, it showed that the incidence of Type I defect was the highest at 76%, followed by Type II at 16% and Type III defect at only at 8%. The spectrum of clinical presentations in patients with GD can range from minimal symptoms to severe heart failure. Those presenting with symptoms may complain of chest pain, effort intolerance, or palpitations and may have features consistent with right heart failure or pulmonary artery hypertension, due to shunting of blood from the left ventricle to the right atrium.

Our patient had a Type I GD. It is likely to be a congenital defect due to its small size and the lack of involvegment of other structures. The small size of the defect would also make it unlikely to cause any symptoms but would be a site for infection. There are no specific recommendations on the management of GD found in the 2015 European Society of Cardiology guidelines on infective endocarditis, the 2018 American College of Cardiology/American Heart Association guidelines on the management of ACHD, or the 2020 European Society of Cardiology (ESC) Guidelines for the management of ACHD.

The consensus available in current literature regarding the decision for treatment of GD is mainly based on symptoms. Asymptomatic GD are normally detected incidentally on echocardiogram and can be managed conservatively with regular follow-up. However, some have argued that all GD should be closed surgically as they rarely close spontaneously and there is an increased risk of developing endocarditis. Surgical closure of GD has been the traditional approach with favourable outcomes. When surgery is performed, a suture repair may be sufficient in small GD and patch repair employed in moderate to large defects. A surgical approach is also the preferred method in specific scenarios like infective endocarditis, as a percutaneous device cannot be implanted during active infection.

Given the extensive damage to our patient’s spinal cord, it was not unreasonable for our initial suspicion for the intermittent AV block to be due to AD. Cardiac rhythm abnormalities post spinal cord injury is not completely understood but is thought to be caused by a loss of supraspinal sympathetic control and increased parasympathetic cardiac control. In AD, a peripheral afferent stimulus below the level of the lesion triggers a cascade of events which eventually leads to an increase in parasympathetic activity resulting in brady or tachyarrhythmias. However, despite the many peripheral stimuli he would have received on ICU, there were no further episodes of bradyarrhythmia. There were also no autonomic abnormalities seen in other organ systems.

With the vegetation located so close to the AV node, it is likely that the mechanism of the AV block is due to oedema associated...
with endocarditis. It is very difficult, maybe even impossible, for us to say for certain if the intermittent AV block was caused by AD or endocarditis or perhaps even a combination of the two. However, the cessation of abnormal cardiac rhythm after an extensive period of antibiotics seems to favour endocarditis as the underlying cause. Furthermore, most high-level spinal cord injury patients experience life-long abnormalities in systemic arterial pressure control,18 this was not seen in our patient.

**Conclusion**

In summary, this case highlights a rare congenital defect presenting in an atypical fashion. Gerbode defects are being detected more frequently now given the sophisticated imaging techniques available. The decision for closure in patients with small defects and without signs of cardiovascular compromise remains an area of contention. However, long-term regular monitoring of the defect should be planned to identify any early signs of heart failure.

**Lead author biography**

Dr Yuen Wei Liao is an Internal Medicine Trainee based in Liverpool. He completed 2 years of Academic Foundation Training in Stoke-on-Trent and is pursuing a career in Cardiology.

**Supplementary material**

Supplementary material is available at European Heart Journal - Case Reports online.

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**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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