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Right-Sided Endocarditis involving Eustachian Valve Following the Use of a Central Venous Line

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Patient:	Male, 27-year-old
Final Diagnosis:	Endocarditis
Symptoms:	Fever
Medication:	—
Clinical Procedure:	Transesophageal echocardiogram
Specialty:	Cardiology • Gastroenterology and Hepatology • Nephrology
Objective:	Rare co-existance of disease or pathology
Background:	Right-sided endocarditis is a well-known condition that occurs predominantly in intravenous drug users and patients with cardiovascular implantable electronic devices, central venous lines, and congenital heart disease. Most cases involve the tricuspid valve apparatus. Eustachian valve endocarditis (EVE) is a very rare and under- diagnosed condition with only a few previously reported cases.
Case Report:	We present a rare case of 2-sided infective thromboembolism from <i>Staphylococcus aureus</i> endocarditis involv- ing both the eustachian and mitral valves in a 27-year-old man with mitochondrial neurogastrointestinal en- cephalopathy disease, which is a rare mitochondrial disease.
Conclusions:	Endocarditis involving the eustachian valve is rare and presents a significant dilemma in diagnosis and treat- ment. Late diagnosis can lead to missed thromboembolic events and can have a significant impact on treat- ment and prognosis. In cases with high suspicion, early use of transesophageal echocardiography and chest CT can greatly advance diagnosis. The international guidelines do not specifically address patients with EVE; therefore, we recommend that the endocarditis team should be involved in any case of EVE to customize a treatment strategy.
MeSH Keywords:	Echocardiography • Echocardiography, Transesophageal • Endocarditis, Bacterial • Eustachian Valve • Thromboembolism
Abbreviations:	EV – Eustachian valve; EVE – Eustachian valve endocarditis; IE – infective endocarditis; IVDU – intrave- nous drugs user; RSIE – right-sided infective endocarditis; MNGIE – mitochondrial neurogastrointesti- nal encephalopathy disease; PEG – percutaneous enterogastrostomy; PICC – peripherally inserted central catheter; CLABSI – central line associated blood stream infection; MSSA – <i>Staphylococcus aureus</i> meth- icillin sensitive; CIED – cardiovascular implantable electronic devices; CXR – chest x-ray; IVC – inferior vena cava; CVC – central venous catheter; TTE – transthoracic echocardiography; TEE – transesophageal echocardiography
Full-text PDF:	https://www.amjcaserep.com/abstract/index/idArt/923465



Background

Right-sided infective endocarditis (RSIE) is a well-known condition that occurs predominantly in intravenous drug users and patients with pacemakers, central venous lines, or congenital heart disease [1,2].

RSIE accounts for 5% to 10% of all endocarditis cases, most of which involve the valvular apparatus. The prognosis of RSIE is usually favorable, and few cases require surgical intervention. However, RSIE is associated with a high rate of complications [3].

According to a study involving the European Endocarditis Registry, embolic events from RSIE occurred in 21% of cases and were significantly associated with tricuspid or pulmonary endocarditis, the presence of vegetation, and *Staphylococcus aureus* [4].

In-hospital death occurred in 17.1% of cases and had a higher frequency in patients with prosthetic valve endocarditis. Independent predictors of mortality are the Charlson comorbidity index, creatinine >2 mg/dL, congestive heart failure, vegetation length >10 mm, cerebral complications, abscess, and failure to undergo surgery when indicated [4].

The eustachian valve is an embryological remnant of the sinus venosus and is located between the inferior vena cava and the right atrium. In adults, it serves no significant function and is considered a benign rudimentary structure [5–8]. Vegetation involving the eustachian valve is rare and only sporadic cases have been reported. Here, we describe an extremely rare case of embolic RSIE involving both the eustachian and mitral valves, without an intra-atrial shunt [9].

Case Report

A 27-year-old man presented with a history of mitochondrial neurogastrointestinal encephalopathy disease, a rare genetic disorder that causes an impairment of mitochondrial function with involvement of the gastrointestinal and central nervous systems. While neurological involvement occurs in the form of peripheral neuropathy and leukoencephalopathy, gastrointestinal involvement mainly involves dysmotility and malabsorption, which lead to severe malnutrition. Our patient needed nutritional support and thus had a percutaneous endoscopic gastrostomy and was treated with total parenteral nutrition via a central catheter line peripherally inserted during the night. He had jejunal diverticulosis with multiple episodes of pseudo-obstructions that required partial small bowel resection. He also had insulin-dependent type I diabetes mellitus.

He presented to the Emek Medical Center Emergency Department with fever, vomiting, and abdominal pain. On clinical examination, the patient looked ill and had a core temperature of 38.5°C, heart rate of 120 beat/min, and blood pressure of 120/70 mmHg. On cardiac auscultation, heart sounds were rapid but regular, with no evidence of additional heart sounds or murmurs. No abnormalities were observed on lung auscultation. Mild abdominal tenderness without guarding was noticed. The percutaneous endoscopic gastrostomy was functioning without signs of infection. Marked redness was noticed around the central catheter line, with pussy secretion. No stigmata of endocarditis was observed.

Laboratory tests showed a marked increase in inflammatory markers (WBC 20 K/uL, range 4.5–11.5, indicating leukocytosis; and C-reactive protein 25 mg/dL, range 0.0–0.5); acute kidney injury (creatinine 1.3 mg/dL, range 0.67-1.17; urea 50 mg/dL, range 17–43) with evidence of diabetic ketoacidosis (serum glucose level 420 mg/dL; positive ketones in peripheral blood); and metabolic acidosis (pH 7.2, range 7.35–7.45; pCO₂ 32 mmHg, range 25–35; and HCO³ 11 mmHg, range 21–28). The initial ECG demonstrated a regular sinus rhythm with an RR interval of 120 and a normal axis without PR or QT segment prolongation. No changes were seen in ST segments or T waves. The chest X-ray was normal for the patient's heart size, and lung fields and abdominal X-ray showed gastric distention without signs of obstruction or perforation.

The working diagnosis was central line-associated bloodstream infection. The central line was removed and sent for culture, as were 3 sets of peripheral blood cultures. The blood cultures were positive for methicillin-sensitive *S. Aureus* (MSSA), and an anti-staphylococcal antibiotic was initiated. The patient developed hypoxemic respiratory failure and was intubated and moved to the intensive care unit.

A chest X-ray showed marked bilateral consolidations with hyperdensities in the apex and base of the right lung (Figure 1). Transthoracic echocardiography (TTE) was performed with a poor acoustic window, showing preserved function, no obvious vegetations, and mild mitral and tricuspid regurgitations. Transesophageal echocardiography (TEE) showed a small mobile mass on the eustachian valve and a large 2×1.5 cm vegetation at the anterior leaflet of the mitral valve without echocardiographic evidence of an intra-atrial shunt (Figures 2, 3).

Over the next days, the patient deteriorated rapidly, and the clinical evidence indicated he had ongoing refractory septic shock. The heart team decided not to perform a surgical intervention at this point and suggested a more conservative approach with a trial of an extended antibiotics regimen.

Positive MSSA cultures continued to grow despite adequate therapy, and further hemodynamic and respiratory deterioration required vasopressor support. A whole-body CT revealed



Figure 1. Chest X-ray. Marked bilateral consolidations with hyperdensities in the apex and base of the right lung.



Figure 2. Transesophageal echocardiography (TEE). A small mobile mass is seen on the eustachian valve (full arrow).



Figure 3. Transesophageal echocardiography (TEE). A large 2×1.5 cm vegetation is seen at the anterior leaflet of the mitral valve (dotted arrow).

massive bilateral consolidations with cavitation on the right upper quadrant with septation, consistent with septic emboli (Figure 4). A brain CT revealed a large frontoparietal infarct consistent with middle cerebral artery occlusion (Figure 5).

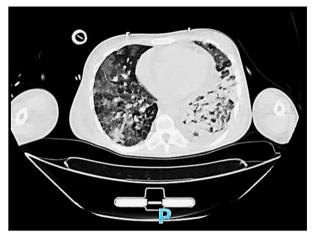


Figure 4. Chest CT. Massive bilateral consolidations with cavitation are seen on RUQ with septation.

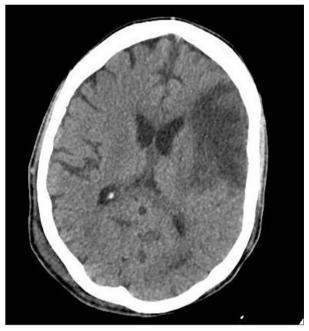


Figure 5. Brain CT, showing a large frontoparietal infarct.

Unfortunately, the patient's condition did not improve and he later developed ventilation-associated pneumonia and disseminated intravascular coagulopathy and died.

Discussion

RSIE is a rare manifestation of infectious endocarditis seen mostly in patients who are at high risk for bloodstream infections, such as intravenous drugs users and patients who have cardiovascular implantable electronic devices or congenital structural heart defects. RSIE mostly involves the tricuspid and the pulmonary valves, and, in rare cases, the sub-valvular apparatus [1–3].

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The eustachian valve is usually a small crescentic fold remnant of the endocardium arising from the anterior rim of the inferior vena cava orifice [5–8]. The eustachian valve is best seen with TEE in the bi-caval view and right-sided horizontal and longitudinal views [11–13].

We present a unique case of RSIE involving a persistent eustachian valve due to MSSA infection, which originated from an infected central venous line. While the proximity of the central line to the eustachian valve probably explains the valve's involvement, the vegetation seen on the mitral valve, without evidence of an intra-atrial shunt, may be explained by the high MSSA infectious burden and virulence.

The fact that the eustachian valve is poorly visualized on TTE makes it very difficult to reveal a small vegetation attached to the right atrium or inferior vena cava wall. Nevertheless, current guidelines do not support the use of TEE for initial examination in the diagnosis of native right-valve endocarditis, which may lead to a missed diagnosis [4]. In our case, TTE failed to demonstrate RSIE, and only TEE clearly demonstrated the vegetation on the eustachian valve.

The antibiotics regimen recommended for double-sided infective endocarditis is the same as that for left-sided infective endocarditis. The duration of antibiotic therapy for uncomplicated RSIE is usually 2 weeks, but the guidelines do not address the treatment duration or the surgical indications in the case of endocarditis involving the eustachian valve [4]. After reviewing the literature, 2 main questions remained unanswered: first, whether EVE has a higher thromboembolic potential than

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uncomplicated RSIE has, and second, whether EVE should be addressed with a more aggressive approach.

In our opinion, effort should be made to fully visualize the eustachian valve and nearby structures in cases of persistent refractory *S. Aureus* bacteremia and in cases with a high suspicion of RSIE, including for intravenous drug users and patients with a central venous catheter line, implantable electronic device, repeated pulmonary embolic phenomena, or congenital heart defect. In addition, TEE is a reasonable next step when TTE has failed. Patients with EVE should be closely monitored for embolic phenomena, and the intra-atrial septum should be visualized to assess an intra-atrial shunt. In cases of clinical suspicion, a chest CT or ventilation-perfusion lung scan can help in the diagnosis.

Conclusions

EVE is rare phenomenon but can have an important clinical impact on diagnosis, treatment, and prognosis. The true incidence of EVE is probably underestimated, and late diagnosis can lead to an unfavorable outcome, which will limit treatment options. Choosing the best modality to visualize EVE is critical because the clinical scenario may not be clear. Guidelines should address the unique features of EVE to increase awareness and guide physicians in customizing medical therapy.

Conflict of interests

None.

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