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INTERMEDIATE

CASE REPORT: CLINICAL CASE

Intraparenchymal Brain Abscess in an Adult Male With Underlying Ebstein Anomaly and Cor Triatriatum Dexter

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ABSTRACT

A 21-year-old man presented with new-onset seizures and brain abscess. Echocardiography and cardiac magnetic resonance imaging revealed underlying Ebstein anomaly, secundum atrial septal defect, and cor triatriatum dexter. The elevated right heart pressures shunting through the septal defect and transient bacteremia were the likely mechanisms for his presentation. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:194–7)

A 21-year-old South American man presented with acute right-sided weakness and paresthesias. He became progressively encephalopathic and experienced a generalized tonic-clonic seizure in the emergency department. He was afebrile, with blood pressure of 114/55 mmHg, pulse of 75 beats/min, respirations of 15 breaths/min, and pulse oximetry of 96% on room air. Examination was significant for aphasia without any other neurological deficits, and a 3/6 holosystolic murmur at the left sternal border with a split S1. No cyanosis or digital clubbing was appreciated.

LEARNING OBJECTIVES

- Echocardiography is an essential tool to rapidly assess cardiac structures and guide management.
- Ebstein anomaly has variable clinical presentations. It may remain undiagnosed and present later in life via secondary manifestations.
- Paradoxical embolism is common in Ebstein anomaly, although it is still a rare cause of brain abscess.

DIFFERENTIAL DIAGNOSIS

The etiology for brain abscess was broad because of a lack of any other medical history, and included bacterial, mycobacterial, fungal, or parasitic organisms. Brain abscess formation is associated with predisposing conditions, such as head trauma, severe immunocompromise due to HIV or immunosuppressive medications, systemic infection in the setting of underlying endocarditis, congenital heart defects, or pulmonary arteriovenous fistulas (1). Echocardiography quickly narrowed the differential by identifying the patient's underlying cardiac abnormalities.

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INVESTIGATIONS

Laboratory analysis did not reveal abnormalities. Electrocardiography (ECG) showed normal sinus rhythm with atrial enlargement and incomplete right bundle branch block. Echocardiography was immediately performed because of a significant systolic murmur on examination that demonstrated a markedly dilated right ventricle (RV) and an apically displaced tricuspid valve. Computed tomography and subsequent brain magnetic resonance imaging (MRI) showed a 2.6- × 2.5- × 2.3-cm abscess in the left frontal lobe with surrounding edema and mid-line shift (**Figures 1A and 1B**). Formal cardiac workup, which consisted of transthoracic and transesophageal echocardiograms, as well as cardiac MRI, showed a severely dilated right atrium, RV, and tricuspid annulus, with an elongated anterior tricuspid leaflet and apically displaced septal and posterior leaflets, which is classic for Ebstein anomaly (**Figure 1C and 1D**). A secundum atrial septal defect (ASD) (**Figure 1E**) and mid-right atrial membrane consistent with cor triatriatum dexter were also appreciated (**Figure 1F**).

MANAGEMENT

Upon discovery of the brain abscess with mid-line shift, the patient was emergently taken for left frontoparietal craniotomy for evacuation by neurosurgery. Cultures grew pan-sensitive *Streptococcus intermedius*. He improved post-operatively with no residual neurological deficits and was discharged on a 6-week course of ceftriaxone and metronidazole with cardiology follow-up.

DISCUSSION

Brain abscess is a challenging diagnosis due to its broad differential in which cardiac etiologies are believed to account for 9% to 43% of cases, with congenital causes consisting of 2% of these (2,3).

Ebstein anomaly accounts for <1% of all congenital cardiac defects, and a secundum ASD or patent foramen ovale is the most common associated defect, occurring in >80% of these patients (4). Cor triatriatum dexter has also been reported in association with other right-sided anomalies, with the degree of septation potentially increasing intracardiac pressures and exacerbating heart failure symptoms (5,6).

In a retrospective study by Attenhoffer et al. (7), paradoxical embolization was seen in adults with Ebstein anomaly and severe tricuspid regurgitation and was strongly associated with the presence of an ASD. Clinical presentation of Ebstein anomaly is otherwise variable and is related to the degree of tricuspid valve leaflet displacement and regurgitation. The Carpentier classification provides a means of describing the degree of malformation severity (8). Our patient's echocardiographic findings were most consistent with type B, a large atrialized right ventricle but with a freely moving anterior leaflet of the tricuspid valve.

As outlined by the American College of Cardiology/American Heart Association guidelines (9), management strategies for Ebstein anomaly, ASD, and cor triatriatum dexter are guided by patient age and presence of clinical heart failure, arrhythmia, or cyanosis. Asymptomatic individuals are managed conservatively, unless they undergo other cardiac surgery, at which point surgical repair is indicated. Yearly monitoring with ECG, echocardiography, and pulse oximetry is recommended, to assess for progressive RV enlargement, atrial tachyarrhythmias, paradoxical embolism, or systemic desaturation from progressive right-to-left shunting. If any of the preceding are present, then surgical repair should be pursued.

Medical management focuses on left ventricular systolic function optimization and rhythm control before surgery (9). Long-term anticoagulation is recommended in patients with evidence of embolization, and endocarditis prophylaxis is advised in those with prosthetic heart valves, a history of endocarditis, or those who are cyanotic.

Early intervention is recommended because delaying surgery until onset of RV dysfunction or heart failure symptoms is associated with worse outcomes. Tricuspid valve repair, plication of the atrialized right ventricle, reduction atrioplasty, and closure of the ASD are classically performed. Treatment for cor triatriatum dexter involves percutaneous balloon dilatation if suitable anatomy is present or surgical removal and repair. Alphonso et al. (10) conducted a retrospective review of 28 patients over a 22-year span with typical and atypical cor triatriatum dexter who underwent surgical repair. At a median

ABBREVIATIONS AND ACRONYMS

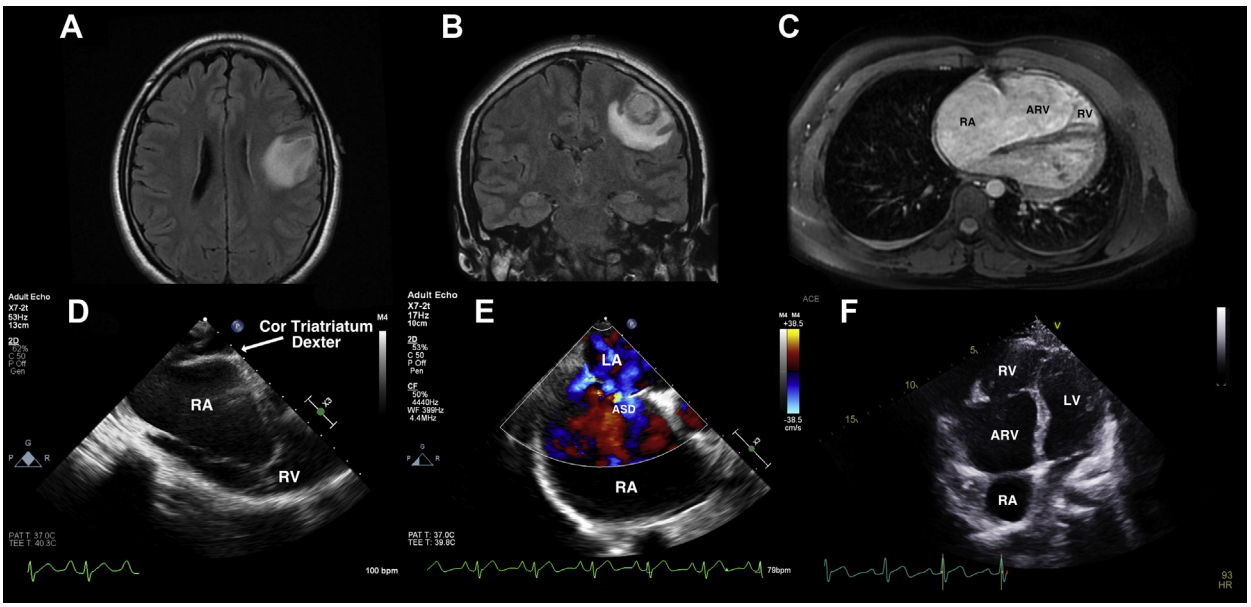
ASD = atrial septal defect

ECG = electrocardiography

MRI = magnetic resonance imaging

RV = right ventricle

FIGURE 1 Imaging and Pathology of Ebstein Anomaly



(A and B) Brain magnetic resonance imaging (MRI) axial and coronal images demonstrating anterior left parietal lobe abscess with surrounding vasogenic edema and mid-line shift. **(C)** Cardiac MRI showing displaced tricuspid valve, atrialized right ventricle (ARV), and reduced right ventricle (RV). **(D and E)** Transesophageal images showing apically displaced tricuspid valve with severe atrialization of the RV, cor triatriatum dexter, and atrial septal defect (ASD) with associated right-to-left shunt. **(F)** Transthoracic 4-chamber view showing true RV, ARV, and true right atrium (RA). The RA and ARV are further separated by the cor triatriatum dexter. LA = left atrium; LV = left ventricle.

follow-up of 98 months, survival was 96% and 88% at 5 and 15 years, respectively, with all patients in New York Heart Association function class I (10).

Our patient had no reported history of previous procedures that might have resulted in bacteremia. Therefore, although extensive thrombotic workup was not pursued, we believe he likely became bacteremic in the setting of an oral infection based on the presence of *Streptococcus intermedius*, with development of infective endocarditis, subsequent embolization, and seeding of the brain parenchyma with eventual abscess formation. Although echocardiography did not show a vegetation, it did not rule out the presence of a previous embolized vegetation. Such patients should undergo a thorough dental examination and pursue corrective care as soon as possible. Because of paradoxical embolization, the patient was recommended evaluation for surgical repair upon completion of his antibiotic course.

Physiologically, increased right atrial pressures stemmed from worsening tricuspid regurgitation that was further compounded by the degree of atrial septation from cor triatriatum dexter. This further reduced RV inflow and worsened right atrial pressures, ultimately increasing the magnitude of right-to-left shunting through the ASD.

FOLLOW-UP

The patient completed his antibiotic treatment and returned to baseline health. Antibiotic coverage was advised for endocarditis prophylaxis for future dental procedures. He was ultimately lost to follow-up, as is the case in a substantial portion of this population. However, patients with congenital heart disease have better outcomes when cared for via a multidisciplinary team with a congenital heart disease specialist as they transition into adulthood (9).

CONCLUSIONS

We presented a rare case of a previously asymptomatic young man who was found to have

numerous underlying cardiac abnormalities, including Ebstein anomaly, ASD, and cor triatriatum dexter, in the setting of a new brain abscess. Our case highlights the value and usefulness of echocardiography because it provided an immediate diagnosis that changed early antimicrobial management. It was primarily responsible for raising the suspicion for a bacterial infection when a parasitic infection initially seemed more likely because of his endemic area of origin.

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The authors have no relationships relevant to the contents of this paper to disclose.

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