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Case Report

Paradoxical septic embolism in an Ebstein's anomaly patient leading to brain abscess: A case report

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ABSTRACT

Ebstein's anomaly (EA), a congenital cardiac anomaly, is characterized by apical displacement of the tricuspid valve leaflet(s) into the right ventricle. We present the case of a 61-year-old female with a history of EA, Wolff-Parkinson-White syndrome, and patent foramen ovale (PFO), who presented with worsening hypoxia and confusion, in the setting of left lower extremity cellulitis and abscess. The computed tomography (CT) scan of the head showed a cerebellar infarct with hemorrhagic conversion. Magnetic resonance imaging of the head showed a satellite lesion raising concern for the embolic nature of infarcts. After ruling out cardioembolic causes of cerebellar infarction, her presenting symptoms were attributed to paradoxical septic emboli from the left leg abscess (demonstrated on CT scan of the leg). She was deemed a poor candidate for surgical closure of PFO due to contraindication to use heparin (due to the presence of hemorrhagic stroke) and underlying comorbidities. Septic embolization is a rare but dreaded complication in EA patients with PFO.

Learning objective:

- Paradoxical emboli can occur in patients with Ebstein's anomaly (EA) and patent foramen ovale (PFO).
- The mainstay of management in case of paradoxical embolism lies with the identification and treatment of the underlying cause, such as infective endocarditis, deep vein thrombosis, or infectious source, as in the present case.
- The surgical correction of PFO in EA patients should be considered when the patient becomes symptomatic with cyanosis, hypoxia, or manifestations of paradoxical emboli.

Introduction

Ebstein's anomaly (EA) is defined as the apical displacement of one or more leaflets of the tricuspid valve (>8 mm/m^2) into the right ventricular cavity, such that the proximal part of the right ventricle (RV) becomes atrialized and the size of the RV (the distal component) significantly decreases. EA is a rare entity since it accounts for <1 % (around 0.5 %) of all congenital cardiac disorders and only around 5 % reportedly survive beyond the age of 50 years [1,2]. Sometimes, this anomaly can go undiagnosed until adulthood when the patient remains asymptomatic because of a well-functioning right heart [3].

The clinical spectrum varies with age. In adults, the presentation ranges from mild symptoms of dyspnea to life-threatening right heart failure and fatal arrhythmias, depending on the complexity and severity of underlying anatomic changes. Here, we report the case of a 61-year-old female, with EA and bicuspid aortic valve, who presented with paradoxical septic emboli via right-to-left shunt (RLS) of patent foramen ovale (PFO) in the setting of left lower extremity abscess.

Case report

A 61-year-old female presented with complaints of fevers, chills, dyspnea, increasing confusion, and lethargy for 5 days. Her medical history included hypertension, hyperlipidemia, diabetes, obesity (body mass index — 31 kg/m^2), bicuspid aortic valve, chronic hypoxemic respiratory failure from RLS via PFO requiring 4–5 L of oxygen therapy, paroxysmal supraventricular tachycardia (SVT) in the form of atrioventricular re-entrant tachycardia (AVRT)/Wolff-Parkinson-White (WPW) syndrome, and paroxysmal atrial fibrillation (PAF).

Her vitals on arrival revealed a temperature of 36.1 °C, blood pressure of 140/92 mmHg, heart rate in the 90s, and respiratory rate of 22/min with an oxygen saturation of 86 % on room air. Physical examination showed non-focal neurological examination with confusion, clear lungs to auscultation, no jugular venous distension, and a grade 3–4 holosystolic murmur was heard on auscultation, loudest in the tricuspid area. The left lower extremity (LLE) was non-erythematous,

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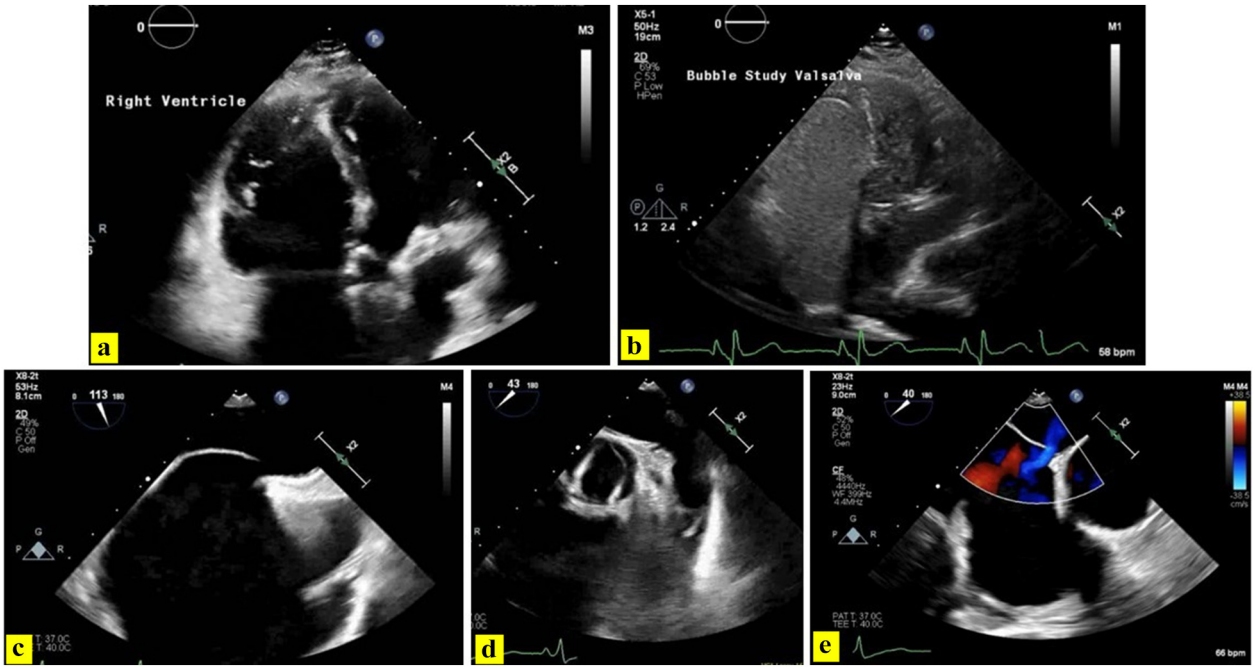

Fig. 1. (a) Transthoracic echocardiography showing right ventricle with apically displaced tricuspid valve leaflets. (b) Transthoracic echocardiography showing agitated saline (bubbles) in both right ventricle and left ventricle suggesting the presence of patent foramen ovale or atrial septal defect. (c) Transesophageal echocardiography showing interatrial septum with patent foramen ovale. (d) Transesophageal echocardiography showing bicuspid aortic valve. (e) Transesophageal echocardiography showing patent foramen ovale with bidirectional flow.

but tender to touch, with no peripheral edema. Admission laboratory results were as follows: White blood cell count 30.4 K/μL (92 % neutrophils), hemoglobin 15.6 g/dL, platelet count 251 K/μL, sodium 133 mEq/L, potassium 3.9 mmol/L, chloride 93 mEq/L, bicarbonate 21 mEq/L, creatinine 2.07 mg/dL (baseline creatinine 0.7 mg/dL), glucose 324 mg/dL, lactic acid 5.5 mmol/L, B-type natriuretic peptide 128 pg/mL, troponin-I level 1.04 ng/mL, and thyroid stimulating hormone 1.41 IU/mL.

The chest X-ray on admission showed cardiomegaly, but no pleural effusion or pneumothorax. Computed tomography (CT) scan of her chest was negative for pulmonary edema, infiltrates, and acute lung parenchymal process. A transthoracic echocardiogram (TTE) revealed a small left ventricle with an ejection fraction of 50 %, enlarged right ventricle with apically displaced tricuspid valve (10.7 mm/m^2 from the tricuspid annulus), severe tricuspid regurgitation (TR), muscular ventricular septal defect (VSD), and stretched PFO with bidirectional shunting/flow (Fig. 1a). The shunting via PFO was confirmed on TTE with a saline bubble study (Fig. 1b). She was admitted to the medical intensive care unit for acute hypoxic respiratory failure and sepsis with the left extremity as a source of infection. She was placed on 15 L of oxygen via high-flow nasal cannula (HFNC) and started on vancomycin and piperacillin-tazobactam. She was also started on furosemide with improvement in her oxygen requirement from 15 L via HFNC to 5 L via nasal cannula. She continued to have intermittent confusion despite improvement in oxygen requirements, leukocytosis, and lactic acidosis, prompting a CT scan of her head which showed a left cerebellar localized lesion with mass effect and edema concerning for hemorrhagic conversion and a tiny chronic left thalamic lacunar type infarct (Fig. 2a). Same-day magnetic resonance imaging of the head confirmed CT head findings and showed a small satellite infarct in bilateral cortical area suggesting the embolic nature of infarcts (Fig. 2b). Transesophageal

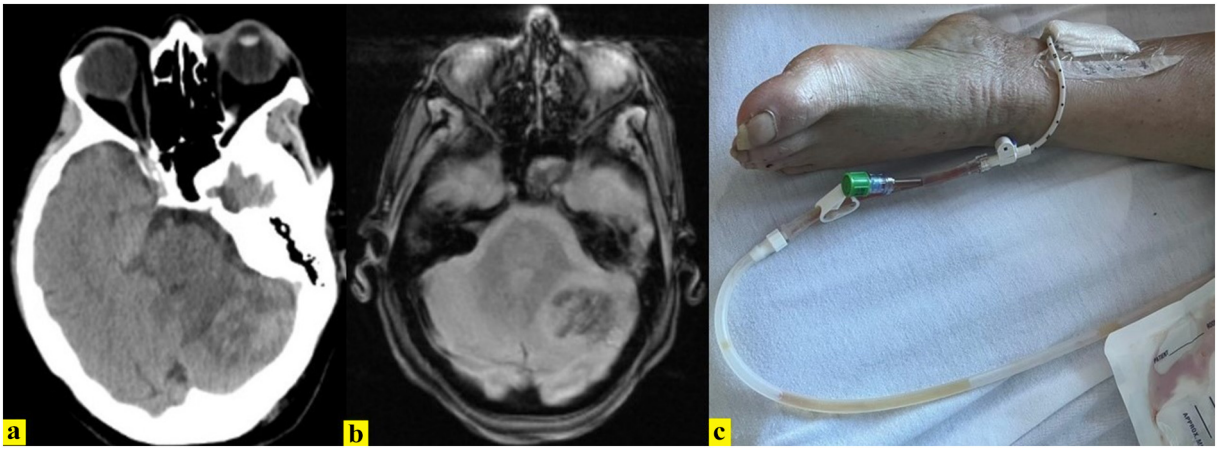

Fig. 2. (a) Computed tomography of the head showing left cerebellar lesion concerning for septic emboli. (b) Magnetic resonance imaging of the head showing large left cerebellar parenchymal lesion, a complication of embolization. (c) Pigtail catheter placed in the left lower extremity to drain the abscess.

echocardiogram confirmed TTE findings and was negative for valvular vegetation and left atrial appendage thrombus (Fig. 1c, d, e) The venous Doppler of both lower extremities was negative for deep vein thrombosis (DVT). CT angiogram of the neck showed minimal calcified plaque but no significant carotid/vertebral artery stenosis. A CT scan of LLE showed fluid collections involving the deep and superficial posterior compartments of the left calf concerning an abscess. No other infectious source was identified on imaging of the chest, abdomen, pelvis, and spine. She underwent fluid drainage with a drain placement for the LLE abscess (Fig. 2c). Her serial blood cultures remained negative.

The baseline electrocardiogram (ECG) showed normal sinus rhythm with age-indeterminate septal and inferior infarct along with accessory pathway (Fig. 3a). She developed multiple episodes of hemodynamically significant SVT. ECG showed orthodromic AVRT with underlying right bundle branch block (Fig. 3b) requiring adenosine and electrical cardioversion to sinus rhythm. Due to recurrent episodes of SVT, she underwent electrophysiology study. She was found to have a posterior/posterolateral accessory pathway with orthodromic AVRT and underwent successful ablation for the same. She underwent right heart catheterization, which showed normal right and left-sided filling pressures (right atrial pressure of 3 mmHg, pulmonary artery pressure of 27/10 mmHg, and pulmonary capillary wedge pressure of 9 mmHg) with cardiac output 3.1 L/min and cardiac index 2.0 L/min/m^2.

Later, fluid culture from her left leg grew *Parvimonas micra*, *Streptococcus pyogenes*, and *Streptococcus anginosus*. She was treated with ceftriaxone and metronidazole for a total of 14 days and discharged to a skilled nursing facility on oxygen therapy of 4–5 L. She was seen by the adult congenital cardiology team and her case was discussed in a regional congenital heart disease board meeting. She was deemed a poor surgical candidate for tricuspid valve replacement ± closure of PFO, the likely cause of paradoxical embolic events. This was due to the presence of multiple comorbidities and the requirement for intra-operative

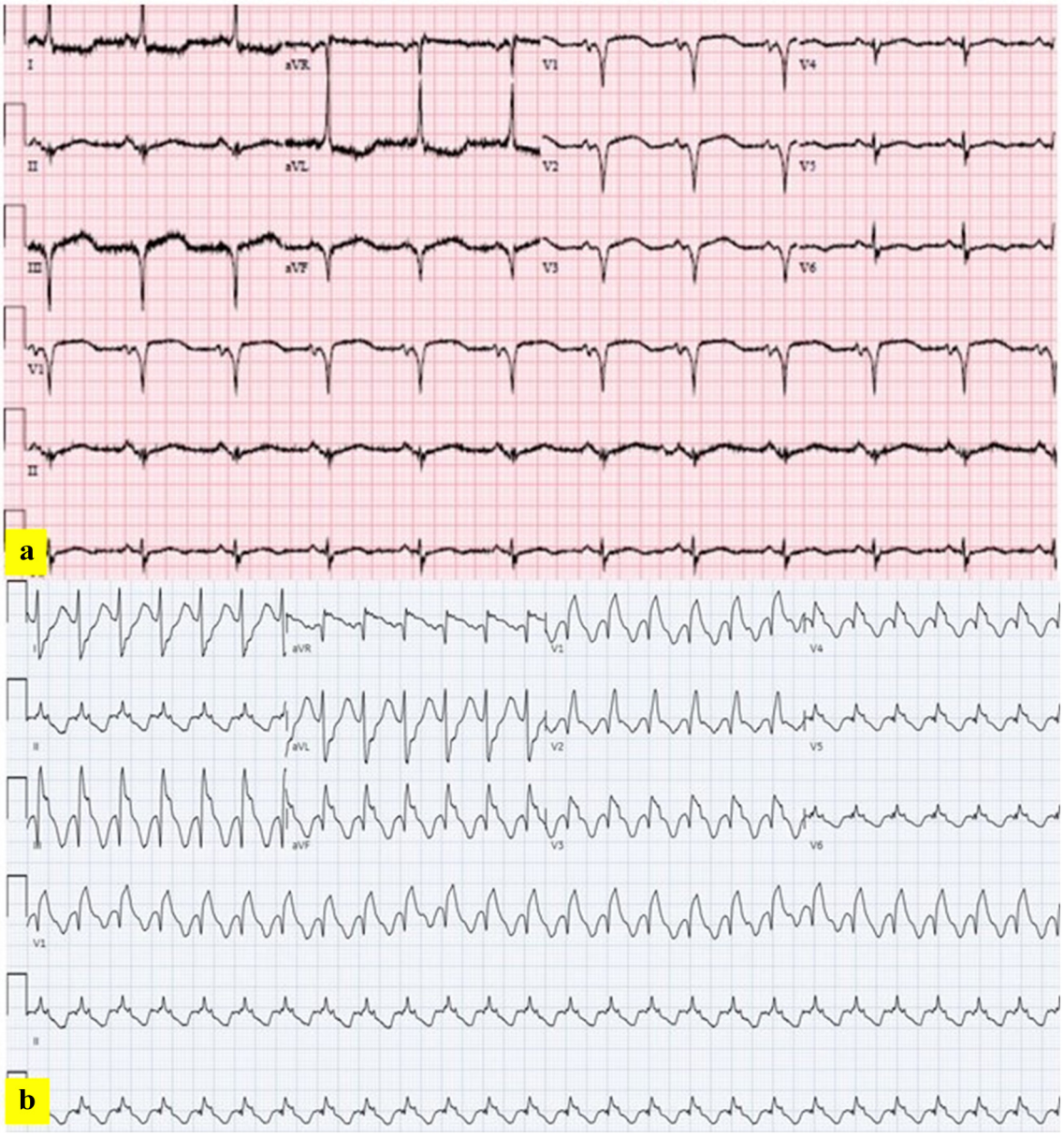

Fig. 3. (a) Electrocardiogram (ECG) shows normal sinus rhythm with age-indeterminate anteroseptal and inferior infarct, and Wolff-Parkinson-White syndrome. (b) ECG showing orthodromic atrioventricular re-entry tachycardia with underlying right bundle branch block.

heparin, which was contraindicated in view of the recent hemorrhagic conversion of her cerebellar infarct. Air filters were placed in all IV lines given the presence of shunt physiology.

Discussion

EA has been commonly reported in association with atrial septal defect (ASD) or PFO, right ventricular outflow tract obstruction, and WPW syndrome, as demonstrated in the present case [2,4]. To the best of our knowledge, this is the 5th case report of EA with PFO/ASD-associated septic paradoxical embolism, but the first case report in the setting of septic emboli originating from a lower extremity abscess [3,5–7].

The principal echocardiographic findings in a patient with EA include right ventricular volume overload, interventricular septal motion abnormality, and apical displacement of one or more tricuspid valve leaflet(s) causing TR, as seen on Doppler examination. Paradoxical embolism is a potential complication in the presence of PFO/ASD. Paradoxical emboli rarely lead to arterial ischemia, but this is true in the present case, where septic paradoxical embolism from the LLE abscess caused multiple ischemic events in the brain [8]. The chronic lacunar infarct noted in CT of the head in our case was most likely due to microvascular ischemic injury in the setting of multiple risk factors (hypertension, hyperlipidemia, and diabetes). Brain abscess is a rare complication in patients with EA. According to a report from the Mayo Clinic, only 5 patients had a history of brain abscess among 968 EA patients [9].

In the present case, hypoxia was due to the mixing of arterial and venous blood in the presence of a bidirectional shunt via PFO. The embolus was likely not of cardio-embolic origin, given the lack of valvular vegetations and left intra-atrial thrombus on echocardiography. The negative lower extremity ultrasound ruled out DVT. The left leg abscess detected on CT imaging was the most likely infectious focus that led to paradoxical septic embolization via RLS, leading to these manifestations in the patient. The treatment of paradoxical septic emboli relies on treating the underlying infectious cause.

A possible explanation for bidirectional shunting seen on echocardiography could be a transient increase in right atrial pressure due to TR from EA leading to transient RLS, which ultimately led to the emboli generated in the systemic veins entering the systemic circulation via intracardiac shunts and bypassing filtering action of the lungs [4]. The closure of PFO in EA patients can be considered in those who are severely symptomatic due to cyanosis, hypoxia, or manifestations of paradoxical emboli. In our case, the decision was made not to close the PFO due to the above-mentioned reasons. When considering surgical PFO closure in EA, the indications for concomitant replacement/repair of the tricuspid valve should be considered. In patients who are not candidates for surgical repair, the percutaneous options for PFO closure can be considered [10].

Conclusion

The presence of septic emboli is an uncommon presentation in patients with EA and PFO, and can be challenging to diagnose. A detailed history and physical examination along with focused diagnostic tests are required to diagnose the source of emboli.

Consent statement

Written informed consent was obtained from the patient for publication of this case report, including accompanying images.

Declaration of competing interest

The authors declare that there is no conflict of interest.

Acknowledgments

None.

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