

Uncorrected Ebstein's anomaly with atrial septal defect complicated by brain abscess in an adult patient

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We report the case of a 56-year-old male patient affected by a severe form of Ebstein's anomaly (type C of Carpentier classification) with secundum atrial septal defect, who presented to the emergency department with impaired consciousness, seizures, and trismus. The brain computed tomography scan showed evidence of a mass located in the frontal lobe, confirmed by brain MRI consistent with brain abscess. Both echocardiography and cardiac MRI showed no evidence of valvular vegetation. This case shows how the combination of increased atrial pressure and bidirectional shunt through atrial septal defect may lead to paradoxical embolization.

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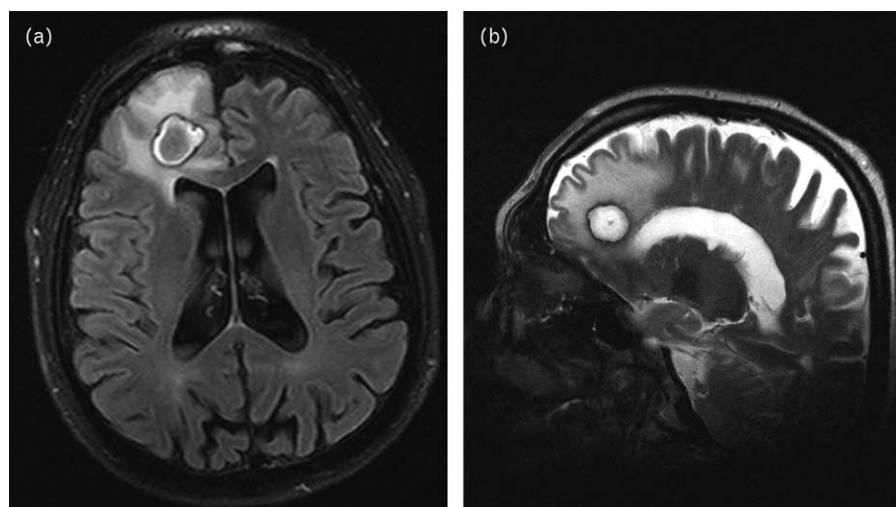
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A 56-year-old patient presented to the emergency department unresponsive with eye deviation, seizures, and trismus. Physical examination showed lip cyanosis and acrocyanosis, mild finger clubbing, normal heart rate, and a holosystolic murmur. The patient was afebrile. Laboratory analysis revealed increased hematocrits and mild leukocytosis. ECG showed right atrial dilatation and complete right bundle branch block. Recent medical history included an episode of gingivitis associated with

fever and sore throat 2 weeks before; after that, the patient underwent professional teeth cleaning.

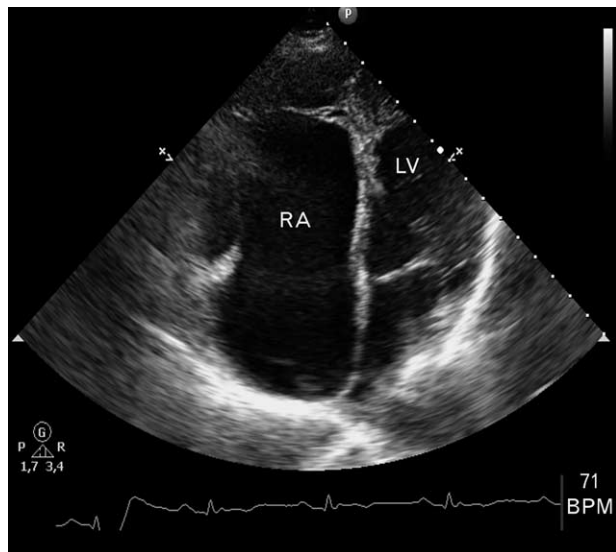
He had been diagnosed with severe Ebstein's anomaly (type C of Carpentier classification¹) and ostium secundum atrial septal defect (ASD) in young age, but he always refused any surgical treatment. In the emergency department, the patient underwent brain computed tomography (CT) scan with evidence of a nodular lesion

Fig. 1



Brain MRI showing brain abscess located in the right frontal lobe with perilesional enhancement consistent with edema and tissutal necrosis. (a) axial view; (b) sagittal view.

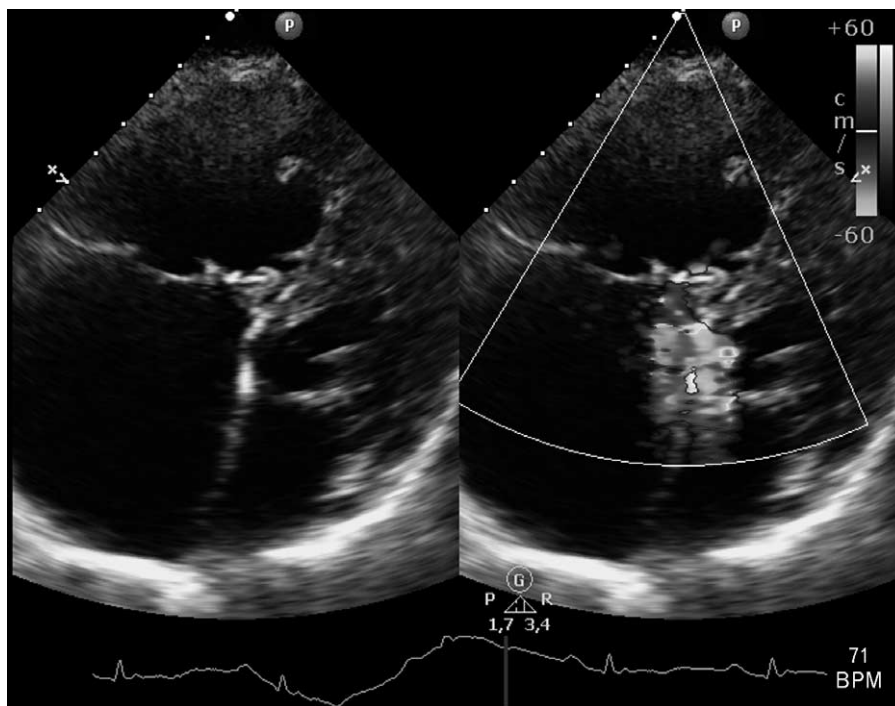
Fig. 2



Four-chamber view of transthoracic echocardiography showing the severe apical displacement of the septal leaflet of the tricuspid valve and the increased dimension of the right atrium and atrialized ventricle.

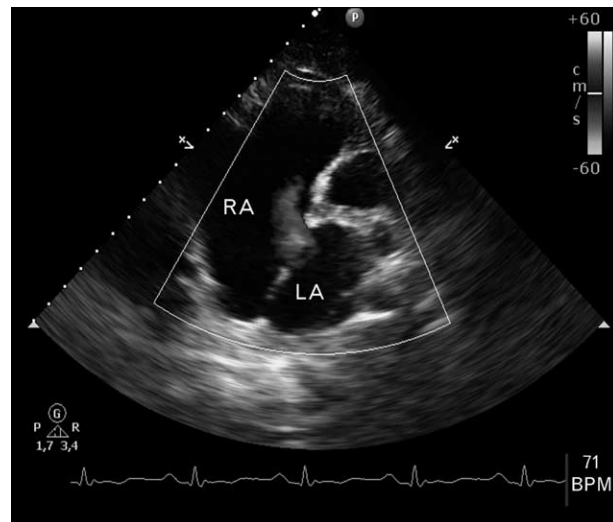
in the right frontal lobe. Further investigation with brain MRI was performed showing a ring-enhancing lesion of 2.5 cm of diameter consistent with brain abscess (Fig. 1).

Fig. 3



Apical four-chamber view of transthoracic echocardiography with evidence of severe tricuspid regurgitation.

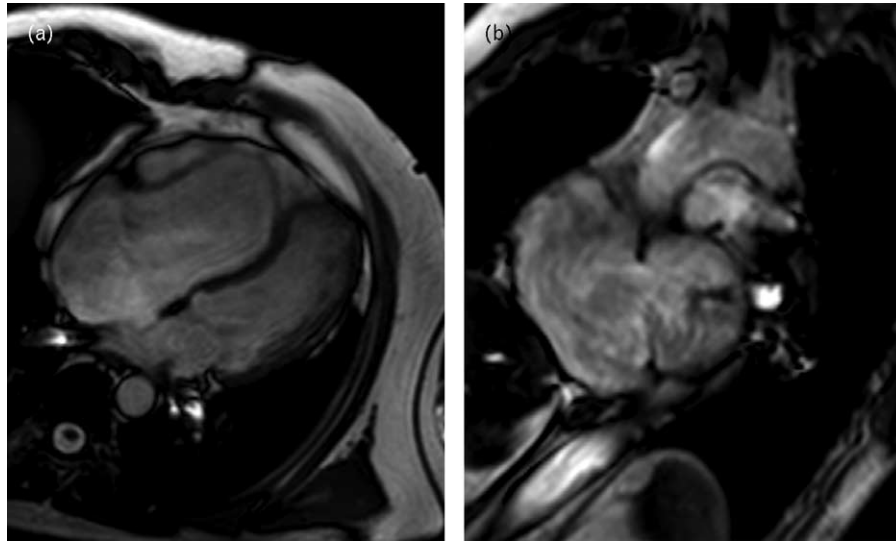
Fig. 4



Color Doppler transthoracic echocardiography showing ostium secundum atrial septal defect with left-to-right shunt.

Transthoracic echocardiography showed a dilated right atrium and an atrialized ventricle with severe displacement of the septal leaflet of the tricuspid valve (Fig. 2). Doppler analysis revealed severe tricuspid regurgitation

Fig. 5



Cardiac MRI. Four-chamber view showing the atrialized part of the right ventricle and the apical displacement of the septal tricuspid valve leaflet (a); atrial short-axis cine with evidence of atrial septal defect (ASD) with bidirectional shunt (b).

and a bidirectional shunt through the ASD (Figs. 3 and 4). Cardiac MRI was performed in order to evaluate better cardiac anatomy (Fig. 5). Both echocardiography and MRI excluded the presence of valvular vegetations.

The patient was treated with ceftriaxone and metronidazole. Blood cultures were persistently negative. The follow-up MRI performed after 1 month of antibiotic therapy showed partial regression of the brain lesion. Serial brain MRI documented a complete regression of the brain lesion after 18 months.

In patients with cyanotic congenital heart diseases, the right to left shunt allows septic emboli or bacteria to reach the cerebral circulation, avoiding the physiological pulmonary filter.² Moreover, increased blood viscosity may lead to low perfusion and microinfarction in some brain areas where microorganism can sustain growth and multiply to form abscess.³ Brain abscess treatment includes antibiotic therapy and neurosurgical treatment, which should be reserved for patient with unstable condition, with large or multiple brain lesions.³

Ebstein's anomaly is often associated with ASD or patent foramen ovale.⁴ Brain abscess due to paradoxical embolization in Ebstein's anomaly has been described by Lønnebakken *et al.*⁵

In our patient, the combination of elevated right atrial pressure, due to intracardiac anatomy and severe tricuspid regurgitation, with a right-to-left shunt through the ASD increased consistently the risk of paradoxical embolization, which may have been caused by an episode of transient bacteremia.

References

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