An adult patient with previously undiagnosed sinus venosus atrial septal defect presenting with brain abscess: a case report

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Background

Brain abscess is a common complication in children with cyanotic congenital heart disease. The presence of an underlying acyanotic congenital heart disease is usually not suspected in an adult patient presenting with brain abscess.

Case summary

A 51-year-old male patient with no known co-morbidities came with complaints of recent onset right lower limb weakness needing support while walking and on evaluation was found to have brain abscess. He underwent robotic endoscope assisted endoport excision of the brain abscess. Two-dimensional transthoracic echocardiography showed right atrial and right ventricular dilatation with mild low-pressure tricuspid regurgitation. Transoesophageal echocardiography (TOE) revealed sinus venosus atrial septal defect (ASD) with left-to-right shunt with the right upper pulmonary vein draining into superior vena cava. Contrast echocardiography revealed a small transient right-to-left shunt. He has been advised to undergo elective surgical closure of ASD with partial anomalous pulmonary venous connection repair.

Discussion

Right-to-left shunting in ASDs can occur in the early systole even in the absence of raised pressures in the right side of the heart, even when the predominant shunt is left to right, but the magnitude of such a shunt is small and transient and is easily missed. Contrast echocardiography and TOE should be done as a part of evaluation of patients presenting with brain abscess.

Keywords

Case report • Atrial septal defect • Sinus venosus atrial septal defect • Sinus venosus atrial septal defect with partial anomalous pulmonary venous connection • Brain abscess

Learning points

• Brain abscess can occasionally be a presenting manifestation of acyanotic congenital heart disease with predominant left to right shunt.
• Right to left shunting in atrial septal defects can occur in the early systole even in the absence of raised pressures in the right side of the heart.
• A complete cardiac evaluation including contrast echo and transoesophageal echocardiography should be done as a part of evaluation of cerebral abscess.

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**Introduction**

Brain abscess is one of the serious complications of cyanotic congenital heart disease.\(^1\) It most commonly occurs in children and adolescents with a peak incidence at 4–7 years of age.\(^1\) Sinus venosus atrial septal defect (SVASD) are located close to the SVC, and usually above the fossa ovalis, and it constitutes about 2–3% of all interatrial communications.\(^2,3\) These defects do not have a superior margin.\(^3\) It is frequently (80–90%) associated with anomalous pulmonary venous return from the right lung, usually of the right upper or middle lobe pulmonary veins to either the SVC or right atrium.\(^2\) In size, the sinus venosus defect may vary from small (and clinically unrecognizable) to non-restrictive.\(^5\) The superior vena caval vein tends to override the defect, allowing for a biatrial superior caval connection.\(^2\) In contrast to other types of atrial septal defects (ASDs), SVASD is not known to close spontaneously.\(^4\) In a study in 115 patients with SVASD by Josh et al., the usual manifestation was NYHA (New York Heart Association) functional class III or IV dyspnoea that was present in 20% of the patients. Apart from dyspnoea, the following symptoms and history were reported: palpitations (31%), angina (19%), history of congestive heart failure (9%), and history of stroke (3%).\(^5\) Diagnosis of SVASD on transthoracic echocardiography (TTE) is challenging and often necessitates the complementary use of more advanced studies, specifically transoesophageal echocardiography (TOE), cardiac magnetic resonance imaging (MRI), cardiac computed tomography, and rarely, cardiac catheterization.\(^6\) We are reporting a case who presented with central nervous system manifestations due to brain abscess and on evaluation was found to have sinus venosus ASD.

*Figure 1* Above (right and left): T1-weighted contrast magnetic resonance imaging of brain showing ring enhancing lesion seen in left parietal lobe with surrounding oedema. Below: T2 FLAIR (below left) and T2-weighted (below right) magnetic resonance imaging images of brain showing post-operative changes in the left parietal lobe with areas of gradient blooming, suggestive of haemorrhage. Surrounding oedema in the left parietal and frontal lobes. Bifrontal pneumocephalus seen.
Timeline

Presentation of illness

• Day 1: Right upper and lower limb numbness
• Day 3: Right lower limb weakness needing support to walk. Admitted to hospital.

Diagnosis of cerebral abscess

• Day 3: MRI brain revealed T1 hypointense, T2/FLAIR hyperintense, contrast ring enhancing lesion in the left parietal region with perilesional oedema extending to motor cortex.

Emergency surgery

• Day 4: He underwent emergency robotic endoscope assisted endoport excision of the lesion under GA.

Two dimensional echocardiography

• Day 7: Mild RA and RV dilatation with mild low pressure TR.

Sinus venous ASD with left to right shunt

• Day 9: Sinus venous ASD with left to right shunt associated with right upper pulmonary vein draining into SVC. Contrast echocardiography revealed a transient right-to-left shunt.

Plan

• Yet to undergo: Elective surgical closure of ASD with PAPVC repair.

Learning point

• A complete cardiac evaluation including contrast echocardiography and TEE should be done, as a part of evaluation of patients presenting with brain abscess.

Case presentation

A 51-year-old male patient with no known co-morbidities came with complaints of recent onset right upper and lower limb numbness and right lower limb weakness needing support while walking. He was conscious and oriented, obeying commands, afebrile with stable vital signs on admission. His bilateral pupils were 2 mm reactive to light. His white blood counts were 12,470 cells/μL with 84.8% neutrophils and was initiated empirically on Ceftriaxone 2 g intravenous (IV) q8h and Amikacin 500 mg IV q12h. His MRI of the brain revealed a brain abscess in the left parietal region with perilesional oedema extending to motor cortex. There were no other infective foci evident on the MRI, hence any direct diffusion from a contiguous cranial infection site was excluded. He underwent emergency robotic endoscope assisted endoport excision of the lesion (Figure 1). Histopathology of the excised lesion showed glial tissue with dense inflammatory infiltrate comprised of lymphocytes, plasma cells, and histiocytes with focal suppurrative inflammation and necrosis. Culture of the aspirate revealed moderate growth of pansensitive streptococcus species and was continued on the antibiotics for 2 weeks. Blood cultures were negative and dental work-up did not reveal any foci of infection. His human immunodeficiency virus status was negative and there was no history of IV drug use. Computed tomography scan of the chest and abdomen did not reveal any foci of infection. Levetiracetam 500 mg

Video 1 Apical four chambered view showing dilated right atrium and right ventricle.

Video 2 Parasternal short-axis view at the mitral valve level showing dilated right ventricle.

Video 3 Colour Doppler across the tricuspid valve showing mild tricuspid regurgitation.
intravenously q8h was initiated for seizure prophylaxis and was con-
tinued on tablet levetiracetam 500 mg q12h after discharge. As a part
of routine evaluation to find out the source of abscess, cardiology
consultation was sought. Two-dimensional TTE showed right atrial
(RA) and right ventricular (RV) dilatation with mild low-pressure tri-
cuspid regurgitation (Videos 1–3, Figure 2) and hence it was decided to
further evaluate the patient with TOE. TOE revealed SVASD with
left to right shunt associated with right upper pulmonary vein draining
into SVC (Supplementary material online, Video S1, Figures 3 and 4).
Contrast echocardiography revealed the presence of a small transi-
ent right-to-left shunt (Supplementary material online, Video S2, Figure 5).
Other pulmonary veins were seen draining into left atrium. There
was no vegetation. The patient was advised elective surgical closure
of ASD with partial anomalous pulmonary venous connection repair
without which the chances of recurrence is high.

**Discussion**

Brain abscess is focal intracerebral collection of pus that results from
contiguous infection or haematogenous microbial spread and is a fre-
quent manifestation of cyanotic congenital heart disease with right to
left shunt. Since ASD patients have left-to-right shunt, haematogen-
ous abscess is uncommon in these patients. In adults with ASD, the
onset of pulmonary hypertension provides right-to-left shunt predis-
posing to paradoxical embolism and brain abscess. However, in our
patient, pulmonary pressures appear normal at echocardiographic as-
essment. Right to left shunting in ASDs can occur in the early systole
even in the absence of raised pressures in the right side of the heart,
even when the predominant shunt is left to right, but the magnitude
of such a shunt is small and transient and is easily missed and such a
shunt was demonstrated in our patient on contrast echocardiogram
(Supplementary material online, Video S2, Figure 5). Hence this can

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**Figure 2** Above left: Apical four chambered view showing dilated right atrium and right ventricle. Above right: Parasternal short-axis view at the mi-
tral valve level showing dilated right ventricle. Below left: Colour Doppler across the tricuspid valve showing mild tricuspid regurgitation. Below right:
Continuous wave Doppler across the tricuspid valve showing a pressure gradient of 23 mmHg. RA, right atrium; RV, right ventricle.
justify the occurrence of cerebral abscess in our patient in spite of the presence of normal right heart pressures and predominant left to right shunt on echocardiogram. Hence a complete cardiac evaluation including contrast echocardiography and TOE should be done, especially if the suspicion is high as in our case where TTE revealed mild RA/RV dilatation, as a part of evaluation of patients presenting with brain abscess.

**Lead author biography**

Dr Navin Mathew received his MBBS degree in 1996 from Calicut University after his training from Trichur Medical College. Subsequently, he got training from the Stanley Medical College, Chennai and received his MD (Internal Medicine) in 2001. He joined the prestigious All India Institute of Medical Sciences in December 2002 for his cardiology training and was awarded the Jagesh Lal Kapila medal for being the best post graduate in AIIMS cardiology 2005. Since 2006, Dr Navin is in the Department of Cardiology at the Amrita Institute of Medical Sciences and is a valuable member of the heart team.

**Supplementary material**

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

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as *Supplementary data.*

**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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