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CASE REPORTS

A very small sinus venosus type of atrial septal defect: A rare but curable cause of recurrent stroke

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Abstract

Sinus venosus is a rare cardiac defect, which may lead to an interatrial shunt. Diagnosis on echocardiography may be difficult requiring an evaluation by a board-certified cardiologist.
We report a case of a 41 year-old male who presented with recurrent episodes of hemiparesis (first left sided, second right sided). Surgical correction of sinus venosus led to resolution of his symptoms.

INTRODUCTION

Atrial septal defects close to the superior vena cava are called sinus venosus atrial septal defects (ASD). They often include apparent or true anomalous entry of one or both of the right pulmonary veins into the right superior vena cava or the right atrium. Thus, this malformation can cause interatrial shunting leading to rightventricular overload. Clinical manifestations range from mild to severe in degree, with most patients having minimal symptoms. Cerebral ischemic events in the setting of ASD with normal right ventricular compliance are rare.[1-3]

We report a case of of a middle aged male with recurrent strokes, who ultimately required surgical correction of a very small sinus venosus type of ASD.

CASE REPORT

A 41-year-old male was admitted to the hospital with dysarthria and left hemiparesis. He had no other medical problem. He did have a 30 pack-year history of smoking. MR diffusion weighted images showed restricted diffusion in the corona radiata of the right middle cerebral artery territory. (Figure 1a). MR angiography was unremarkable. (Figure 1b).

Laboratory results showed that hemoglobin was 15.5gm/dL, total leukocyte count was 9,700/mm^3 with 70% neutrophils, and platelet count was 229,000/mm^3. Liver and renal function tests, electrolytes, urine examination, and coagulation parameters were within normal limits. Antithrombin3 was 86.5%, lupus anticoagulant was negative, protein S was 60%, and protein C was 97%. Peripheral blood smear was normal. Values for antinuclear antibody, double-stranded DNA antibodies, ANCA and ACA IgG were all within normal limits.

Electrocardiography showed normal sinus rhythm. 24-hour Holter monitoring was unremarkable. Transthoracic echocardiogram demonstrated left atrial enlargement and minimal tricuspid regurgitation. Transcranial Doppler sonography was negative. The initial impression was an ischemic stroke due to small vessel occlusion, and the patient was started on aspirin 300 mg/day. Two months later he came back to our hospital with transient right hemiparesis. MR diffusion weighted images revealed focal restricted diffusion in the superior frontal lobe of the right anterior cerebral artery territory. There was no attributable lesion that could explain the transient right hemiparesis on diffusion weighted imaging. (Figure 1c) Therefore, transesophageal echocardiography (TEE) was performed, which revealed a right-to-left shunt after microbubble injection intravenously and the presence of very small sinus venosus type interatrial communication. (Figure 2a, 2b) There was no abnormal drainage of the right upper pulmonary vein into the superior vena cava on computed tomographic angiography.

Surgical correction of this abnormality was performed under extracorporeal circulation without any complications. After right atriotomy,

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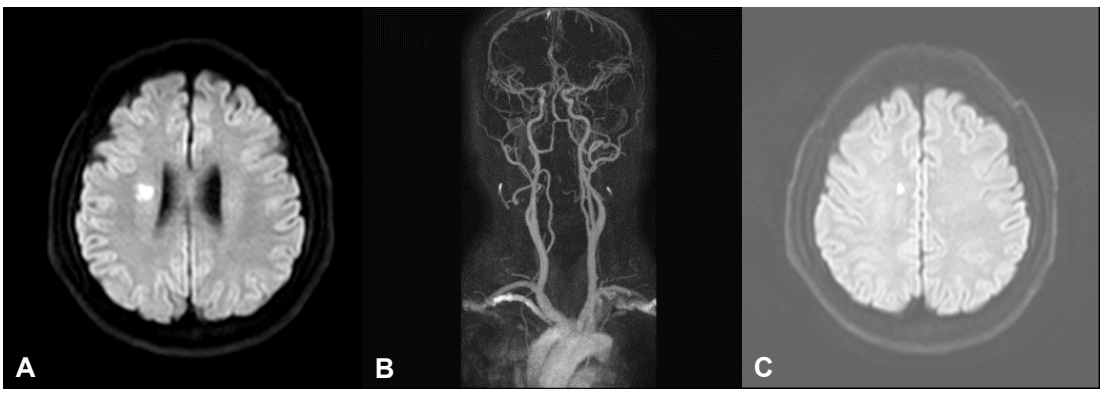

Figure 1: Brain MRI images. Initial diffusion-weighted MRI (A) showing restricted diffusion in the right corona radiata. MR angiography with gadolinium enhancement (B) was unremarkable. Follow up diffusion-weighted MRI (C) demonstrating a new area of restricted diffusion in the right high frontal lobe, which was not consistent with the patient's right hemiparesis.

the interatrial defect was closed. (Figure 2c) Post-operative transesophageal echocardiography did not show any signs of intracardiac shunt, even after injecting microbubble injection intravenously. There was no recurrent stroke after the operation for over a period of 3 years. Follow-up TEE and electrocardiography were normal.

DISCUSSION

Sinus venosus ASD is found in 2% to 10% of patients with ASD. Main cardiac disturbances include pulmonary hypertension, arrhythmias and extrinsic compression of the pulmonary artery when a giant remnant valve of sinus venosus is present.[3,4] Most patients remain asymptomatic during most of their childhood. During their second decade of life, exertional dyspnea or palpitations are common. Arrhythmias and pulmonary hypertension can develop with age.[5] The mechanism of stroke in cases of sinus venosus ASD is not well established. Paradoxical embolism may occur as in patent foramen ovale. Direct embolization from thrombi formed within the aneurysm is another possibility with an associated wall aneurysm. Embolism from ventricular arrhythmia or atrial arrhythmia may occur; possibly enhanced by the transient arrhythmias observed in sinus venosus ASD patients.[3] A false-negative TTE is common in sinus venosus ASD due to its superior location. A TEE performed by specialists in cardiac malformations may be useful in diagnosing and treating this rare condition as surgical repair may be of significant benefit.[3,6] ECG gated multislice CT appears to be a promising tool in exploring abnormalities of cardiac structure and has been described recently in a case of sinus venosus ASD as a supplemental diagnostic tool to echocardiography.[7]

Our patient's initial neurological symptoms occurred in the right middle cerebral artery

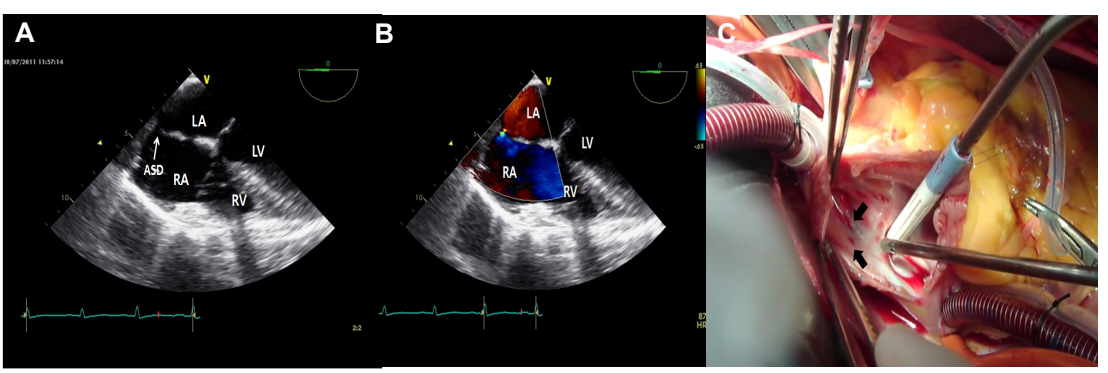

Figure 2: Two-dimensional (A) and color Doppler (B) echocardiographynshowing a small sinus venosus type of atrial septal defect with left to right shunt. During the operation, an internal view of right atrium of heart (C) showing two small sinus venosus types of atrial septal defect (black arrows).
ASD, Atrial septal defect; LV, left ventricle; LA, Left atrium; RV, right ventricle; RA, right atrium.

territory but the following neurological symptoms could not be explained on the basis of right middle cerebral artery stenosis. This feature supports the hypothesis of an embolic process among these patients. A very small sinus venosus ASD was discovered in TEE. This sinus venosus ASD was not hemodynamically significant nor caused any changes in right atrial structure. However, our patient has no additional strokes for the following 3 years after closure of sinus venosus ASD. Closure of sinus venosus ASD is indicated when there is a hemodynamically significant shunt causing enlargement of the right heart. A hemodynamically significant shunt is defined as a pulmonary-to-systemic flow ratio greater than 1.5. As in our case, other indications for intervention include suspicion of paradoxical embolism in the absence of other causes.[8]

This case emphasizes the usefulness of TEE in cryptogenic stroke especially in relatively young patients. The prevalence of patent foramen ovale may be as high as 56% in patients younger than 55 years of age who have had a cryptogenic stroke.[9] Closure of patent foramen ovale has not been shown to be more beneficial than medical therapy alone in CLOSURE-1 trial.[10] However, there have been no randomized controlled trials in patients with sinus venosus ASD. Even small sinus venosus ASD may have a higher risk of embolism than patent foramen ovale. Diagnosing sinus venosus ASD may be challenging but can lead to decisive surgical management. This type of malformation may be overlooked while performing conventional transthoracic echocardiography.

In conclusion, even a very small sinus venosus ASD may be a potentially treatable cause in recurrent cryptogenic stroke. Further clinical trials are required to demonstrate the efficacy of sinus venosus ASD closure in preventing cryptogenic stroke. But clinical trial may not be possible as the condition is very rare.

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