Sinus Venosus–Type Atrial Septal Defect
A Rare Curable Cause of Recurrent Transient Neurological Deficits

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Background and Purpose—Sinus venosus defect is a rare cardiac abnormality, provoking an interatrial shunting outside the interatrial septum. Echographic diagnosis is difficult and may require examination by a specialized cardiologist. Summary of Case—We report the case of a young woman who presented with repeated episodes of hemiparesis. Conclusions—Surgical correction of sinus venosus defect led to disappearance of neurological symptoms. (Stroke. 2006; 37:2385-2386.)

Key Words: cerebral embolism • heart septum • sinus venosus

Sinus venosus defect (SVD) is a rare cardiac abnormality consisting of a disturbed connection of the vena cava to the right atrium, with abnormal pulmonary vein connection to the vena cava. Thus, the malformation provokes an interatrial shunting outside the interatrial septum.1 SVD is found in 2% to 10% of patients with cardiac atrial septal defects. Clinical manifestations range from benign to severe, with most patients having minimal, if any, functional limitation to exercise. Main cardiac disturbances include pulmonary hypertension, arrhythmias,2 and extrinsic compression of the pulmonary artery when giant remnant valve of sinus venosus is present.3 Life expectancy of patients with SVD, however, seems to be shortened. Reports of cerebral complications of SVD are rare, including ischemic stroke.2 Brain abscess4 has been described, possibly attributable to shunting of the natural filter of the pulmonary capillary system, as in Rendu-Weber-Osler disease.5

Case Report
A 14-year-old girl, with personal and familial history of migraine without aura, first presented in 1999 with 3 episodes of transient left-sided hemiparesis and paresthesia within 3 months, followed by 2 episodes of pure motor left-sided hemiparesis within 2 months. Symptoms lasted between 30 and 60 minutes. Transient ischemic attacks (TIAs) were suspected. Brain MRI showed small hyperintense images on T2-weighted images, involving the nucleus caudatus, putamen, and internal capsule of the right hemisphere, consistent with ischemic lesions. Sonography of the cervical arteries showed no abnormality. Transthoracic echocardiography showed no abnormality. Transthoracic echocardiography showed only slight mitral insufficiency. The following tests were negative: head and neck MR angiography, ECG, 24-hour cardiac rhythm monitoring, hypercoagulable screening, lumbar puncture, and mitochondrial DNA screening for MELAS syndrome. Diagnosis of idiopathic regressive ischemic stroke was proposed, and aspirin 100 mg/d was prescribed. Because additional transient left-sided episodes were occurring ~3 to 4 times a year, sometimes accompanied or followed by headache, transesophageal echocardiography (TEE) was requested 1 year later to be performed by a specialist in cardiac malformations; it revealed a massive right-to-left shunting after microbubble injection in an arm vein and the presence of sinus venosus–type interatrial communication, associated with abnormal drainage of the right upper pulmonary vein to the superior vena cava (Figure). No giant valve was observed. Surgical correction of this abnormality was performed under extracorporeal circulation, without any complications. After right atriotomy, interatrial defect was closed with the use of a Dacron device. Enlargement of the superior vena cava was performed with a pericardiectomy, which allowed closure of right atriotomy. No TIA was observed after the operation over a period of 6 months.

Discussion
Probable neurological diagnoses include TIAs and migraine. In both cases, the cardiac origin of neurological problems is highly probable because the surgical correction of the cardiac malformation interrupted the neurological process. Other interatrial shunting diseases are known to predispose to ischemic stroke, including patent foramen ovale.6 Pulmonary shunts in patients with Rendu-Weber-Osler may also lead to ischemic strokes, although this disease mainly predisposes to brain hemorrhage or abscess.3 The mechanism of stroke in cases of SVD is not well established. Paradoxical embolism is possible, as in patent foramen ovale. Embolism from local...
lesions is more probable, possibly enhanced by the transient arrhythmias observed in SVD patients. The importance of cephalalgia must be noted. Indeed, a migrainous syndrome was present in the family. A relationship with the malformation is possible. A higher prevalence of migraine in patients with patent foramen ovale has been described, even if the causal relationship is not clearly understood. A suggested explanation is that cerebral microemboli might favor spreading depression of cortex and induce migraine. A similar mechanism might take place in SVD. In our patient, neurological symptoms occurred in the left side exclusively. This feature does not favor the hypothesis of a cardiac embolic process. However, the attacks did not fulfill current criteria for hemiplegic migraine or migraine with aura.

This case emphasizes the usefulness of TEE in the setting of transient neurological deficits, especially in young patients. Discovery of SVD in this setting is rare, but it can lead to decisive surgical management. This type of malformation may be overlooked at conventional transthoracic echocardiography. A TEE performed by specialists in cardiac malformations may be useful to discover and cure this rare disease because surgical repair may be of significant benefit.

ECG gated multislice CT appears to be a promising tool in exploring cardiac morphological abnormalities and has been described recently in a case of SVD as a supplement to echocardiography.

References


A, Sinus venosus–type interatrial communication consists of abnormal SVC connection to right atrium, with SVC overriding the superior border of interatrial atrial septum. Walls of SVC are then continuous with both left and right atrial chambers. An abnormal pulmonary vein connection to the SVC is present in nearly all cases. (Modified from Bozio A. Le doppler couleur dans l’étude des retours veineux pulmonaires anormaux. In: Davignon A, Hery E, Scheuble C, Bozio A, van Doesburg N, eds. Le doppler couleur dans le diagnostic des cardiopathies congénitales. Paris, France: Pradel; 1992:210.) SVC indicates superior vena cava; ASD, atrial septal defect; RA, right atrium; LA, left atrium; IVC, inferior vena cava; and RPA, right pulmonary artery. B, TEE aspect of our patient, showing the SVC overriding the superior border of interatrial septum and subsequent interatrial communication. Note that this frame is not exactly the same as that in A. RV indicates right ventricle.
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