Cerebral Infarction and Ventricular Septal Defect

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With the availability of contrast echocardiography, patent foramen ovale is frequently detected in patients with stroke, especially in those with no clear etiology and/or the young patient with stroke. Before this report, an association of stroke with ventricular septal defect had not been reported. In this communication, we describe a 38-year-old patient who developed an occipital lobe infarction and who, on investigation, was found to have a ventricular septal defect. Other investigations, which included four-vessel cerebral angiography, collagen disease workup, and coagulation profile, were all normal. We believe this case further extends the spectrum of cerebral ischemic events that may occur with intracardiac shunts. (Stroke 1989;20:957–958)

Despite rapid improvements in imaging technology, the diagnosis of “stroke—uncertain etiology” remains a significant problem. In fact, it is believed that “cryptogenic” stroke may be more common than cerebral infarction secondary to atherosclerotic narrowing of the carotid arteries or thrombosis. Any improvement in diagnostic methods may, therefore, prove very helpful in better understanding the multiple etiologies of stroke. Until recently, paradoxical embolism from a patent foramen ovale (PFO) was considered a relatively rare event, but the introduction of noninvasive contrast echocardiography has made the diagnosis of PFO easy and safe. A number of reports have since appeared that suggest an increased incidence of PFO in patients with stroke, especially where no cause is apparent or the patient is young. In one recent study, the incidence of PFO was 40% in patients with stroke and 10% in control subjects ($p<0.001$). Paradoxical embolism from the venous circulation is presumed to be the underlying mechanism in such cases. In contrast to PFO, other causes of intracardiac shunts have until now not been reported in association with stroke.

In this communication we describe a patient who developed an occipital lobe infarction and, on investigation, was found to have a ventricular septal defect (VSD) by contrast echocardiography.

Case Report

A 38-year-old male was assessed for visual problems that had developed 1 day before admission. Soon after the occurrence of the visual symptoms, he developed a right-sided pounding headache that was present at admission 24 hours later. He had a 2-year history of migraine accompaniments that consisted of 5- to 15-minute episodes of scintillating scotomas in both visual fields that would appear and resolve rapidly. No episodes were ever followed by a headache, nor was there a history of headaches occurring without visual symptoms.

Examination showed a dense left homonymous hemianopia and no other neurological, systemic, or cardiac abnormalities. Cranial computed tomographic scan revealed a hypodense lesion in the right occipital lobe consistent with a cerebral infarction. Four-vessel cerebral angiography was normal. For contrast echocardiography, six injections of normal saline and 5% glucose were recorded. A dense opacification of the right ventricle was immediately followed by the appearance of echoes in the left ventricle during the systole. These findings were consistent with a diagnosis of a transventricular shunt. Hematologic investigations including prothrombin time, partial thromboplastin time, serum immunoglobulins, antinuclear antibody, and erythrocyte sedimentation rate were all normal. Investigations for occult deep vein thrombosis were not done because there was no clinical evidence of its presence. The patient made a gradual recovery and was treated with aspirin; after 3 years, he has developed no new symptoms. However, he still has infrequent episodes of migraine accompaniments.

Discussion

Since the introduction of contrast echocardiography, the detection of pulmonary-systemic shunts
has become very simple. The technique requires the intravenous introduction of echo-dense material, such as air bubbles, which then moves to the right side of the heart. This material is subsequently detected as contrast over the myocardium by conventional two-dimensional echocardiography. Usually air bubbles are contained in 5–10 ml isotonic saline and are introduced by means of an antecubital intravenous injection. Two-dimensional echocardiography can then pick up the air bubbles through the four-chamber view in the right ventricle. In patients with PFO, there is an immediate appearance of the air bubbles in the left atrium. In cases with a VSD, however, there are three patterns that are thought to be diagnostic. These are the appearance of the contrast transversing the ventricular septum in systole, diastolic appearance of the contrast in the left ventricle with left ventricular appearance in the left atrium, and a negative contrast effect seen in the right ventricle. Because there may be no flow in small shunts, a negative study is not uncommon.

Our patient had a history of migraine accompaniments. Although there appears to be an increased frequency of cerebral infarction with migraine, such an increase has until recently not been described with migraine accompaniments. Furthermore, VSD is very rarely associated with cerebral ischemic events, and these usually occur in the setting of advance disease with associated pulmonary hypertension, both of which were absent in this patient. We could not rule out the fact that the infarct was not in some way related to migraine accompaniment or even to a small arteriovenous malformation with previous hemorrhage and spontaneous resolution.

The mechanism leading to stroke in patients with PFO is, at present, unclear. The following mechanism is repeatedly postulated. The primary event is the presence of a thrombus in the venous side of the circulation, commonly thought to develop in the leg veins. Under normal circumstances, atrial and ventricular pressures are higher in the left side of the circulation. During coughing or Valsalva maneuver, transient increases in the right-sided pressures could cause a thrombus that was present on the right side to cross over into the systemic circulation and embolize into the cerebral blood vessels. A mechanism similar to this could also be possible with a VSD. The true incidence of VSD has been difficult to calculate. Incidence in autopsy studies has varied from 0.25 to 1.0%. Because the majority of patients with VSD survive and in many the defect closes spontaneously, autopsy data do not give a true picture of the prevalence of the abnormality. The clinical spectrum of the disease is also very variable. Most patients either have a loud systolic murmur or develop symptomatic heart disease and pulmonary hypertension. A few patients may, however, remain asymptomatic; in these patients, the diagnosis is made with invasive cardiac testing or, more recently, with noninvasive methods such as contrast echocardiography, as in our patient. Treatment in asymptomatic cases such as ours is difficult. Further experience with better identification of milder cases and long-term follow-up may be helpful in deciding if surgical intervention is indicated.

This case further extends the spectrum of cardiac septal defects that could potentially cause cerebral infarction through cerebral embolism and be detected by noninvasive methods. Caution is necessary, however, in interpreting these results because there is still very little evidence that directly links these shunts to subsequent embolic events. PFO is a common condition and may be present in up to 40% of routine autopsies and could thus be present incidentally. VSD usually does not result in cerebral events until cardiac complications develop. In our patient, the associated migraine accompaniments may have contributed to the cerebral infarction. Furthermore, although clinical evidence for venous thrombosis is not present in most patients, this does not rule out the presence of a clot because approximately 50% of patients with venous thrombosis show no clinical evidence of this condition.

References

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