Paradoxical embolization in an adult cystic fibrosis patient

Nabil M Al Lawati MD MRCP (UK), Pearce Wilcox MD FRCPC

CASE REPORT

Cystic fibrosis patients with an implantable venous access device (IVAD) and a patent foramen ovale (PFO) are at an increased risk of developing paradoxical embolism. A 33-year-old patient who had a cerebrovascular accident in the above setting is described. She had been anticoagulated because she had thrombosis of the tip of the indwelling catheter, and her PFO was closed percutaneously followed by replacement of her IVAD. She made a full neurological recovery. Echocardiography and prophylactic closure of the PFO, when present, as primary prevention for paradoxical embolism may be warranted in cystic fibrosis patients before placement of an IVAD.

Key Words: Cystic fibrosis; Paradoxical embolism; Patent foramen ovale

Cerebrovascular accident (CVA) in patients with cystic fibrosis (CF) is a rare event. The occurrence of such an event should prompt the treating physician to look for an intracardiac lesion, such as a patent foramen ovale (PFO), which is a known risk factor for transient ischemic attack and stroke. We report the case of a 33-year-old woman who had a CVA secondary to paradoxical embolism through a PFO.

CASE PRESENTATION

A 33-year-old woman with CF, delta F508 homozygous, complicated by recurrent pulmonary exacerbations, diabetes mellitus, gastroesophageal reflux and pancreatic insufficiency presented with an abrupt onset of headache, diplopia, blurring of vision, temperature fluctuation and gait unsteadiness. Four days before presentation, she had developed intermittent fever and worsening of her long-standing cough. Further review of her past medical history revealed that she had had two transient episodes of visual disturbance over the past few years, for neither of which medical attention had been sought. She had a right-sided venous access device (Port-a-Cath, Smiths Medical MD Inc, USA) implanted 15 years earlier. There had been no overt problems with this access device and it adherent to the tip. An urgent echocardiogram with a bubble study showed a patent catheter with a small thrombus adherent to the tip. An ultrasound Doppler study of the lower limbs showed no deep vein thrombosis. Angiography of the Port-a-Cath showed a patent catheter with a small thrombus adherent to the tip. An urgent echocardiogram with a bubble study showed valsalva- and cough-induced marked right to left shunt through a PFO. The cardiac chambers were normal in size, and the colour flow Doppler examination was normal. Pulmonary artery pressure could not be measured.

On clinical examination, the patient was alert, oriented, afebrile and breathing comfortably with peripheral oxygen saturation of 94% on 2 L/min of oxygen. The site of the Port-a-Cath was unremarkable. She had clubbing, bilateral coarse crackles on auscultation of the chest, and her heart sounds were normal with no murmurs or added sounds. An abdominal examination was normal.

Examination of the central nervous system had revealed a difficulty with short-term recall and no difficulty with long-term memory or attention. Her gait was slightly unsteady, with a tendency to veer to the right. In addition, the patient had slight difficulty with tandem gait, and swayed slightly on Romberg testing.

Tests for antiphospholipid antibody, factor V leiden and prothrombin gene mutation were all negative. An electrocardiogram was normal and an urgent computed tomography scan of the brain showed a 12 mm hypodensity in the medial aspect of the left thalamus that was in keeping with an infarction (Figure 1). An ultrasound Doppler study of the lower limbs showed no deep vein thrombosis. Angiography of the Port-a-Cath showed a patent catheter with a small thrombus adherent to the tip. An urgent echocardiogram with a bubble study showed valsalva- and cough-induced marked right to left shunt through a PFO. The cardiac chambers were normal in size, and the colour flow Doppler examination was normal. Pulmonary artery pressure could not be measured.

A diagnosis of paradoxical embolization was made in view of the above and the patient was started on anticoagulant therapy. Thrombolytic therapy was deferred because of the acute cerebrovascular accident. The patient showed significant improvement, both subjectively as well as objectively, over the next few days. Transesophageal echocardiogram
right to left shunt. The right-sided pressure can further increase with paroxysms of cough, such as the ones our patient had, increasing the risk of and/or worsening a right to left shunt.

While IVADs remain a convenient and relatively safe means of providing venous access for repeated antibiotic therapy in CF patients, there are a number of reported complications of these devices; the most common of these was thrombosis (40%) (7). With such a finding and with the apparently high prevalence of PFO, one should consider echocardiography on CF patients, especially those with severe disease, before placement of IVADs. Although controversial, prophylactic closure of PFO, if present, in an attempt to prevent the potential complication of CVA secondary to a paradoxical embolism, may be justified. There is no evidence for routinely starting CF patients with IVADs on anticoagulant therapy. There are a number of case reports of mechanical embolization of catheter parts; however, we are aware of only a single case of pulmonary embolism secondary to catheter thrombosis in a CF patient (8). Hence, at this point, one cannot recommend routine prophylactic anticoagulation given that this is not a risk-free therapy.

Percutaneous transcatheter device closure of PFO is the preferred means of closing PFO. It has been reported to be safe and effective, with efficacy ranging between 86% and 100% (9,10). The Amplatzer device, such as the one used in our patient, had no thrombus formation compared with other devices in two reviews (11,12).

CONCLUSION

PFO is a common cardiac lesion which is apparently a common echocardiographic finding in CF patients. It is an important risk factor for transient ischemic attack and stroke secondary to paradoxical embolism, especially with indwelling venous access devices. Prophylactic closure as the primary prevention for CVA may be warranted in CF patients before placement of IVADs.

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