68-year-old patient was admitted with right-sided hemiplegia and dysarthria of sudden onset. His medical history included bipolar disorder, past alcohol and cigarette abuse, and untreated arterial hypertension. Clinical examination revealed a pulsatile mass in the upper epigastric region and diastasis recti, which were congenital, according to the patient. The cardiac rhythm was atrial fibrillation. Chest radiography showed rightward deviation of the heart (Figure 1A). Thoracoabdominal computed tomography disclosed protrusion of the left ventricular apex into the epigastric region, containing a round hypodense mass suggestive of a thrombus (Figure 1B). Cerebral computed tomography showed an occlusive thrombus of the proximal left middle cerebral artery without any early sign of brain ischemia (Figure 2A).

Intravenous thrombolysis was clinically unsuccessful. Because of severe hypotension, the patient was intubated. Endovascular mechanical thrombectomy resulted in recanalization of the occluded vessel with delayed filling (final Thrombolysis in Cerebral Infarction score 2). The next day, after extubation, neurological examination revealed global aphasia and right-sided hemiparesis. Brain magnetic resonance imaging (MRI) showed an acute ischemic stroke of the left temporal and parietal cortex, posterior lenticular nucleus, and internal capsule (Figure 2B).

Transesophageal echocardiography and cardiac MRI confirmed the presence of a 3.8-cm-long outpouching of the left ventricular apex through a narrow neck, containing a 1.6×1-cm mobile mass (Figure 3 and Movies I through IV in the online-only Data Supplement). Left ventricular contractility was homogeneously reduced, with 26% ejection fraction. The free wall of the outpouching showed hyperintense signal, late gadolinium enhancement (Figure 3D), and late systolic contraction on cine images (Movies III and IV in the online-only Data Supplement). MRI confirmed the midline thoracoabdominal defect. Anticoagulation and heart failure treatment were introduced. Two weeks later, control echocardiography and cardiac MRI no longer showed a mobile mass in the outpouching. Coronarography performed later in another hospital revealed normal coronary arteries. Six months later, the patient’s hemiparesis and aphasia improved significantly (Modified Rankin Scale score 2). There was no further embolic event.

Congenital left ventricular outpouchings include diverticula, aneurysms, and hernias. Diverticula have a thick wall made up of histological normal heart wall tissue and are connected to the main left ventricular chamber through a narrow neck, whereas aneurysms are thin walled, fibrotic, noncontractile, and have a wide communication with the main chamber. Cardiac hernias are defined as myocardium protruding through a pericardial defect.1,2 Cardiac MRI is a sensitive method for detecting left ventricular protrusions, but the distinction between aneurysm, diverticulum, and hernia may be difficult, and sometimes the final diagnosis can only be established intraoperatively and histologically.

Figure 1. A, Chest x-ray showing rightward deviation of the heart. B, Thoracoabdominal computed tomography, sagittal reconstruction, showing protrusion of the left ventricular apex of the heart into the epigastric region (arrow).
Our patient’s left ventricular apical outpouching associated with a thoracoabdominal midline defect suggested Cantrell’s pentalogy, a malformation syndrome characterized by congenital left ventricular diverticulum associated with defects of the pericardium, anterior diaphragm, lower sternum, and midline abdominal wall.1 The hyperintense magnetic resonance signal and late gadolinium enhancement of the outpouching wall suggested fibrosis, which might result from chronic compromise of blood supply through the narrow hernial neck.3

The prevalence of congenital left ventricular outpouchings was 0.76% in a retrospective study of 12,271 adults undergoing left ventricular catheterization and found to have normal coronary arteries.4 In that study, 5 of 79 patients, all of them with diverticula, suffered embolic stroke attributed to the diverticulum during a follow-up period of 56 months, suggesting that outpouchings present a significant risk of stroke. Other complications included arrhythmia and rupture.

The optimal management of congenital left ventricular outpouchings diagnosed in adulthood is unknown. Surgical removal might improve left ventricular systolic function. Perioperative risk in the case of surgical removal versus risk of recurrent embolism with or without antithrombotic agents should be taken into account. Our patient elected for long-term anticoagulation unless recurrent embolism or symptomatic heart failure should occur. Long-term anticoagulation would also be indicated because of atrial fibrillation.

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Disclosures
None.

References

Figure 2. A, Cerebral computed tomographic arteriography, coronal reconstruction, showing an occlusive thrombus in the proximal left middle cerebral artery (arrow). B, Cerebral diffusion-weighted magnetic resonance imaging showing ischemia in the territory of the left middle cerebral artery.

Figure 3. A, Transthoracic echocardiography, apical view, showing a thrombus inside the apical left ventricular protrusion (arrow). Coronal (B) and sagittal (C) cardiac magnetic resonance imaging showing herniation of the left ventricular apex into the epigastric region. D, Sagittal cardiac magnetic resonance imaging showing late gadolinium enhancement of the wall of the protrusion (arrow).