Rare Case of Recurrent Cardioembolic Stroke Attributed to Multiple Congenital Cardiac Malformations

Gino Chesini, DO, Rajveer Sangera, DO, Trent Howard, DO, David Hockmuth, MD, and David Laughrun, MD, *Des Moines, Iowa*

INTRODUCTION

Congenital left ventricular aneurysms and diverticula (CLVAD) are rare and have a wide range of symptomatology and no established treatment guidelines.[1-9] This case of a 38-year-old woman with recurrent cerebrovascular accidents adds to the growing recognition of a distinct form of cardiomyopathy with an array of potential clinical manifestations. CLVAD may be seen on a variety of noninvasive and invasive imaging studies, but the findings may sometimes be misinterpreted as manifestations of ischemic heart disease. When correctly identified, findings on noninvasive imaging studies are critical for guiding management.

CASE PRESENTATION

A 38-year-old woman was admitted to the hospital with an ischemic stroke. Cardiology consultation was requested to evaluate for possible cardiac source of thromboembolism.

Review of the patient's medical history revealed that a systolic murmur was detected when she was 27 years of age, attributed to mitral valve prolapse with mild to moderate mitral regurgitation by transthoracic echocardiography.

At 34 years of age, the patient was evaluated for an episode of right-sided chest pain at rest that occurred 1 hour after a run. Electrocardiographic findings and cardiac marker levels were normal. A treadmill technetium-99 myocardial perfusion scan was ordered and was interpreted as suggesting probable mid anterior wall infarct and probable small area of apical ischemia (Figure 1). Cardiac catheterization was performed, revealing patent coronary arteries with no obstructive coronary artery disease and an anterior left ventricular aneurysm (Figure 2). Referral was made for cardiac magnetic resonance (Figure 3), which confirmed a midanterior left ventricular aneurysm as well as several areas of late gadolinium enhancement remote from the aneurysm. The latter findings were interpreted initially as two nontransmural and one transmural myocardial infarctions. The possibility of thromboembolic myocardial infarctions was considered, and a hypercoagulable workup was done, which was unremarkable. A lipid panel showed a high-density lipoprotein level of 39 mg/dL and a low-density lipoprotein level of 126 mg/dL. The patient was started on aspirin 81 mg/d and atorvastatin 20 mg/d.

At 37 years of age, the patient presented with left-hand hemiparesis and aphasia. Her symptoms resolved spontaneously within 24 hours. Magnetic resonance imaging of the brain showed a hyperintense signal on the surface of the right cerebral hemisphere, interpreted by radiology as probable ischemic injury. Electrocardiography and telemetry showed sinus rhythm with no atrial fibrillation, carotid duplex was normal, and repeat hypercoagulable workup was unremarkable. Transthoracic echocardiography showed a left ventricular ejection fraction of 56% and mitral valve prolapse with moderate mitral regurgitation. The patient had not been taking aspirin, and it was recommended that she resume aspirin at a dose of 325 mg/d.

Four months later, still at 37 years of age, the patient had <24 hours of right-sided weakness. Head computed tomography showed no evidence of acute stroke, and a diagnosis of transient ischemic attack (TIA) was made. She was advised to continue aspirin 325 mg/d.

At 38 years of age, the patient developed aphasia, left-sided paralysis, and hypoesthesia. Immediate head computed tomography showed evidence of a prior left frontal cortical infarct with no intracerebral hemorrhage. Follow-up magnetic resonance imaging of the brain showed an acute right middle cerebral artery distribution infarct in addition to old changes in the left frontal lobes. Brain and neck magnetic resonance angiographic findings were normal. Electrocardiography and telemetry showed sinus rhythm with no atrial fibrillation.

Cardiology consultants recommended transesophageal echocardiography (Figure 4) to look for cardiac source of embolism. This showed no evidence of thrombus, vegetation, or atherosclerosis. There was no apparent patent foramen ovale (PFO) on a bubble study. A thin-walled aneurysm was present on the anterior wall of the left ventricle, and there were four thicker walled diverticula with preserved contractility of their walls in the inferior distribution. There was mitral valve prolapse with moderate mitral regurgitation.

Because of recurrent stroke and one TIA despite antiplatelet therapy in the setting of the left ventricular aneurysm and multiple diverticula, anticoagulation with warfarin was initiated. Cardiovascular surgery was consulted to consider left ventricular aneurysmectomy to remove the presumed source of her recurrent stroke and TIA. Multiple discussions were had regarding the risks, potential benefits, and alternatives to surgery. The lack of randomized clinical trial data to guide our recommendation was acknowledged, and seeking a second opinion was encouraged. The shared decision was ultimately made to proceed with

From the Mercy Medical Center, Des Moines, Iowa.

Keywords: Left ventricular aneurysm, Thromboembolic stroke, Congenital cardiac malformations

Conflicts of interest: The authors reported no actual or potential conflicts of interest relative to this document.

2468-6441

https://doi.org/10.1016/j.case.2017.09.002

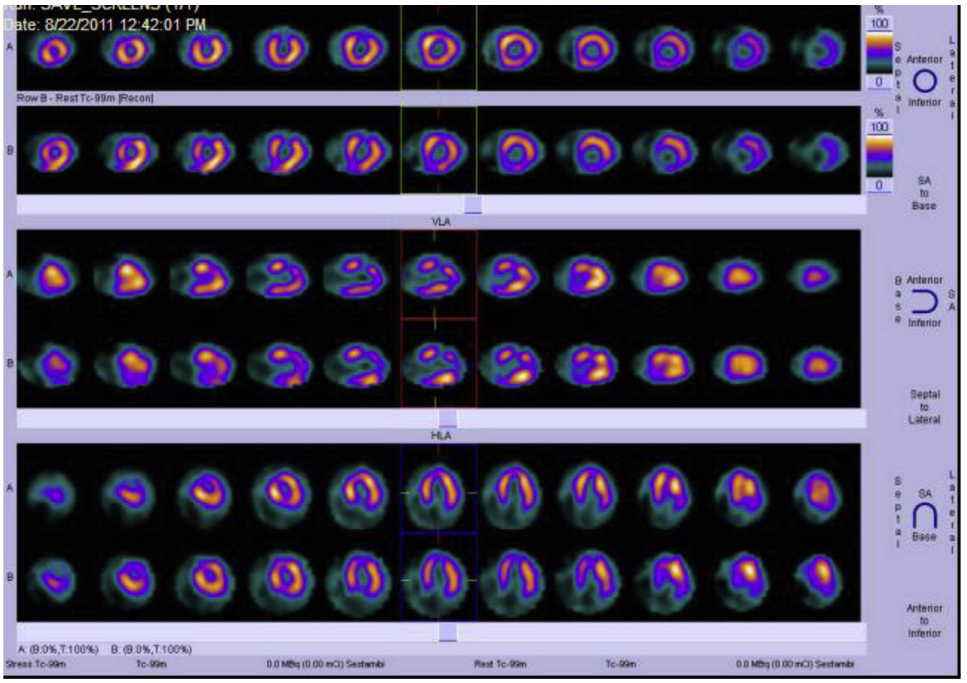

Figure 1 Myocardial perfusion technetium-99 scan demonstrating a fixed anterior defect in the left ventricle, initially interpreted as myocardial infarction but later confirmed as congenital aneurysm.

surgery. Intraoperative findings confirmed an obvious fibrotic aneurysm sac on the anterior surface of the left ventricle (Figure 5), multiple left ventricular diverticula, and fibrotic changes involving the anterolateral papillary muscle. A PFO was present (the initial transesophageal echocardiographic bubble study produced false-negative results). The surgeon proceeded to repair the left ventricular aneurysm with a Hemashield Dacron patch (Figure 5), repair the mitral valve with commissuroplasty and a 27-mm ATS Flex-C band annuloplasty, close the PFO, and ligate the left atrial appendage. Preoperative and postoperative transesophageal echocardiography was performed on the day of surgery to demonstrate aneurysm closure (Figure 6). Heart tissue was sent to pathology for evaluation of the specimen. The pathology report on the excised tissue was consistent with aneurysm. The histologic slides (Figure 7) demonstrated that pathologic fibrosis was apparent, as well as some isolated myocardial fibers. The postoperative course was uneventful. Considering the newly placed left ventricular Dacron patch and pending further clinical follow-up, it was decided to continue warfarin at discharge.

The patient was seen in cardiology outpatient follow-up 6 weeks after surgery and again 1 year after surgery. She was free of any further neurologic symptoms. A shared decision was made to change her therapy from warfarin to clopidogrel.

DISCUSSION

Left ventricular diverticula and aneurysms are outpouching malformations of the ventricular wall.[1] A diverticulum is defined as a muscular outpouching containing all three layers of myocardium that exhibits synchronous contraction.[1] Aneurysms are defined as thin segments that consist primarily of fibrous tissue with minimal unorganized muscular fibers that exhibit either akinesia or dyskinesia.[1-5,10-13] Both malformations are associated with an increased risk for ventricular wall rupture, recurrent thromboembolism, valvular abnormalities, arrhythmias, and heart failure.[1-13] The prevalence of these malformations has been estimated at 0.76%.[12,14]

Formation of congenital ventricular outpouchings begins in the fourth embryonic week.[12,13] Congenital malformations are diagnosed by exclusion when a precipitating event, such as myocardial infarction, trauma, Chagas disease, hypertrophic cardiomyopathy, mucopolysaccharidosis, sarcoidosis, in utero viral infections, connective tissue defects, and Cantrell syndrome, cannot be identified.[1] In our patient's case, none of these alternative diagnoses was present.

The most common presentations of patients with these abnormalities were syncope, rhythm disturbances, and chest pain.[10] Treatment varies from surgery to conservative medical management and must be individualized.

We hope the case presented here will add to the existing case reports in heightening the awareness and understanding of this condition. As awareness increases, earlier and more accurate diagnosis should follow. For example, our patient's presentation with chest pain led to a series of tests, some of which, in hindsight, may not have been necessary if the characteristic findings of CLVAD had been recognized by transthoracic echocardiography. Her myocardial perfusion scan and cardiac magnetic resonance imaging results were both interpreted as indicating ischemic heart disease with prior infarcts and probable ischemia. Notably, the aneurysm detected by single-photon emission computed tomographic and cardiac magnetic resonance imaging did not fit a typical coronary distribution. It was only