Unusual Presentation of Congenital Pseudoaneurysm of Mitral Aortic Intervalvular Fibrosa in a Healthy Child

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INTRODUCTION

Pseudoaneurysm of the mitral aortic intervalvular fibrosa (PA-MAIFV) is a rare condition defined as a pseudoaneurysm at the interannular zone between the mitral and aortic valves and its communication with the left ventricular outflow tract between the left coronary or noncoronary aortic cusp and the anterior leaflet of the mitral valve. It is most commonly associated with infective endocarditis (IE), aortic valve surgery, and chest trauma. Bicuspid aortic valve has also been associated with PA-MAIFV formation. Most cases have been reported in the adult population, with very few cases reported in the pediatric population. We describe a rare case of a previously healthy child who presented with new-onset stroke related to acute IE of the native aortic valve caused by direct valve damage due to turbulent flow from contained PA-MAIFV. We review the possible mechanism, clinical presentation, and management of this unusual disease.

CASE PRESENTATION

A 4-year-old child was referred to our hospital with new-onset left hemiparesis and facial weakness. She was previously healthy, although she had cough and nasal congestion with no apparent fever for 2 weeks before presentation. Initial head computed tomography (CT) revealed an acute infarct in the right middle cerebral artery territory secondary to thrombus in the right internal carotid artery. She was started on heparin infusion for ongoing management of stroke. As part of her evaluation, echocardiography was performed and demonstrated normal cardiac anatomy except for a large cyst-like structure in the mitral aortic intervalvular fibrosa measuring about 4 x 2 cm, consistent with pseudoaneurysm of mitral aortic intervalvular fibrosa, which was protruding into the left atrium and was associated with intact ascending aorta and aortic arch (see Figures 1–3 and Videos 1 and 2).

Other significant findings included moderate mitral regurgitation and free aortic insufficiency related to flail secondary to valve perforation extending to the commissure between the left coronary and noncoronary cusp. No obvious vegetations or clots were found. Urgent cardiac CT was performed, and it was initially reported as possible left sinus of Valsalva rupture into a pseudoaneurysm confined to the extracardiac space with intact ascending aorta and aortic arch. However, the image quality was significantly limited, and on posterior review the cardiac radiologist reported the study as nonconclusive.

The patient was taken to the operating room and underwent pseudoaneurysm resection, aortic root repair, and aortic valve replacement. Intraoperative transesophageal echocardiography confirmed the diagnosis of rupture of pseudoaneurysm of mitral aortic intervalvular fibrosa (see Figure 4 and Videos 3–5).

Direct visual inspection showed a large pulsating pseudoaneurysm completely filling the aortic transverse sinus. It was deemed to be a chronic process, as the walls of the pseudoaneurysm were thick and well formed. With the aortic root opened, it was clear that the origin of the pseudoaneurysm was in the triangle of the left coronary and noncoronary cusps, immediately beneath the hinge points. The aortic valve was completely destroyed because of acute endocarditis process, with friable vegetations all over the valve. This was the suspected source of the acute stroke (see Figures 5A and 5B).

The presumed sequence of events is as follows: remote contained PA-MAIFV that originated from the triangle of the left coronary and noncoronary cusps beneath the hinge points, followed by acute IE of the native aortic valve secondary to turbulent flow that damaged the aortic valve.

The patient’s postoperative course was unremarkable. According to the guidelines, culture-negative endocarditis therapy was completed. Negative infectious workup included multiple bacterial and fungal cultures, viral polymerase chain reaction, and 16s ribosomal deoxyribonucleic acid polymerase chain reaction. From the neurologic perspective, the patient remained stable, and she was started on seizure prophylaxis with levetiracetam, and her heparin infusion was transitioned to enoxaparin. Routine head CT was performed 11 days after admission and revealed cortical laminar necrosis and cortical hemorrhagic transformation in the area of previous stroke. Follow-up head CT revealed interval improvement with decrease in the area of hemorrhagic transformation and necrosis. Electroencephalography did not show seizure activity. Intense occupational and physical therapy was started immediately after admission, and the patient showed significant neurologic improvement. Eighteen days after her admission, she was discharged to a rehabilitation facility to optimize her neurologic recovery.

DISCUSSION

PA-MAIFV is a rare condition that most commonly affects adults. A review by Sudhakar et al. found only <5% of 89 reported cases that involved children <10 years of age. A few cases are reported in
the literature in the pediatric population, most of them in older children than our patient, with the exception of a case of suspected congenital PA-MAIVF in a 3-year-old that did not require surgical intervention. It is unclear whether our patient developed PA-MAIVF secondary to IE or had congenital PA-MAIVF that led to IE secondary to turbulent flow damage of the aortic valve. The former is the most common etiology of PA-MAIVF. The suspected mechanism involves injury of the MAIVF, which is a relatively avascular
fibrous structure, that can result in a pseudoaneurysm formation, which can extend between the left atrium, ascending aorta, and the pulmonary artery into the pericardial space. In our patient the lack of previous infectious focus supports congenital as the most likely etiology for the PA-MAIVF. Additionally, direct intraoperative visualization was significant for a pseudoaneurysm with thick and well-formed walls, which is consistent with a chronic process. Congenital PA-MAIVF as initially described by Abrahams et al. in 1962 is a subaortic aneurysm formed within the aortic annular ring. This intervalvular fibrosa area has been identified as the origin of the majority of left ventricular outflow tract aneurysms despite different predisposing factors. The aneurysm forms a dehiscence between the fibrous ring and the muscular ventricular wall and presents with aneurysmal openings into the left ventricle with subsequent enlargement leading to compression of adjacent structures, including the left atrium, coronary arteries, and pulmonary artery.

One of the differential diagnoses in this case was a sinus of Valsalva aneurysm; this diagnosis was entertained in view of the initial cardiac CT report. By definition, sinus of Valsalva aneurysm is an enlargement of the aortic root area between the aortic valve annulus and the sinotubular ridge. The risk factors and clinical presentation are similar to those of PA-MIVF, but in our case direct visualization of the pseudoaneurysm confirmed its origin below the aortic valve in the intervalvular fibrosa area, which supports the diagnosis of PA-MIVF. Additionally, a rupture of sinus of Valsalva into a pseudoaneurysm would be confined to the extracardiac space; instead, in our patient the pseudoaneurysm was completely filling the aortic transverse sinus.

PA-MAIVF is usually asymptomatic by itself. Symptoms are associated with complications. We suspect that our patient developed an ischemic cerebrovascular event related to septic thrombi secondary to IE of the aortic valve, which was injured by turbulent flow from congenital PA-MAIVF. Neurologic events are some of the most devastating complications of IE and can affect up to 40% of patients. They can range from transient ischemic attacks to ischemic or hemorrhagic strokes, meningitis or encephalitis, and cerebral abscesses. In this setting, cardiac surgery carries high risk for mortality because of the possibility of catastrophic hemorrhagic transformation of the stroke associated with bypass and anticoagulation. We decided to proceed with the surgical approach considering that most recent reports aim for early surgical intervention in cases of ischemic strokes. Even though hemorrhagic transformation of the stroke occurred later after the surgery, it did not have a significant impact on our patient’s clinical status and her neurologic recovery.

CONCLUSION

PA-MAIVF is rare condition that usually affects adults, with very few cases reported in children. The most common etiology is IE, but it can be congenital. Patients are typically asymptomatic, with exceptions related to complications.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2018.02.006.

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