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Case report

A case of pseudoaneurysm of the mitral-aortic intervalvular fibrosa in a pediatric patient ☆☆☆

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ABSTRACT

Pseudoaneurysm of the mitral-aortic intervalvular fibrosa is a rare and potentially deadly aberrance of the cardiac architecture, with few reported pediatric cases. Complications can include rupture into the pericardium, ongoing infective endocarditis, arrhythmias, valvular dysfunction, thrombus formation, and compression of the coronary arteries. Although there have been cases which have gone solely with surveillance, the majority of these cases will have surgical intervention to reduce the risk of complications. We present a case in a pediatric patient with a history of bicuspid aortic valve, who presented with recurrent fevers after a dental procedure despite treatment with antibiotics. Infective endocarditis was suspected based on clinical exam, and imaging revealed pseudoaneurysm of the mitral-aortic intervalvular fibrosa. The patient was treated with surgical resection and homograft replacement.

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Introduction

Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (P-MAIVF) is a rare occurrence, with its most frequently described causative associations being active or prior endocarditis, prosthetic valves, or native valves with anomalies. The mitral-aortic intervalvular fibrosa is fibrous tissue between the aortic annulus and anterior mitral valve leaflet; given its avascularity and thin nature, it is susceptible to infection as well as damage during valvular surgeries [3]. Since 1966, the number of cases

identified in living patients is numbered near one-hundred, with less than 10% being pediatric cases [8]. Complications from P-MAIVF can include rupture into the pericardium, ongoing infective endocarditis, arrhythmias, valvular dysfunction, thrombus formation, and compression of the coronary arteries [8]. Due to severity of the associated complications, expedient surgical intervention is the treatment of choice [9].

We present an uncommon case of a pediatric patient with history of bicuspid aortic valve who presented with recurrent fevers despite antibiotics after a dental procedure, who was found to have P-MAIVF which was treated surgically.

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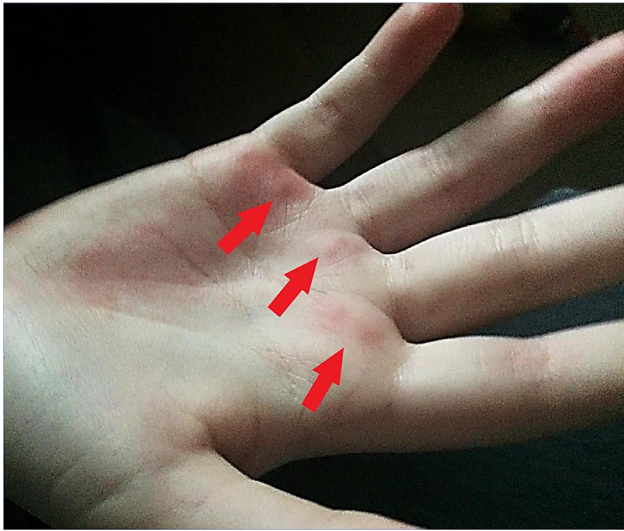


Fig. 1 – Photograph of patient's hand showing Janeway lesions (red arrows) on the palmar aspect of the metacarpophalangeal joints on day of admission (Color version of figure is available online)

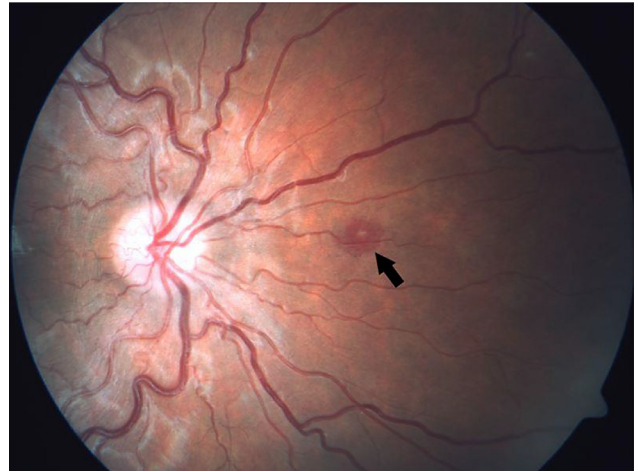


Fig. 2 – Photograph from fundoscopy obtained on hospital day 2 showing Roth spot (black arrow) (Color version of figure is available online)

Case report

Our patient is a 10 year-old male with a history of bicuspid aortic valve and aortic insufficiency who presented with three weeks of daily fever up to 103 F and fatigue. Early in the course of illness after a positive rapid strep throat swab he was treated first with azithromycin and then with cefdinir after a second positive rapid strep test. He re-presented to clinic after 1 week of cefdinir with ongoing fevers and new hand lesions (Fig. 1). His primary care physician suspected endocarditis, and the patient was taken to cardiology for a transthoracic echocardiogram, which noted a new, more echogenic area on the non-coronary cusp of his aortic valve (images not available). The patient was admitted to the Pediatric ward for treatment of infective endocarditis in consult with Pediatric Infectious Disease and Pediatric Cardiology. Further history revealed the patient had invasive dental work about one month prior to onset of symptoms

Blood cultures were drawn prior to the initiation of broad spectrum antibiotics and were repeated every eight hours. Fundoscopic examination on hospital day 2 revealed Roth spot (Fig. 2). From admission until hospital day three, all blood cultures grew *Streptococcus sanguinis*. Cultures remained negative subsequent to hospital day 4, however the patient continued to fever after eleven days of appropriate therapy. This prompted cardiac MRI to be obtained out of concern for cardiac abscess. MRI results were inconclusive for aortic root abscess vs aneurysm (Figs. 3.1 and 3.2); diffusion weighted imaging was not obtained. Subsequent gated cardiac CT with contrast definitively showed P-MAIVF (Fig. 4).

After 48 hours without fever, the patient was discharged with home antibiotic infusion therapy. 7 day follow up echocardiogram identified persistence and enlargement of the

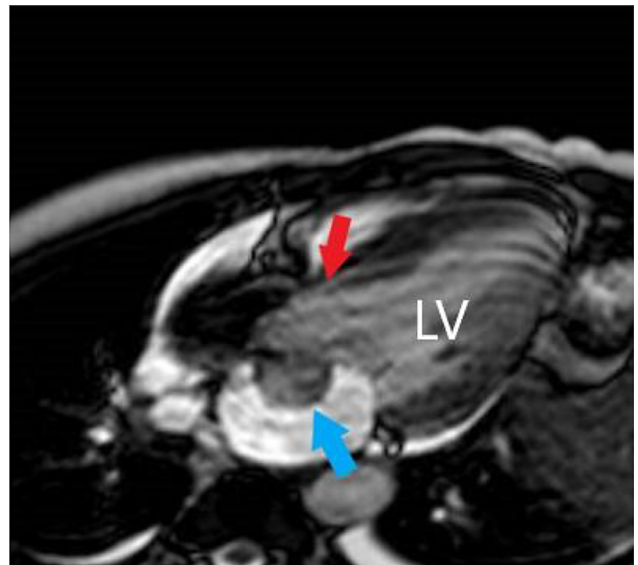


Fig. 3.1 – MR bright blood sequence through the left ventricular outflow tract plane showing the false lumen (blue arrow) along the left ventricular outflow tract (red arrow). Left ventricle (LV) labeled (Color version of figure is available online)

P-MAIVF (Figs 5.1 and 5.2); fever also returned. Given these complications, a decision was made to operate urgently prior to completion of his antibiotic course. He underwent resection of the pseudoaneurysm (Fig 6) as well as homograft replacement of aortic root and valve without complication. Patient recovered well from a cardiovascular standpoint postoperatively.

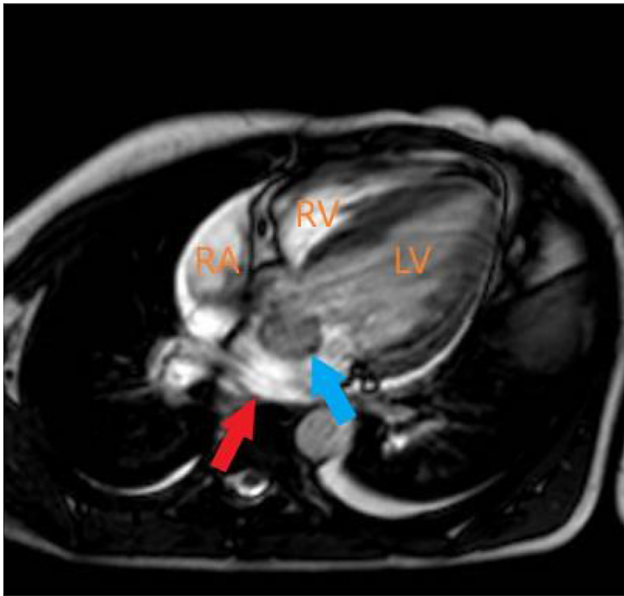


Fig. 3.2 – MR bright blood sequence in four chamber view showing the false lumen (blue arrow) at the mitral-aortic intervalvular fibrosa, expanding towards the left atrium (red arrow). Right atrium (RA), right ventricle (RV), and left ventricle (LV) labeled (Color version of figure is available online.)

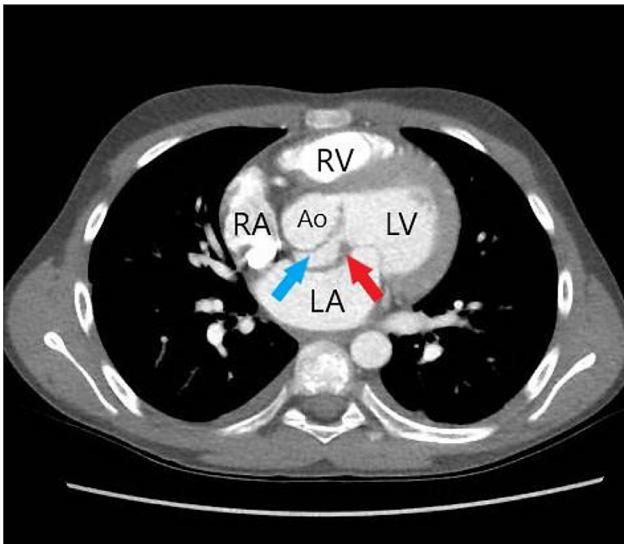


Fig. 4 – Computed tomography angiogram in axial plane showing the false lumen (blue arrow) at the mitral-aortic intervalvular fibrosa (red arrow) communicating with the left ventricular outflow tract, compatible with pseudoaneurysm. Right atrium (RA), right ventricle (RV), left atrium (LA), left ventricle (LV), and aorta (Ao) labeled (Color version of figure is available online)

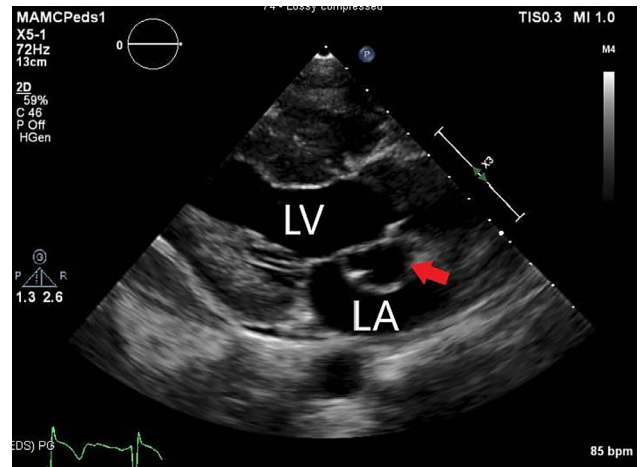


Fig. 5.1 – Echocardiogram with false lumen (red arrow) along the left ventricular outflow tract, enlarged during systole. Left ventricle (LV) and left atrium (LA) labeled (Color version of figure is available online)

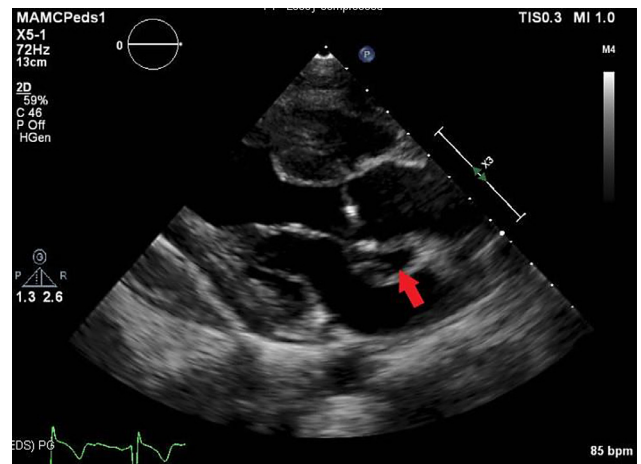


Fig. 5.2 – Echocardiogram with collapse of the false lumen (red arrow) along the left ventricular outflow tract during diastole (Color version of figure is available online)

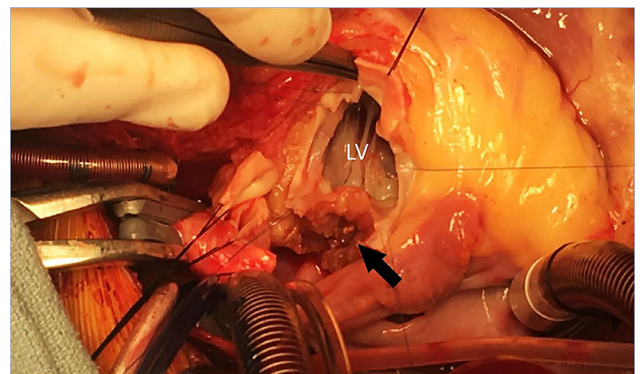


Fig. 6 – Intraoperative photograph during resection and repair of the pseudoaneurysm (black arrow). Left ventricle (LV) labeled (Color version of figure is available online)

Discussion

Pseudoaneurysm of the mitral-aortic intervalvular fibrosa is a rare and potentially fatal complication of endocarditis and very few cases in pediatric patients have been reported to date. To our knowledge, this is the first reported case in a pediatric patient with a bicuspid aortic valve. The complications of such aneurysms in children, especially with aortic regurgitation, have the potential for high mortality [7]. Complications can include fistulae to the atria or aorta, recurrent endocarditis, and compression of adjacent structures, including the coronary arteries, leading to myocardial infarction [9]. Rupture of the P-MAIVF into the pericardium, leading to fatal pericardial tamponade, effusion and pericarditis has also been reported [8]. Additionally due to the close location of the atrio-ventricular node to the MAIVF, atrio-ventricular blocks, requiring pacemaker placement, have been described [5].

Imaging for P-MAIVF is typically performed with echocardiography, with transesophageal being superior to transthoracic [1]. The pseudoaneurysm will typically present as a pulsatile false lumen along the left ventricular outflow tract, expanding and collapsing with systole and diastole [9, 2]. CT and MRI are valuable for preoperative evaluation, as both modalities can show more detailed anatomy of the pseudoaneurysm, as well as surrounding structures [6, 9]. Echocardiography and MRI can also be used to assess for mitral or aortic regurgitation, which is commonly present with P-MAIVF [9].

Due to the potential of complications if left untreated, surgical intervention is usually recommended. The procedure typically involves resection and closure of the pseudoaneurysm and may also include aortic valve replacement [8]. Most patients who have had this diagnosis go on to surgery, and thus there is limited information on the natural progression without surgery. Of the limited cases available, some of the patients who have not had surgery went on to not have any major complications [4]. These cases typically are patients who either refused surgery or were considered a high surgical risk, unlikely to survive the surgery, and instead were managed with close clinical surveillance and serial imaging [4,8]. This type of management would not be recommended for those with high risk features of P-MAIVF, which include active endocarditis, size greater than 3 cm, bicuspid aortic valve, aortic regurgitation, presence of fistula, thrombus formation, or compression of adjacent structures [8]. If a patient does not have surgery, follow up with transesophageal echocardiography is adequate to evaluate for progression of pseudoaneurysm size or development of complications [8]. In our case, given the complications of persistent endocarditis and ongoing fever, surgery was chosen for definitive management.

Patient Consent

Informed consent was obtained from the patient's parents for publishing this case by Dr. Liesemer.

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