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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Rapidly Enlarging Aortic Root Pseudoaneurysm in a Child With Endocarditis and Repaired Congenital Heart Disease

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ABSTRACT

A child with repaired double outlet right ventricle presented with *Staphylococcus aureus* bacteremia. Despite unsuspecting echocardiography on admission and clinical improvement on antibiotics, repeat routine echocardiography detected an aortic pseudoaneurysm, requiring a Ross-Konno operation. In repaired congenital heart defects with bacteremia, close echocardiographic surveillance is required to detect aortic pseudoaneurysm. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:1716–1718) Crown Copyright © 2021 Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

INTRODUCTION

The incidence of infective endocarditis in children, particularly those with a history of cardiac surgery, seems to have increased in recent years (1). The complications of infective endocarditis requiring surgical intervention have had a favorable outcome in the last few decades (2); however, if left undiagnosed or untreated, they can be potentially fatal. Aortic root pseudoaneurysm has been very rarely reported in children (3,4), and thus the actual incidence is unknown. We report a case of an aortic root pseudoaneurysm that rapidly evolved during the first week of illness requiring urgent surgical intervention. We highlight the role of multi-modal imaging to detect this complication.

LEARNING OBJECTIVES

- To recognize that aortic root pseudoaneurysm can be a rapidly evolving complication of infective endocarditis despite adequate antimicrobial therapy.
- To recommend regular imaging in children on treatment for infective endocarditis with underlying cardiac disease for timely diagnosis of aortic root pseudoaneurysm.
- To advocate urgent surgical intervention in aortic root pseudoaneurysm for excellent results.

HISTORY OF PRESENTATION

A 3-year-old boy, with history of surgically repaired ventricular septal defect in the presence of double outlet right ventricle at 6 weeks of age, presented to his local hospital with a 1-day history of fever, rash, and inability to bear weight. He had multiple

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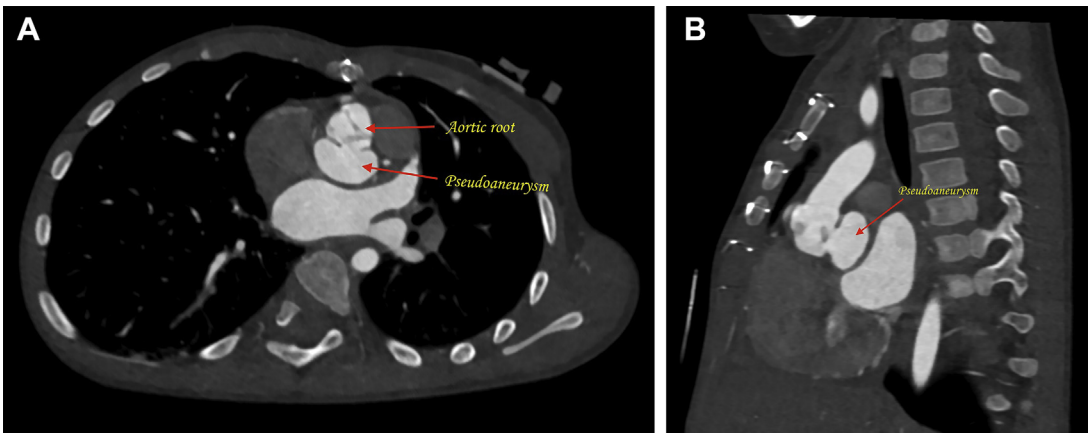
FIGURE 1 Computerized Tomography Images Demonstrating the Aortic Pseudoaneurysm



(A) Axial view of computed tomography scan showing a contrast-filled pseudoaneurysm of the non-coronary cusp with no evidence of rupture. **(B)** Sagittal view of computed tomography scan showing a pseudoaneurysm posterior to the aortic root with a small neck with no evidence of rupture.

nonblanching petechiae with no evidence of splinter hemorrhages or Osler's nodes. On auscultation, the child had normal heart sounds with a long, harsh, late peaking ejection systolic murmur, best heard at the left lower sternal border, radiating to the neck. Both his knee joints were swollen, tender, and warm to touch, right more than left.

PAST MEDICAL HISTORY

Antenatally diagnosed double outlet right ventricle was repaired at 6 weeks of age. During surgery, it was felt that the ventricular septal defect was wide and shallow with a ridge of conal tissue separating the pathway from the ventricular septal defect to aorta and the pulmonary valve. Thus, a part of the conal septum was resected, and a left ventricle to aortic polytetrafluoroethylene baffle was created. Postoperatively, there was mild left ventricular outflow tract obstruction with mild aortic regurgitation. Over the next 2 years, the left ventricular outflow tract obstruction progressed to moderate; however, there was no left ventricular hypertrophy.

DIFFERENTIAL DIAGNOSIS

Initial investigations revealed an elevated C-reactive protein level of 111 mg/L and a normal white blood cell count. Two separate blood cultures were positive for methicillin-susceptible *Staphylococcus aureus*. Empiric intravenous ceftriaxone was changed to intravenous flucloxacillin based on microbiological sensitivity testing. Given the patient's cardiac history and persistent knee pain, he was transferred to our cardiac service for further evaluation and management. The differential diagnosis was bacterial endocarditis complicating septic arthritis.

INVESTIGATIONS

An initial transthoracic echocardiogram (Video 1) showed moderate left ventricular outflow tract obstruction with thickened aortic valve leaflets and mild aortic regurgitation, although no vegetation was evident. Magnetic resonance imaging of the lower limb showed bilateral knee effusions with the right knee aspirate positive for methicillin-susceptible *S aureus*.

There was clinical improvement with resolution of fever and knee pain. C-reactive protein levels decreased, with repeat blood culture results negative for bacterial growth after 3 days of flucloxacillin administration. A routine transthoracic echocardiogram (Video 2) performed 6 days after admission showed an increase in the severity of aortic regurgitation to moderate with an outpouching posterior to the aortic root. For further delineation, a transesophageal echocardiogram was performed (Video 3), which raised the possibility of an aortic root abscess or pseudoaneurysm. To differentiate, a cardiac computed tomography angiogram (**Figure 1,** Video 4) was performed, which showed a pseudoaneurysm of the non-coronary cusp measuring 25 × 20 × 11 mm with the neck measuring 8 mm. A contained rupture could not be excluded.

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MANAGEMENT

Given the rapidity of development of the pseudoaneurysm and the possibility of imminent rupture, the child underwent an urgent Ross-Konno operation, 18 h after the diagnosis was confirmed on computed tomography angiogram, with a pulmonary homograft placed between the right ventricle and the pulmonary artery. The surgery was performed 10 days after the infective endocarditis diagnosis and 7 days after the negative blood culture results. The surgical findings confirmed rupture of the aorta at the left and noncoronary cusp commissure with the formation of a thin-walled pseudoaneurysm adherent to the left atrial roof. The left coronary cusp was thickened and infected. The left ventricular outflow tract was tubular with a subaortic membrane present.

The patient's post-operative course was uneventful. Magnetic resonance imaging of the brain was performed to exclude septic cerebral emboli. Postoperative transthoracic echocardiogram showed excellent function of the aortic autograft and pulmonary homograft. The child completed the recommended course of intravenous flucloxacillin and was discharged with complete resolution of symptoms.

DISCUSSION

An aortic root pseudoaneurysm is a rare complication in adults. However, the incidence in the pediatric population is extremely rare. Mehmet et al (3) reported a case of a giant mycotic aortic aneurysm protruding over the sternal notch in a child with previous cardiac surgery that was successfully surgically excised. Azhar and Abu-Ouf (4) reported a similar case in a 13-year-old child with no history of cardiac disease. Isolated cases of pseudoaneurysm of the left ventricle (5) and left atrium (6) have been reported as a complication of infective endocarditis.

In our case, the initial transthoracic echocardiogram showed no evidence of a pseudoaneurysm; however, within 1 week of illness, the pseudoaneurysm rapidly evolved to a considerable size with increasing aortic regurgitation and likely imminent aortic rupture. Although the antibiotic treatment was effective in controlling the acute infection, the initial bacterial virulence was sufficient to damage the aortic root wall. The flow disturbance related to the preexisting left ventricular outflow tract obstruction might have altered the structure of the aortic root wall, predisposing to an aortic pseudoaneurysm. This case highlights the importance of close imaging surveillance in patients with bacterial endocarditis and pre-existing repaired congenital heart disease, even in the setting of apparently "controlled" clinical signs of infection. Our case also highlights the value of multi-modal imaging to differentiate an aortic root abscess from a pseudoaneurysm.

FOLLOW-UP

The child was last seen 1 year after hospital discharge. He was well, with good aortic autograft and pulmonary homograft function. Lifelong bacterial endocarditis prophylaxis has been recommended.

CONCLUSIONS

Aortic root pseudoaneurysm can be a potentially lethal complication of infective endocarditis in children with a history of cardiac surgery. The rapidity of its growth, despite improvement in symptoms and laboratory markers, warrants regular screening for evidence of its development. Surgical repair in a timely fashion provides extremely good outcomes.

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KEY WORDS double outlet right ventricle, endocarditis, echocardiography

APPENDIX For supplemental videos, please see the online version of this paper.