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EDITORIAL COMMENT

Infected Pseudoaneurysm in Congenital Heart Disease*

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Infective endocarditis is a known but rare entity in children, even in those with congenital heart disease. Over the past decade, there has been minimal increase in incidence despite significant changes in prophylaxis recommendations (1,2). When present, infective endocarditis can have devastating effects on the heart and surrounding structures, including atrioventricular and semilunar valve destruction, abscess formation, conduction abnormalities, repair dehiscence, and pseudoaneurysm formation.

In this case report, Singh et al (3) present a young child with double outlet right ventricle with subaortic ventricular septal defect repaired with ventricular septal defect baffle in infancy who developed fever, rash, and joint pain with elevated inflammatory markers and a positive blood culture for methicillin-susceptible *Staphylococcus aureus*. His presentation was remote from his surgical procedure. Although symptoms and inflammatory markers improved and blood cultures became negative with intravenous antibiotic therapy, a routine transthoracic echocardiogram at day 6 of illness revealed the development of a large aortic root pseudoaneurysm that was not present on the initial echocardiogram. Transesophageal echocardiography and cardiac computed tomography confirmed the diagnosis and it was noted that the pseudoaneurysm had rapidly enlarged over the previous days. Urgent surgery with Ross-Konno operation was performed, and surgical inspection confirmed rupture of the aortic root wall between aortic leaflet commissures with contained pseudoaneurysm. After surgery, the child was treated with intravenous antibiotics for the usual course and did well.

Pseudoaneurysm is a well-recognized but unusual complication of infective endocarditis and can occur within the heart (eg, ventricular or mitral-aortic intervalvular fibrosa, right ventricle to pulmonary artery conduit) or at the level of the great arteries (eg, aortic root, descending aorta) (4-6). In the setting of infection, vulnerable regions of the heart likely develop weakening that leads to outpouching and ultimately to contained rupture. Many congenital heart defects, such as tetralogy of Fallot and bicuspid aortic valve, are known to be associated with intrinsic aortopathy and aortic root dilation, which likely makes this region of the heart vulnerable to infective pseudoaneurysm (7,8). The authors surmised that the left ventricular outflow tract obstruction may have damaged the aortic root wall similar to what is seen in bicuspid aortic valve disease.

This case report demonstrates how confirmed endocarditis can progress rapidly and become life-threatening even as a patient appears to show clinical improvement. The lesson learned is that frequent echocardiographic surveillance is warranted in cases of infective endocarditis in congenital heart disease to ensure that vegetative lesions have not progressed and that new lesions have not developed. Rapidly enlarging pseudoaneurysm in the setting of infective endocarditis is at significant risk of rupture because the wall of the pseudoaneurysm is only scar tissue; such a finding is a surgical emergency. Singh et al (3) should be commended for their meticulous care of this patient, timely diagnosis, accelerated timing of intervention, and his ultimate good outcome.

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REFERENCES

1. Baltimore RS, Gewitz M, Baddour LM, et al. Infective endocarditis in childhood: 2015 update. *Circulation*. 2015;132:1487–1515.

2. Rushani D, Kaufman JS, Ionescu-Ittu R, et al. Infective endocarditis in children with congenital heart disease: cumulative incidence and predictors. *Circulation*. 2013:1412–1419.

3. Singh AA, Gnanappa GK, Orr Y, et al. Rapidly enlarging aortic root pseudoaneurysm in a child with endocarditis and repaired congenital heart disease. *J Am Coll Cardiol Case Rep*. 2021;3:1716–1718.

4. Ahmed A, Shivaram P, Zakaria D. Pseudoaneurysm of the mitral-aortic intervalvular fibrosa following endocarditis and aortic valve surgery in an infant—case report and exhaustive systematic review of pediatric cases. *Echocardiography*. 2020;37:1495–1505.

5. Jeng EI, Martin TD. Simultaneous aortic valve endocarditis and mycotic pseudoaneurysm of aortic coarctation. *J Card Surg*. 2020;35:1714–1716.

6. Tomar M, Bhan A. Rapidly progressing giant aneurysm of right ventricular outflow tract with severe conduit obstruction: report of two cases. *Images Paediatr Cardiol*. 2018;20:1–7.

7. Niwa K, Perloff JK, Bhuta SM, et al. Structural abnormalities of great arterial walls in congenital heart disease: light and electron microscopic analyses. *Circulation*. 2001;103:393–400.

8. Stock S, Mohamed SA, Sievers HH. Bicuspid aortic valve related aortopathy. *Gen Thorac Cardiovasc Surg*. 2019;67:93–101.

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