Case Report

Infective endarteritis in a case of supravalvular aortic stenosis

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\textbf{ABSTRACT}

Supravalvular aortic stenosis is the rarest form of congenital left ventricular outflow tract obstruction. It can be associated with Williams syndrome or it may be an isolated anomaly. Although infective endocarditis is common in valvular aortic stenosis, in supravalvular aortic stenosis infective endocarditis/endarteritis is rarely reported. We report a case of infective endarteritis in a case of supravalvular aortic stenosis due to resistant enterococci, causing prolonged fever. Blood culture-directed antibiotic therapy cured the infection.

\textbf{Learning objective:} Infective endarteritis in supravalvular aortic stenosis is a rarely reported but treatable entity. Although it may respond to empirical antibiotics, occasionally some resistant organism, causing infective endarteritis, may not respond to empirical therapy. Multiple blood culture with culture-directed antibiotic administration may be lifesaving, as in our index case.

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\textbf{Introduction}

Supravalvular aortic stenosis (AS) is the rarest form of congenital left ventricular outflow tract obstruction. It can be associated with Williams syndrome or it may be an isolated anomaly \cite{1,2,3}. The incidence of supravalvular AS is six in 10,000 to one in 20,000 live births \cite{4}. Although infective endocarditis is common in valvular AS, in supravalvular AS infective endocarditis (IE)/endarteritis is rarely reported in the literature. Here we report a case of isolated supravalvular AS with infective endarteritis having multiple vegetations in the whole of the ascending aorta. He was cured with proper antibiotic administration.

\textbf{Case report}

A six-year-old boy presented with documented fever for 4 months despite being treated empirically with multiple antibiotics prior to admission. On physical examination, he had pallor and splenomegaly. An ejection systolic murmur was audible over the precordium, best audible over aortic area with radiation to carotid. His facies and intelligence were normal. His chest X-ray was unremarkable. Electrocardiography showed no gross abnormality. Echocardiography revealed normal aortic valve (tricuspid, normal, and symmetric cusp opening) (Fig. 1A) but supravalvular narrowing just distal to the aortic sinus of Valsalva (Fig. 1B and Video 1). The suprasternal view confirmed the supravalvular aortic stenosis with turbulence in the ascending aorta. The peak and mean gradient across the narrowing were 58 mmHg and 35 mmHg, respectively (i.e. moderate AS) (Fig. 2A). Almost whole of the ascending aorta starting from the aortic sinus up to the origin of brachiocephalic artery was full of multiple varying sized mobile vegetations (Fig. 2B, Videos 1 and 2).

Other valves and septum were normal. No pulmonary stenosis was present. No intra-cardiac vegetation was seen. The transesophageal echocardiography (TEE) confirmed the highly mobile numerous vegetations extending from the aortic sinus up to origin of the brachiocephalic artery (Fig. 3).

His blood culture grew enterococci (on two separate occasions) which were penicillin-resistant (minimum inhibitory concentration > 0.5 μg/ml) and was treated according to culture sensitivity. He became afebrile after 10 days of antibiotic therapy with vancomycin and gentamycin. Antibiotics were continued for 4 weeks.

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His blood calcium level was normal. He did not undergo gene analysis as his phenotype was normal with normal pulmonary valves.

Discussion

Supravalvular AS is the rarest of the three varieties of congenital AS. It is associated with Williams syndrome in 30–50% of cases. About 20% of cases are familial but without features of Williams syndrome, and the remaining cases (i.e. about half of all patients) appear to be sporadic [1–3]. Williams syndrome is associated with a microdeletion in chromosome region 7q11.23, encompassing elastin gene. Reports suggest that this microdeletion of elastin gene is responsible for typical vasculopathy of Williams syndrome, supravalvular aortic stenosis (SVAS), and pulmonary artery stenosis [5,6]. Our index case neither had any feature of Williams syndrome nor any family member affected, thus falls under the sporadic group.

Although the literature, in general, has described supravalvular AS as a predisposing factor for IE, there is only a single case report till date by Maruyoshi et al. [7], describing a case of intractable endocarditis associated with supravalvular AS. That
case also had intracardiac vegetations with mitral and aortic valve involvement. Our index case had vegetations only in the ascending aorta without involvement of any other intracardiac structure. Supravalvular narrowing with turbulent jet seemingly had predisposed him to infective endarteritis. It is a rare but treatable entity. Although they may respond to empirical antibiotics, occasionally some resistant organisms, causing IE, may not respond to empirical therapy. Multiple blood culture with culture-directed antibiotic administration may be lifesaving, as in our index case.
**Conflict of interest**

The authors declare no conflict of interest.

**Appendix A. Supplementary data**

Supplementary data associated with this article can be found, in the online version, at http://dx.doi.org/10.1016/j.jccase.2014.09.003.

**References**


Fig. 3. Trans-esophageal echocardiography showing multiple mobile vegetations in aortic root and ascending aorta. Ao, aorta; LV, left ventricle.