

Non ST-Elevation Myocardial Infarction in a Patient with Supravalvular Aortic Stenosis. Role of Multi-Modality Imaging

Dear Editor,

Supravalvular aortic stenosis (SVAS) is a congenital narrowing of the ascending aorta above the level of sinus of Valsalva. It is most often commonly associated with William's syndrome although it can arise without syndromic associations as well.^{1,2} Left main coronary artery (LMCA) occlusion or stenosis has been seen to be associated with the condition, and can be a cause of sudden cardiac death in such patients.^{3,4,5,6}

Case Report

A 29-year-old female was admitted to our centre with chest pain and shortness of breath while performing strenuous aerobic exercises. She had a history of non-syndromic congenital supravalvar aortic stenosis (SVAS) and was non-compliant to follow-ups. She was previously offered surgery but had refused surgical intervention.

Upon presentation, her electrocardiogram was consistent with that of a left main coronary artery (LMCA) occlusion and showed for global deep ST depressions with reciprocal ST elevations in aVR. Her symptoms abated promptly with rest in the Emergency Department, with resolution of accompanying electrocardiographic changes (Fig. 1). A bedside echocardiogram showed intact wall movements. Serially trended troponin I measurements peaked at 31.6 ug/L (normal <0.039).

She was admitted to the Coronary Care Unit and managed conservatively with close monitoring of haemodynamics. Cardiac computed tomography (Fig. 2) showed normal

coronary vessels with an elongated left coronary cusp extending to the sinotubular junction, in close relation to the left main coronary ostium, as well as narrowing of the sinotubular junction in keeping with SVAS. On cardiac magnetic resonance imaging (Fig. 3), there was no late gadolinium enhancement in keeping with a myocardial infarction, or evidence of myocardial oedema. The sinotubular junction was also noted to be narrowed at 13 mm, whereas the annulus was 18 mm, sinus of Valsalva was 22 mm and the ascending aorta was 16 mm in diameter. The peak gradient across the supra-aortic and aortic valve segments was 63 mmHg, consistent with severe stenosis.

The patient recovered well with conservative management and was discharged with advice against excessive exercise. She was given an early appointment to consult with a view to early surgical intervention of SVAS.

Discussion

SVAS has been well described in the literature, and has associations with LMCA occlusion.^{3,4,5,6} There have also been reported cases of sudden cardiac death from occlusion of the right coronary ostium.⁷ It is most classically associated with William's Syndrome although it can exist independently of syndromic conditions as well.^{1,2} A case series of 9 patients described 3 patterns of LMCA occlusion including ostial narrowing (type 1), cusp-ridge fusion (type 2), as well as fusiform narrowing of the LMCA (type 3), and suggested differing surgical approaches for each pattern.⁸

Presentations of LMCA occlusion in association with

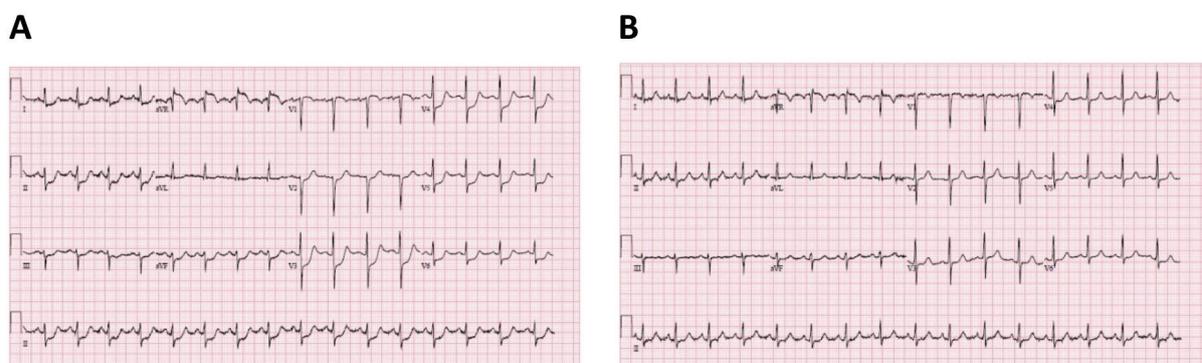


Fig. 1. Electrocardiograms upon presentation and after rest. A) Electrocardiogram shows global ST depressions with a reciprocal ST elevation of lead aVR. B) Electrocardiogram shows for resolution of ST changes with rest.

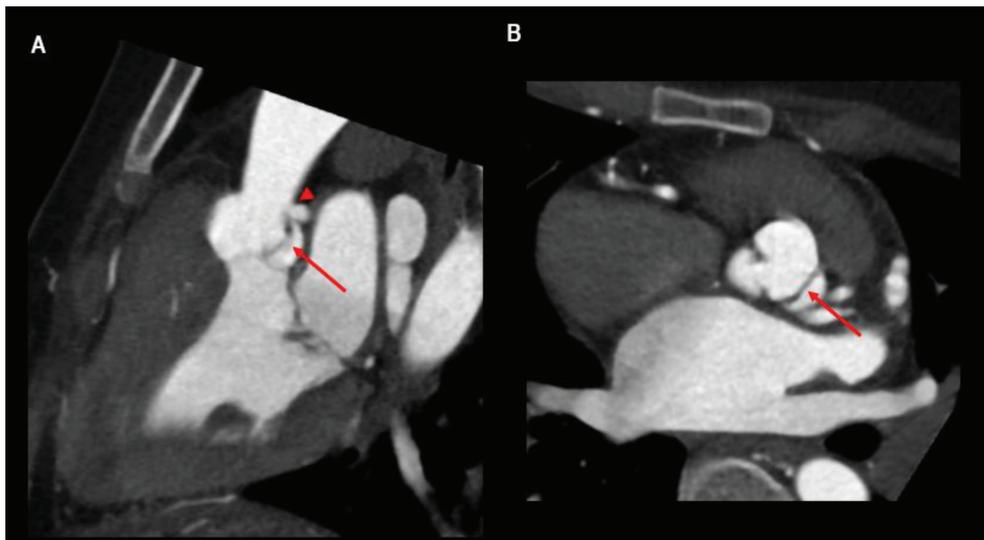


Fig. 2. Cardiac computed tomography of SVAS and origin of LMCA. Multiplanar reformats from the CT coronary angiogram study in A) left ventricular outflow tract view and B) enface view of the aortic valve at the level of the sinuses of Valsalva. This shows the elongated left coronary cusp (arrow). The tip of the left coronary cusp is seen in close relation to the ostium of the left main coronary artery (arrowhead).

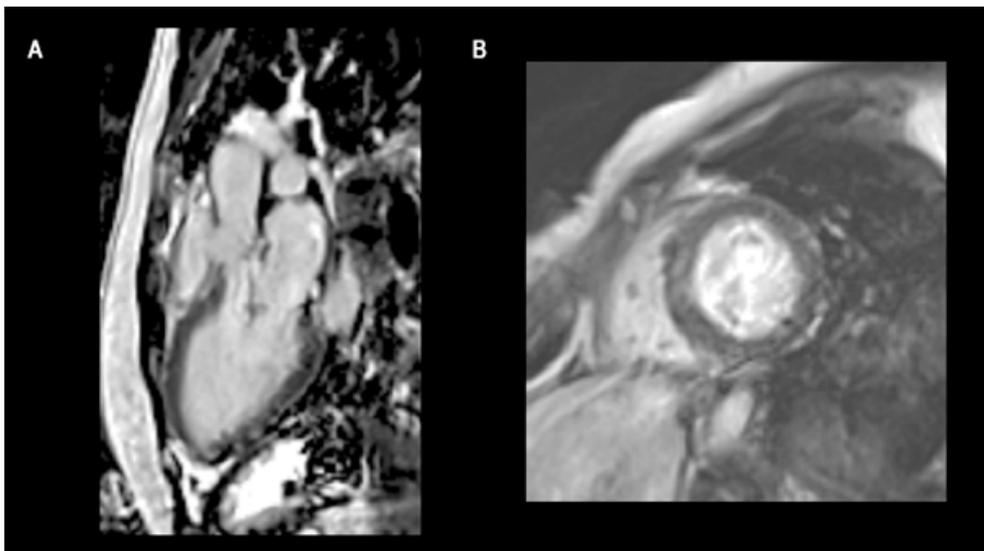


Fig. 3. Late gadolinium enhanced magnetic resonance images of the heart. Post-contrast high resolution inversion recovery magnetic images of A) 3-chamber and B) basal short axis views do not show any late gadolinium enhancement to suggest myocardial infarction, or any regional wall motion abnormalities. The sinotubular junction was also noted to be narrowed at 13 mm, whereas the annulus was 18 mm and the sinus of Valsalva was 22 mm. The peak gradient across the supra-aortic and aortic valve segments was 63 mmHg.

SVAS can be as a myocardial infarction or sudden cardiac death in a paediatric population.^{3,4,5,7} Treatment tends to favour a surgical approach, combining an aortic root reconstruction for SVAS as well as either surgical repair of the left main coronary ostium or emplacement of a bypass graft.⁸ In this specific case of LM coronary occlusion from cusp-ridge fusion (type 2) and SVAS, the recommended approach was excision of the valve leaflet from the aortic wall with preservation of all possible leaflet tissue, followed by an aortotomy and single patch aortoplasty with native pericardium.⁸

In the above patient, we believe that the strenuous exercises undertaken had resulted in a dynamic obstruction of the left main coronary ostium due to her unique anatomy with an ensuing presentation of chest pain with features suggestive of LMCA occlusion. Coronary artery stenosis or obstruction should hence be considered in all patients with SVAS.

The authors suggest that a non-invasive approach with close monitoring and early cardiac imaging as the above could be a viable and safe alternative to invasive coronary angiography in patients with such a presentation.

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