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**TITLE:** Intramyocardial calcification in a 37 year old patient with severe aortic stenosis

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**Short Running Title:** Intramyocardial calcification in a 37year old patient with severe aortic stenosis

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TITLE: Intramyocardial calcification in a 37-year-old patient with severe aortic stenosis

CASE REPORT
A 37-year old man with symptoms of progressive dyspnoea and intermittent chest pain over a one year period with a longstanding history of aortic stenosis (AS) was referred for aortic valve replacement. His personal cardiovascular history was unremarkable, most notably for tuberculosis, rheumatic heart disease, pericarditis or myocardial ischaemia. Family history was non-contributory. Examination revealed an ejection systolic murmur consistent with aortic stenosis. Vital signs revealed an early warning score of zero, laboratory investigations were found within normal parameters and Electrocardiogram (ECG) was in sinus rhythm. Echocardiography (ECHO) demonstrated an ejection fraction of 55% and confirmed severe AS and moderate aortic incompetence with peak and mean gradients of 90 and 50mmHg respectively. Extensive myocardial calcification on ECHO (Figure 1) and coronary angiography (Figure 2) prompted further investigation with CT. CT was notable for a thick band of intramyocardial calcification extending from beneath the left coronary cusp to half way down the interventricular septum (Figure 3). Calcification was not noted in any other organ system and there was no evidence of malignancy. Serum calcium levels were within normal range. Cardiac catheterisation revealed no evidence of coronary artery disease.

Aortic valve replacement was performed. Intraoperative findings revealed a heavily calcified aortic valve. The native valve was meticulously excised and replaced using a 25mm carbomedics mechanical prosthesis with interrupted sutures. Post-operative echocardiogram was satisfactory and postoperative recovery was uneventful.

DISCUSSION
Epidemiological studies reveal that aortic stenosis increases with age, 25% of people aged over 65 have aortic sclerosis, and 3% over 75 have severe stenosis. Approximately, 16% of patients with sclerosis progress to stenosis in seven years [1]. The overwhelming cause of aortic stenosis is calcific degeneration, 2% of the population account for congenital bicuspid valves, rheumatic disease is now
extremely rare [2]. Cases of myocardial calcification which are not associated with myocardial infarction or metastatic deposition are extremely rare [3, 4]. Calcification of the myocardium can occur secondary to two mechanisms. Firstly, dystrophic calcification occurs in dead or degenerative tissue in the presence of normal calcium/phosphate balance such as previous myocardial infarction. Other potential causes of dystrophic calcification include myocarditis, ventricular aneurysms, tuberculosis, sarcoidosis and haemorrhage [5-9]. Secondly metastatic calcification, which occurs when a derangement of calcium phosphate metabolism results in calcium deposition in normal tissue, such as chronic renal failure or hyperparathyroidism [3]. Our patient had no known history or aberrant biochemical analyses to suggest that any of the differentials mentioned were the causative pathology. Extensive calcification and porcelain heart due to endomyocardial fibrosis has also been reported in the literature [10]. Endomyocardial fibrosis is characterised by the presence of fibrous tissue in the endocardium eventually extending into the myocardium, a finding not identified in our patient. Mitral annular and leaflet calcification occurs frequently with degenerative aortic stenosis and is known to reduce leaflet opening and result in significant mitral stenosis in 25% of patients with calcific aortic stenosis [11]. In our patient, however, no involvement of the mitral valve was noted.

CONCLUSION

This is an unusual case as significant calcification of the interventricular septum developed in the absence of any known aetiologic factor or evidence of pre-existing myocardial injury. Valvular heart disease, is an underappreciated yet serious and growing public health problem

Keywords: Aortic Stenosis, Intramyocardial calcification, Aortic valve replacement, cardiothoracic surgery, Symptomatic Valvular Diseas

Conflict of Interest

None
LEARNING POINTS

- Intramyocardial calcification is extremely rare in the absence of cardiovascular history.
- Intramyocardial calcification can be associated with dystrophic and/or metastatic calcification like chronic renal failure or hyperparathyroidism.
- It requires a thorough diagnostic work up with CT and coronary angiogram evaluation to determine the extent of calcification and underlying valve disease.
- Surgery is generally only indicated if there is significant valve pathology, such as in this case.

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degenerative aortic stenosis: real-time 3D transoesophageal

ABREVIATIONS
ECG Electrocardiogram
CT Computed Tomography
AS Aortic Stenosis
ECHO Echocardiography

FIGURE LEGENDS

Figure 1: Pre-operative transthoracic echocardiogram demonstrating abnormal
calcification in the interventricular septum

Figure 2: Coronary angiogram demonstrating intramyocardial calcification relative to
the left anterior descending and circumflex arteries

Figure 3: CT thoracic angiogram (coronal section) demonstrating aberrant
calcification extending from the aortic valve to the interventricular septum
Figure 1: Pre-operative transthoracic echocardiogram demonstrating abnormal calcification in the interventricular septum (LA: Left Atrium, Ao: Aorta)
Figure 2: Coronary angiogram demonstrating intramyocardial calcification relative to the left anterior descending and circumflex arteries.

Figure 3: CT thoracic angiogram (coronal section) demonstrating aberrant calcification extending from the aortic valve to the interventricular septum.