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ORIGINAL RESEARCH

"Coconut atrium": A case report and review of the literature

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Summary

"Coconut atrium" or complete dystrophic left atrial wall calcification is rare and occurs almost exclusively in the middle-aged and the elderly, often following chronic conditions such as rheumatic heart disease, end-stage renal disease and tuberculosis. Hyperparathyroidism, lipid storage disease and use of xenografts are known causes of incomplete intra-cardiac dystrophic calcification in older children. The present report describes a rare case of cardiac calcification demonstrated by Trans-thoracic 2-D Echocardiography in a two-year old Indian boy who presented at a private hospital in Hyderabad, India with breathlessness. Unfortunately, his parents declined further extensive investigations and treatment, but this case was instructive due to the rarity of complete dystrophic left atrial wall calcification, the exclusion of the common known aetiologies and the extremely young age of the patient.

Key words: Atrium of stone, Coconut heart, Dystrophic cardiac calcification, Left atrial calcification, Porcelain atrium, Porcelain heart, Stone heart.

Introduction

"Coconut atrium" is a relatively rare medical entity. [1] It has variously been described as atrium of stone, coconut heart, heart of stone, porcelain atrium, porcelain heart and stone heart. [2 - 6] This form of cardiac calcification is frequently diffuse and occurs predominantly among older patients [7 - 9] and usually those with chronic inflammatory conditions such as rheumatic heart disease, [10, 11] in end-stage renal disease, [4] following endocarditis, [12] and as a complication of radiotherapy for malignancy. [13]

Dystrophic cardiac calcification has also been reported, though rather infrequently, in children following hyperparathyroidism, [14] tuberculosis, [15] and myocardial damage arising from corrective surgery for cyanotic congenital heart disease with xenografts. [16] Intra-cardiac calcification had earlier been reported in a 12-year old Indian girl with Gaucher's disease, a rare genetic lysosomal storage disorder. [17] Cardiac calcifications, which may involve the coronary arteries, have also been documented in children with end-stage renal disease, [18] haemochromatosis [19] and some idiopathic cases. [20]

This report describes a preschool aged child who had a massive left atrial calcification associated with bicuspid aortic valve. To the best of the authors' knowledge, such severe complete left atrial calcification has never been reported in children this young. Therefore, the case is reported to create awareness and with a review of the existing literature.

Case Description

A 27-month old Indian boy presented at the Krishna Institute of Medical Sciences (KIMS) Hospital, Hyderabad, India with a month history of progressively worsening laboured breathing. History of fever and weight loss, were denied. He had an apparently uneventful past medical history and his parents were peasant farmers. The physical examination was not particularly remarkable except for being underweight; the body weight was 8.8 kg (-3.87 z), the height was 75 cm (-4.14 z), the body surface area (BSA) was 0.43 m^2 and the Body Mass Index (BMI) was 15.6 kg/m^2 (- 0.69 z or 25th percentile). [21] There was no significant peripheral lymph node enlargement.

The peripheral oxygen saturation (SPO_2) of 96% and normal arterial blood pressure of 89/52 mmHg were normal. The plain chest radiograph (Figure 1) was not particularly remarkable, whereas the electrocardiogram revealed sinus tachycardia, left axis deviation and left ventricular hypertrophy.

However, Trans-thoracic echocardiogram revealed a tiny patent foramen ovale with a left-to-right shunt, mildly enlarged left atrium with calcification of the entire left atrial wall and the left atrial side of the inter-atrial septum, mitral valve and mitral annular calcification but without mitral regurgitation or stenosis. The aortic valve was thickened, calcified and bicuspid and there were moderate aortic regurgitation. [22] There was concentric left ventricular hypertrophy, but no coarctation of the aorta. There was good bi-ventricular function and normal pulmonary arterial blood pressures. Details of the measured echocardiographic indices are shown in Table I. The anthropometric z-scores were computed using the Ped (z) Pediatric Calculator©. [21]

Figure 1: Plain chest radiograph that revealed the cardiac calcifications.

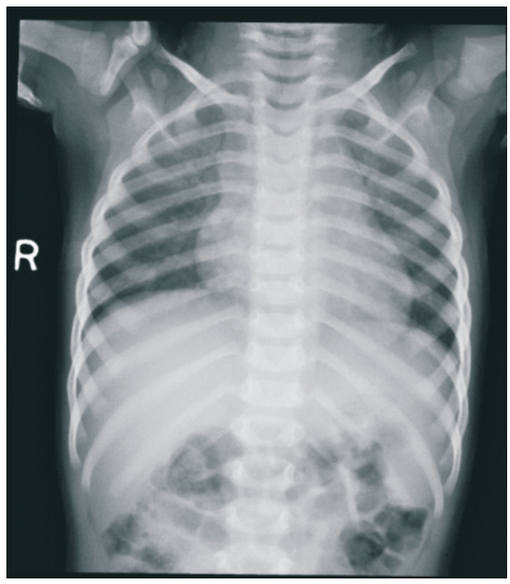


Table I: Trans-thoracic 2-D and M-Mode echocardiography measurements.

Parameters	*Size*	*Z-Score*[21]
Inter-ventricular septal in Diastole (IVSd)		9.41mm 6.92z
Left Ventricular Internal Diameter in Diastole (LVIDd)	20.8mm	-2.19z
Left Ventricular Posterior Wall in Diastole (LVPWd)	6.72mm	3.58z
Inter-ventricular septal in Systole (IVSs)	9.63mm	2.27
Left Ventricular Internal Diameter in Systole (LVIDs)	9.86mm	-3.19z
IVS/LVPW (MM)	1.40	N
Left Ventricular Mass (cubed) LVM	35.0g	1.12z
End Systolic Volume [M-Mode Teich] (ESV)	1.98mL	
End Diastolic Volume [M-Mode Teich] (EDV)	14.1mL	
Fractional Shortening [M-Mode Teich] (FS)	42.0%	N
Ejection Fraction [M-Mode Teich] (EF)	75.4%	N
Pulmonary Valve Annulus	10.5mm	-0.89z
Left Ventricular Mass Index	82g/m^2	N
Main Pulmonary Artery	9.3mm	-1.44z
Right Pulmonary Artery	8.6mm	1.14z
Left Pulmonary Artery	8.0mm	1.39z
Aortic Isthmus	7.75mm	0.11z
Aortic Valve Annulus	9.18	-0.91z
Sino-tubular junction	13.6	1.25z
Ascending Aorta	14.0	1.32z
Left Atrial Diameter	18.8	N

Figures 2 and 3 show still images of left long parasternal axis echocardiogram. These images revealed complete left atrial calcification, to include the atrial septum and the aortic and mitral valves. Figure 4 is a subcostal image showing the left ventricular hypertrophy as well as left atrial and aortic calcifications.

Figure 2: Trans-thoracic left parasternal long axis echocardiogram with colour Doppler.

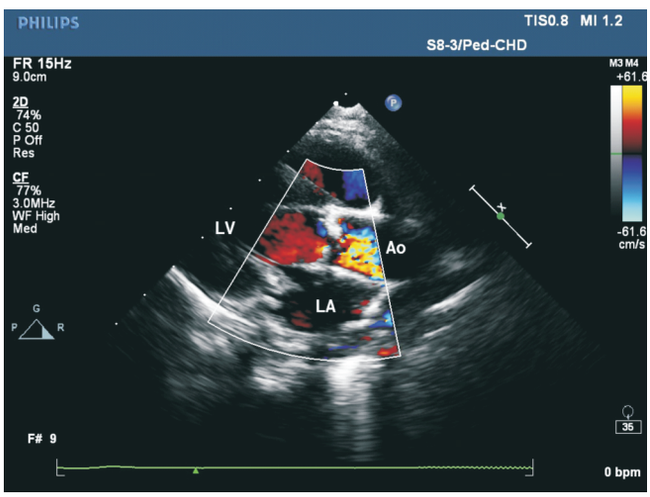
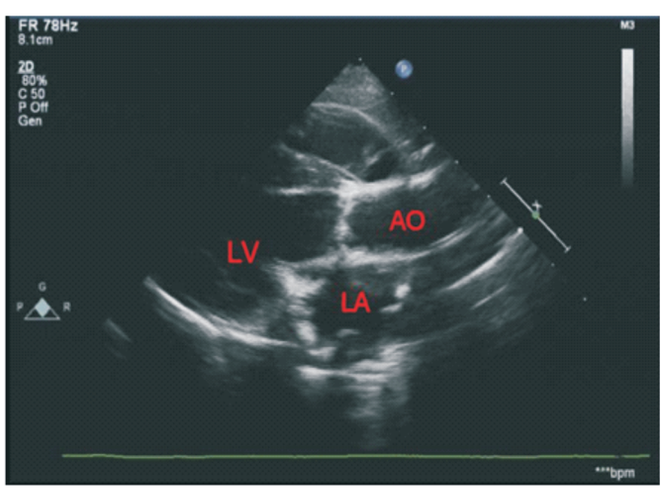


Figure 3: Trans-thoracic left parasternal long axis B-mode echocardiogram.



The abdomino-pelvic ultrasonogram, complete blood cell counts and differential counts, serum calcium, magnesium, phosphate and other serum electrolytes and urea, renal function and parathyroid tests were all essentially normal. Iron studies and CT scan were yet to be conducted. Upon counselling of his parents about our initial findings, written informed consent for the use of the information in scientific publication was obtained. However, the boy has since not been re-presented for follow-up care and attempts to reach the parents over the telephone proved abortive.

Discussion

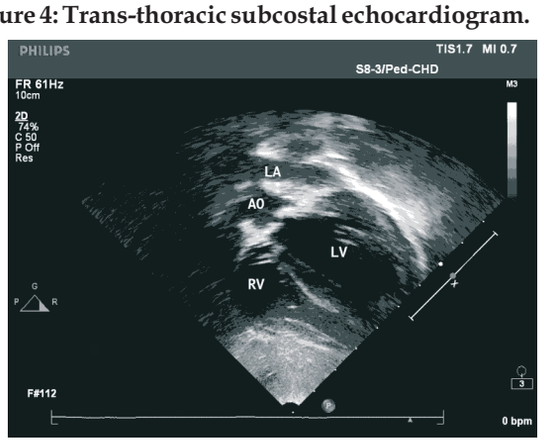
Cardiac, intra-cardiac and pericardial calcifications are well described in the literature. [23] The calcifications are, in most instances, pathologic. [23] Left atrial (LA) calcifications are among the less commonly encountered subset of cardiac calcifications. [23]

LA calcifications were originally described in 1898. [2, 9] Other cases as reported by Oppenheimer in 1912, MacCallum in 1924 and various others across the globe, were all complications of rheumatic heart disease. [23, 24] Most of these cases previously reported were among middle-aged and elderly and females. The index case was a preschool male child who had calcified bicuspid aortic valves.

LA calcifications usually involve the left atrial appendage, left atrial free wall, mitral valve apparatus or the inter-atrial septum discretely. [2] The term "Porcelain atrium" involves the LA appendage, the free wall of the LA and the mitral valve apparatus, except the inter-atrial septum, whereas "coconut" atrium is more severe with the involvement of all areas of the LA. [7] Septal calcification makes surgical interventions challenging and portends a worse prognosis. [25, 26] The cardiac calcifications are either metastatic calcification or dystrophic calcification. [1]

Metastatic calcification is typically seen in patients with a disturbance of calcium and phosphorus metabolism, often due to renal dysfunction. In the index case, renal and parathyroid functions were normal, suggesting that the calcification in his left atrium was dystrophic rather than metastatic. [4] In addition, the index case had severe dystrophic calcification of the entire LA including the atrial septum, the mitral and aortic valves.

Figure 4: Trans-thoracic subcostal echocardiogram.



Calcification of the inter-atrial septum had been reported as a potential contraindication for mitral valve surgery. [6, 27, 28] The index case had no history of surgical exposure. It has been associated with complicated valvular stenosis, cardiac arrhythmias, cardiac block and abnormal cardiac haemodynamics. [4]

The incidence of normally functioning bicuspid aortic valves is 0.6% to 0.9%. [29, 30] The calcification of bicuspid aortic valves is usually age-related and often seen only after the second decade of life. [29] The index case had a dystrophic cardiac calcification as he had normal levels of serum calcium. [13, 24]

There are fewer reports of massive dystrophic LA calcifications following other diseases besides rheumatic heart disease. These include cases associated with chronic kidney disease, radiotherapy for neuroblastoma, endocarditis and Gaucher's disease. [4, 12, 13, 17] The index case did not have a clinical history nor physical or laboratory features suggestive of any of these conditions. In our instance, we did not have the opportunity for more extensive diagnostic investigations and thus, we did not arrive at a definitive underlying aetiology beyond the bicuspid aortic valve. Nonetheless, we have succinctly excluded all the previously documented possibilities.

Conclusions

Following extensive review of the literature, we conclude that such a case has not been previously reported. Therefore, this case is worthy of awareness creation among physicians and other health care staff who attend to young children. To the best of the authors' knowledge, this is the first report that identifies the complete dystrophic left atrial calcification associated with bicuspid aortic valve in a preschool aged patient.

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