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Case Report

Computed tomography imaging characteristics of shone syndrome

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ABSTRACT

Shone syndrome was first described in 1963 by Dr JD Shone. It is a constellation of congenital abnormalities compromising approximately 0.6% of all cases of congenital cardiac abnormalities. Shone syndrome is also known as Shone complex, involving several characteristic cardiac abnormalities: coarctation of the aorta, subaortic stenosis, supralvalvular mitral ring, and a parachute mitral valve. Given the uncommon nature of the disease, we present this case to illustrate potential postsurgical appearances of Shone syndrome, specifically on computed tomography imaging.

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Introduction

We present a case of Shone syndrome after multiple cardiothoracic surgeries. Shone syndrome was first described in 1963 by Dr JD Shone. It is a constellation of congenital abnormalities compromising approximately 0.6% of all cases of congenital cardiac abnormalities. Shone syndrome is also known as the Shone complex, involving several characteristic cardiac abnormalities: coarctation of the aorta, subaortic stenosis, supralvalvular mitral ring, and a parachute mitral valve (PMV).^{1,2} Given the uncommon nature of the disease, we present this case to illustrate potential postsurgical appearances of Shone syndrome, specifically on computed tomography (CT) imaging. This report will focus on imaging after sur-

gical intervention to illustrate the subtle details encountered in radiologic practice.

Case report

A 24-year-old female presented to our facility after referral from an outside facility for work-up of her chronic cardiac problems. Further history from the patient reports that she was born with an unknown congenital heart defect. Throughout her childhood, she has suffered from repeated episodes of respiratory infection, fatigue, peripheral edema, and shortness of breath eventually culminating in her recent diagnosis of congestive heart failure. She states that she had multiple

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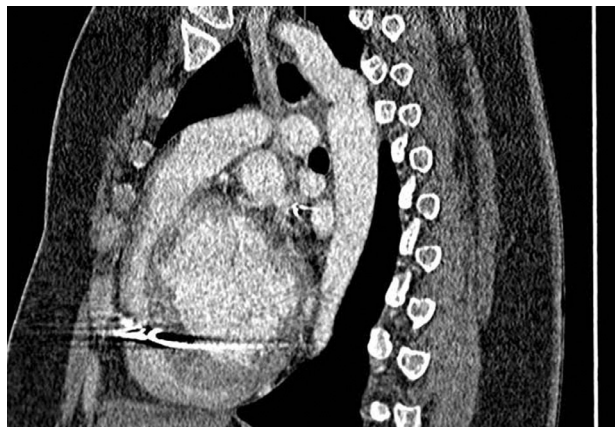


Fig. 1 – Sagittal reconstruction of CTA chest showing repaired aortic coarctation.

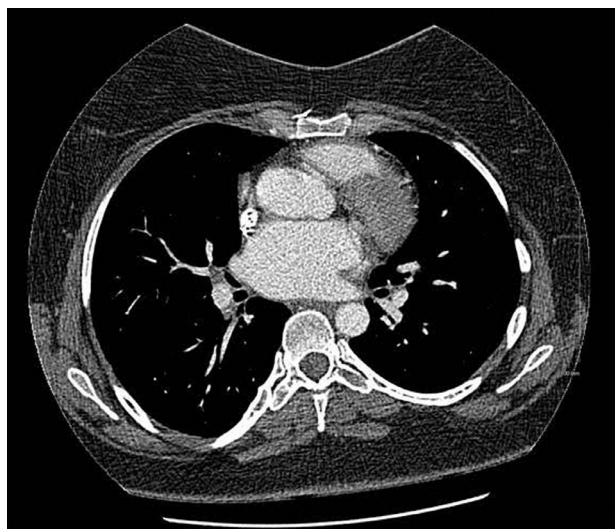


Fig. 2 – Axial image of CTA of the chest of our patient demonstrating subaortic stenosis.

previous open heart surgeries and recently had a pacemaker implantation.

The patient underwent CT angiography of the chest, abdomen, and pelvis at our facility, which showed severe dilatation and hypokinesis of the left ventricle (with an ejection fraction of 29%) and a mildly dilated ascending aorta measuring 4.1 cm. There was abnormality of the contour of the descending aorta at the aortic isthmus, consistent with repaired aortic coarctation. There was also a focal area of luminal narrowing immediately proximal to the aortic valve, indicative of subvalvular aortic stenosis. PMV was suspected but difficult to resolve, possibly beyond the spatial resolution of CT imaging. A membrane-like ridge was seen overlying the mitral valve, consistent with supravulvular ring. There was CT evidence of pulmonary arterial hypertension.

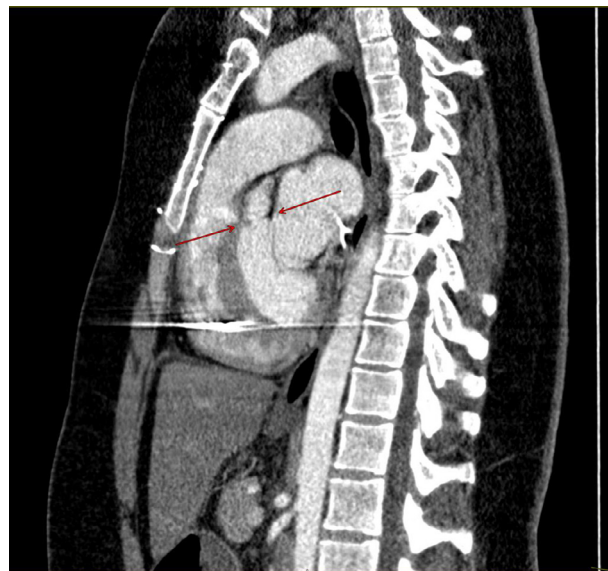


Fig. 3 – Sagittal reconstruction of CTA of the chest of our patient demonstrating subaortic stenosis.

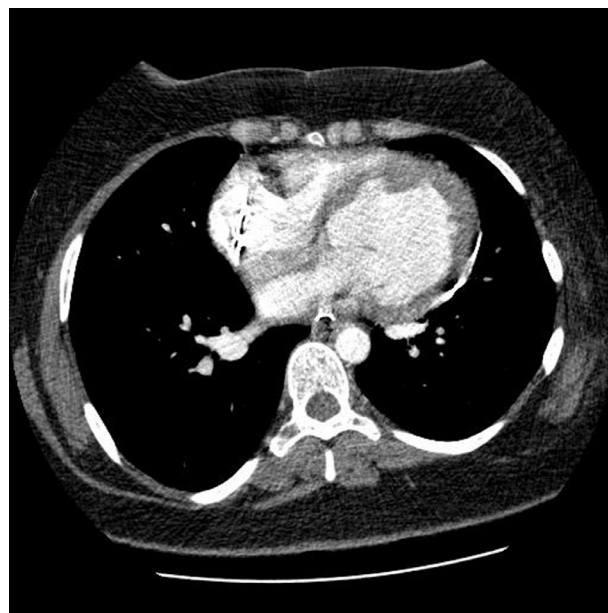


Fig. 4 – Axial image of CTA of the chest of our patient demonstrating possible parachute mitral valve.

Discussion

Shone complex is a congenital heart disease described by Dr JD Shone et al. in 1963. It typically comprises of 4 characteristic lesions of the left heart including PMV, supravulvular mitral ring, subaortic stenosis, and coarctation of aorta.¹ According to a study in 2008 by Popescu et al., Shone disease has an incidence of 0.6% of all cases of congenital heart disease. During early embryogenesis, mitral valve obstruction is the

triggering pathological event. Subsequent underdevelopment of the left ventricle leads to varying degrees of left ventricular outflow tract (LVOT) obstruction and aortic coarctation.² When only 2 or 3 of the abnormalities are present, Shone complex is diagnosed as the incomplete form. Alternatively, there have been reported cases with additional lesions including fused chordae, single papillary muscle, and congenital mitral stenosis.³

Patients can become symptomatic as early as 2 years of age. Typical symptoms include dyspnea, nocturnal cough, tachypnea, poor feeding, failure to thrive, fatigue, and signs and symptoms of heart failure with reduced cardiac output. Patients can also present with recurrent episodes of wheezing and respiratory tract infections due to pulmonary congestion, edema, and exudative pleural effusions.

PMV develops because the chordae-tendinae from both mitral valve leaflets converge onto a single papillary muscle instead of normally diverging onto 2 papillary muscles.¹ The singular attachment of the chordae-tendinae results in a restricted valve opening and subvalvular obstruction and regurgitation.⁴

Supravalvular mitral ring is a membrane-like peripheral ridge arising from the left atrial wall overlying the mitral valve and frequently found attached to the mitral valve. This attachment to the valve may impair the opening of the leaflets causing mitral valve inflow obstruction. The ring may also be large enough to protrude into the mitral valve inflow and cause obstruction. Turbulence can cause a progressive increase in the supravalvular membrane or ridge thickness, worsening mitral inflow obstruction.

Coarctation of the aorta is a congenital narrowing of the aorta. There are 3 types: preductal, ductal, or postductal; named according to their location in reference to the site where the ductus arteriosus inserts.

Valvular and subvalvular aortic stenosis is a narrowing of the aortic valve and the channel below the aortic valve connecting the left ventricle to the aorta. This results in anatomic obstruction of blood across the LVOT.

To date there has been 2 retrospective studies analyzing outcomes of surgical management of patients with Shone syndrome. Dr St. Louis et al. reviewed 28 cases between 1988-2004 from The Children's Medical Center at the Medical College of Georgia, Augusta, Georgia. Dr Malhotra et al. described 43 cases between 1987-2007 from Children's Hospital, Denver, Colorado.

Dr St. Louis et al. reported that of the 28 cases, 2 patients were lost to follow-up. Of 26 remaining patients, 2 died after a second operative intervention. The average follow-up of 24 patients was 6.3 years (range, 1-16 years). Overall survival was 93%. All surviving patients are in class I or II congestive heart failure. They concluded that long-term survival of patients diagnosed with Shone complex is excellent, operative strategies for this complex group should be individualized, and mitral interventions may generally be deferred.⁵

Dr Malhotra et al. reported that of their 43 cases, there was 1 in-hospital death (2.5%) and 6 late deaths (14.2%). 5- and 10-year survival for staged surgical and transplantation was 88% versus 61.3% and 83.1% versus 61.3% ($P = .035$). At a mean follow-up of 7.9 years, freedom from mitral reoperation was 83.3% and freedom from reoperation for subaortic

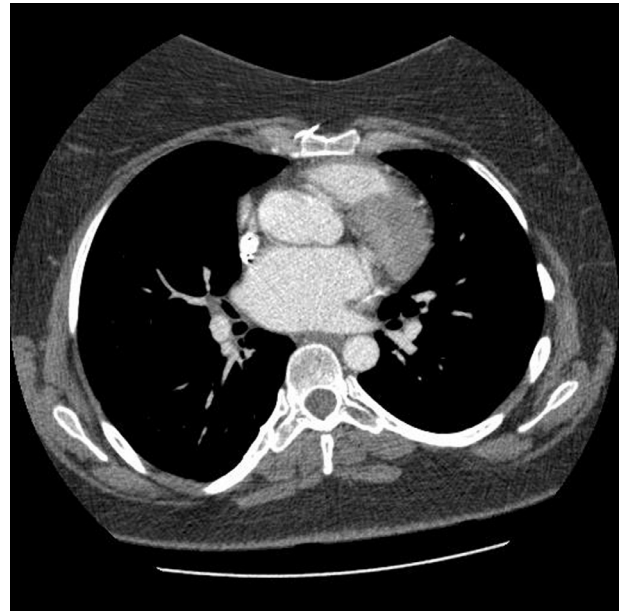


Fig. 5 – Axial image of CTA of the chest of our patient demonstrating supravalvular mitral ring.

stenosis was 78.0%. Wait-list mortality was 13.3% (2 of 13). Wait-list time exceeding 90 days was an incremental risk factor for death after transplantation ($P = .005$). They concluded that despite the challenges of a reparative strategy for Shone complex, favorable survival and durability outcomes can be expected. Heart transplantation, although avoiding the pitfalls of staged repair, confers increased risks from ongoing physiologic derangements due to uncorrected left heart inflow and outflow obstructions during the wait for donor heart availability.⁶

Patients with Shone complex can have a good outcome with early surgical intervention before the onset of pulmonary hypertension. Poor outcome during surgical management is dependent on the degree of involvement of the mitral valve and the presence of secondary pulmonary hypertension.⁷ Surgical management has improved with better understanding of valve pathology and expertise in reparative techniques. Lesions causing obstruction of the LVOT should be addressed first and surgical intervention on the lesions causing inflow obstruction may be postponed. Repair of valves should be done whenever possible, with replacement being reserved for failure of repair.⁸

Conclusion

Shone syndrome is a rare congenital heart disease that is readily recognized by the cardinal left-sided heart defects. Early recognition of this entity through imaging is important in sustaining positive outcomes with early surgical intervention. By reporting this case in the literature we hope to raise additional awareness of Shone syndrome via CT as a potential diagnostic modality (Figs. 1-5).

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