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July 2018

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Recommended Citation

Rasool, F., Khan, M. A., Amanullah, M. (2018). Sinus of valsalva aneurysm rupturing into main pulmonary artery: A rare paediatric cardiac emergency. *Journal of Pakistan Medical Association*, 68(7), 1113-1114.

Available at: https://ecommons.aku.edu/pakistan_fhs_mc_surg_paediatr/38

Sinus of valsalva aneurysm rupturing into main pulmonary artery: A rare paediatric cardiac emergency

Faiz Rasool,¹ Mohammad Asim Khan,² Muneer Amanullah³

Abstract

Congenital Sinus of Valsalva aneurysm is a rare congenital cardiac disease. Most common site of origin is the right sinus. It ruptures into right ventricle or right atrium most of the times. Only in less than 2% of the cases it ruptures into the pulmonary artery. We report a rare case of right sinus of valsalva aneurysm rupturing into pulmonary artery.

Keywords: Ruptured sinus of valsalva aneurysm, Aortopulmonary tunnel, aortic root abscess.

Introduction

Sinus of valsalva aneurysm is a rare disease, accounting for less than 1 % of all congenital cardiac diseases.¹ It becomes clinically apparent when it ruptures. Most of the times it arises from right sinus of valsalva. As aorta occupies a central position, sinus of valsalva aneurysm can rupture into any chamber of the heart, interventricular septum, pericardium and pulmonary artery. While most of the times it ruptures into the right ventricle or right atrium, rupture into the pulmonary artery is rare,² accounting for less than 2 % of all ruptured sinuses of valsalva. We report a rare case of ruptured aortic sinus of valsalva aneurysm into the main pulmonary artery.

Case Report

A 12 years old boy who was referred from an Adult cardiac surgery tertiary care centre with fever and dyspnoea of recent origin and a discharging wound at the left foot. Physical examination revealed a heart rate of 116/minute, BP of 140/50mmHg and temperature of 100°F. A continuous murmur with thrill was heard across the precordium. Examination of the foot revealed exposed medial malleolus with purulent discharge. Provisional diagnosis of infective endocarditis was made. Routine blood investigations were normal. A chest X-ray showed an enlarged cardiac shadow and pulmonary congestion. Blood cultures were negative. C Reactive Protein was

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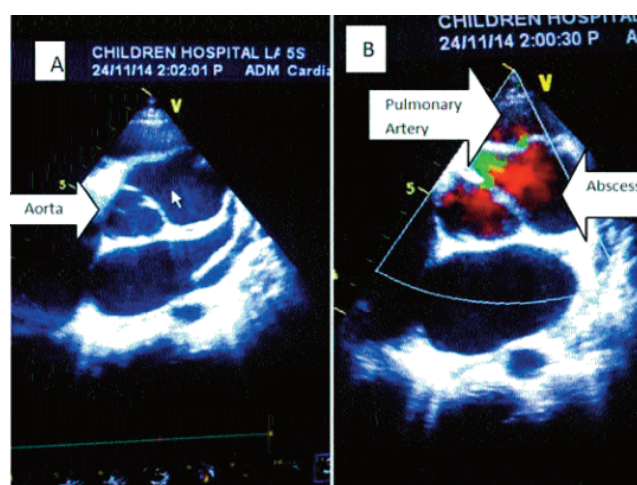


Figure-1: Echocardiogram (A: without contrast, B: with contrast) showing para aortic cavity with communication with aorta and main pulmonary artery.

48mg/dl (normal < 6). Transthoracic Echocardiogram showed a multiloculated para aortic abscess cavity communicating with the MPA. (Figure-1). While a Cardiac CT scan showed the abscess cavity posterior to the MPA with a communication.

An emergent surgery was planned. After a median sternotomy and heparinization, aortobicaaval cannulation was performed and cardiopulmonary bypass established. After cross clamping the ascending aorta, the aorta was transected and antegrade cold blood cardioplegia was infused through the coronary Ostia. No para-aortic abscess was identified. There was a fistulous communication between the aortic right sinus of valsalva and left posterior sinus of pulmonary artery with the dilated part of the fistula between aorta and MPA. Intra-operative diagnosis of a ruptured sinus of valsalva aneurysm into the MPA was made (Figure-2). Both ends of the fistulous openings were closed with patches of glutaraldehyde fixed autologous pericardium. The aorta was closed and Cardiopulmonary bypass was discontinued with ease. Post operative course was uneventful and he was discharged on the 5th post-operative day. At last follow-up (2 months after surgery) he was asymptomatic and his latest echo showed that

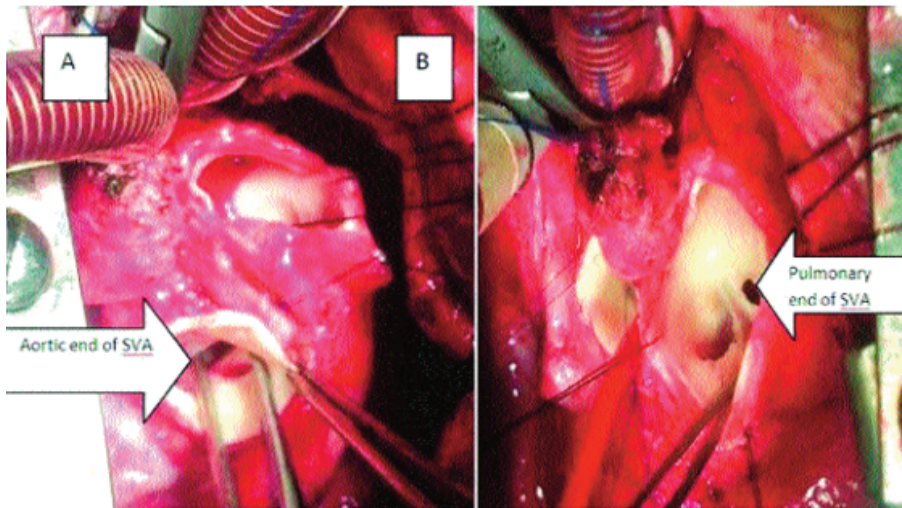


Figure-2: Aortic (A) and pulmonary (B) ends of fistulous tract.

there was no aneurysm, no aortic or pulmonary insufficiency with good biventricular function.

Discussion

Congenital SVA is due to the failure of the fusion between the aortic media and the heart at the level of the fibrous annulus of the aortic valve. The acquired variety of sinus of Valsalva aneurysm is seen in connective tissue disorders like Marfan's syndrome, Behcet's disease or aortic valve endo-carditis and, rarely, chest trauma.

A total of 90-95% of the congenital aneurysm originates in the right or non-coronary sinus and project into the right ventricle or into the right atrium.³ Almost all aneurysms arising in the non-coronary sinus rupture into the right atrium and those arising in the right coronary sinus generally communicate with the right ventricle and occasionally with the right atrium.

The mean age of ruptured congenital sinus of Valsalva aneurysms is 25-40 years. Fewer than 15% of cases occur before the age of 20 years. Our patient presented at the age of 12 years.

Because of its rarity and unusual presentation and the

presence of an infected wound at the medial malleolus, accompanied with fever, we diagnosed it pre-operatively as endocarditis of the aortic valve with an abscess on both echocardiogram and cardiac CT scan.

Heilman⁴ reported the first case of ruptured sinus of Valsalva aneurysm into pulmonary artery in 1985. Since then only 12 cases have been reported. There was only one case report⁵ of paediatric patient that was diagnosed as aorta to pulmonary artery tunnel. We report a case of ruptured sinus of Valsalva aneurysm into main pulmonary artery that occurred in paediatric age group i.e. 12 years.

Although rare but one should keep in mind the possibility of Sinus of Valsalva aneurysm rupturing into pulmonary artery in the setting of a child presenting with machinery murmur.

Disclaimer: None.

Conflict of Interest: None.

Funding Disclosure: None.

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