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Multidedector CT Assessment of Sinus Venosus Type Atrial Septal Defect Combined with Partial Anomalous Pulmonary Venous Return

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Authors' contributions

This work was carried out in collaboration between all authors. Author LP wrote the draft of the manuscript. Author UT designed the figures, managed literature searches and contributed to the correction of the draft. Author BA managed the literature searches. Authors GY, MG and OU provided the case, the figures and supervised the work. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

We present two cases of sinus venosus type atrial septal defect associated with right anomalous pulmonary venous return to the superior vena cava with echocardiography and computed tomography angiography. Sinus venosus type atrial septal defect is a rare anomaly and can be diagnosed with transesophageal or transthoracic echocardiography. However it is often difficult to detect associated pulmonary venous return anomalies and other congenital anomalies with echocardiography. Multidedector CT is a non-invasive technique that may provide useful information on localization and dimensions of ASD and associated anomalies. It is also important to detect these anomalies prior to cardiac and aortic surgical procedures.

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1. INTRODUCTION

Sinus venosus type atrial septal defect (SVASD) is a malformation that was described by Peacok in 1858 and by Waggstaffe in 1868 [1]. Atrial septal defects (ASD) represent 10-15% of congenital cardiac abnormalities. Sinus venosus type of ASD accounts for 4-11% of ASDs and only 1% of all congenital heart abnormalities. Ninety percent of all SVASD cases also have partial anomalous pulmonary venous return (PAPVR) abnormality. Patients with PAPVR are usually asymptomatic and this abnormality is found incidentally [2]. SVASD is a rare congenital defect, so it is of importance to detect this defect and possibly the accompanying PAPVR anomalies prior to cardiac or aortic surgical procedures [3]. SVASD can be diagnosed with echocardiography. However, if accompanied by PAPVR multidetector CT or MRI are useful.

In this article we present two cases with SVASD and PAPVR, together with their transesophageal echocardiography (TEE), transthoracic echocardiography (TTE) and 64-slices-multidedector computed tomography (MDCT) findings.

2. CASE 1

A 37 years old male patient was admitted to hospital with chest pain and effort dyspnea. Blood test results were normal except of high LDL level, which was 126 mg/dL (3.258 mmol/L). TTE and then TEE studies showed mitral valve prolapse, right atrial and ventricular enlargement, enlargement of main pulmonary artery, mild pulmonary valve insufficiency and SVASD with a diameter of 13 millimeters (Figs. 1a and b). An ECG-gated MDCT was performed to detect further abnormalities. MDCT-angiography showed that superior vena cava was draining into the right atrium above the level of SVASD, and also the right superior pulmonary vein was found draining into superior vena cava.

3. CASE 2

A 31 years old male patient was admitted to hospital with chest pain. Both TTE and TEE tests showed no disorders of heart valves and their motions. Also, left ventricular wall thickness, motions and systolic functions were found normal. Additionally, aortic root and main pulmonary artery diameters were normal. An SVASD with a diameter of 16 millimeters was seen at the interatrial septum. The right atrium and ventricle were enlarged and a mild pulmonary arterial hypertension was present. An ECG-gated MSCT was performed to check for further abnormalities. MDCT showed the SVASD and also right superior pulmonary vein draining into superior vena cava (Figs. 2a and b).

4. DISCUSSION

Both of the presented cases have classic symptoms of congenital SVASD and concomitant PAPVR. SVASD causes left-to-right shunt, while PAPVR aggravates the shunt. SVASD diagnosis is important because in ASD patients stroke and ischemic events occur due to paradoxical embolism. Furthermore, ASD patients may develop pulmonary artery hypertension. Both cases had right atrial and ventricular enlargement, and moderate pulmonary artery hypertension, all related to left-to-right shunt. Both SVASD's were diagnosed with echocardiography. However, PAPVR was not shown by echocardiography. MDCT shows the localization and dimensions of ASD, it also shows the drainage of right superior pulmonary vein into vena cava superior. It has also enabled the detection of additional anomalies [4].

Although the exact embryology of sinus venosus defects is controversial, a failed right superior pulmonary vein to overlie interatrial septum has been suggested as the mechanism [5]. Ostium primum and ostium secundum type ASDs are the most common types. The ostium primum type has an incidence of 15% and is characterized by a defect in lower portion of interatrial septum. The ostium secundum ASD has an incidence of 75% and the defect is in the middle of interatrial septum. In the SVASD the defect is in the superior portion of the interatrial septum and it includes 4-11% of all ASD cases [2,3,5].

SVASD is thought to develop in utero, during the pulmonary vein development phase at week 7 [5,6]. Sinus venosus opens into the primitive right atrium of the embryonic heart. During the development, main pulmonary vein is formed from the sinus venosus and then moves toward the left atrium, thus, sinus venosus forms the posterior walls of atria. The posterior wall of main pulmonary artery and right superior pulmonary vein participate in the formation of superior

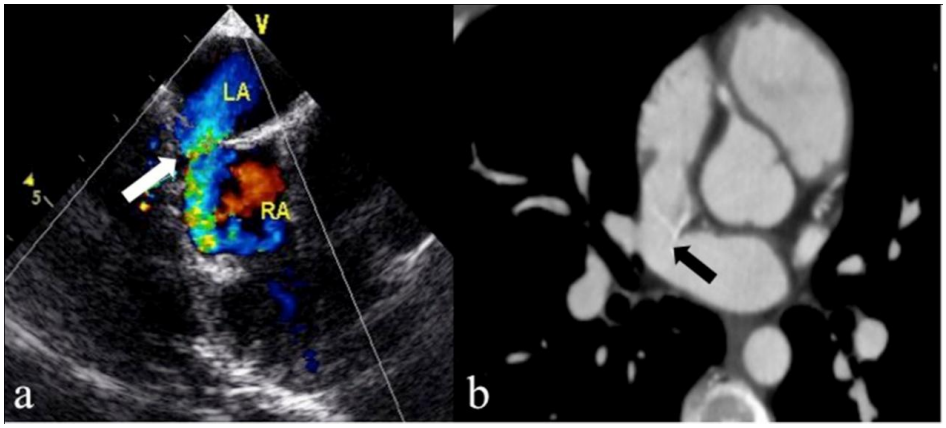

Fig. 1a,b. (a)Transesophageal echocardiography shows the sinus venosus type atrial septal defect and left to right shunt (white arrow)(LA: left atrium, RA: right atrium), (b) Axial CT image demonstrates the sinus venosus type atrial septal defect (black arrow)

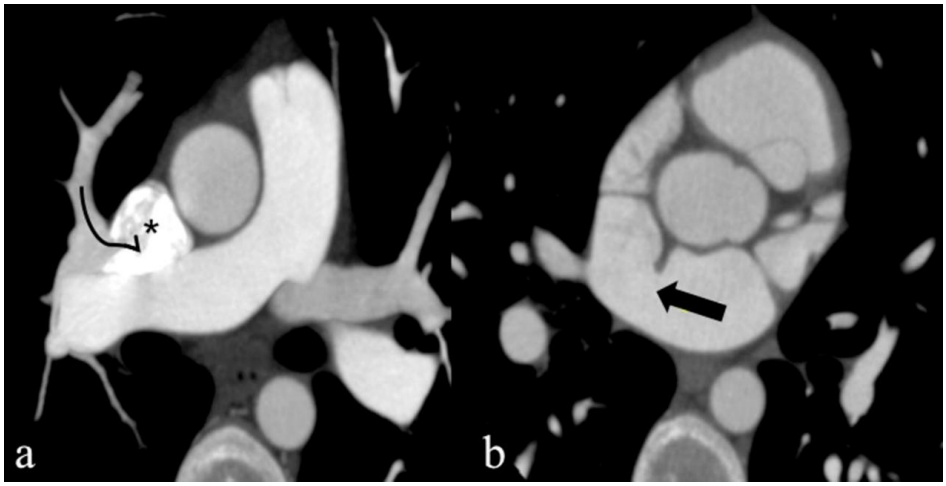

Fig. 2a, b. (a) Right superior pulmonary vein (arrow) drains into the vena cava superior (star). (b) Axial CT image shows the sinus venosus type atrial septal defect (black arrow)

part of the interatrial septum. A defect in this wall leads to a defect in the superior part of the interatrial wall also right superior pulmonary vein drains into the right atrium or vena cava superior [6].

Most of SVASD cases are not diagnosed until fourth or fifth decades of life when pulmonary hypertension or signs of right-to-left shunt develop. Patients with SVASD are prone to atrial dysrhythmias and thromboembolic complications [4,7].

While SVASD has a poor prognosis compared to different types of ASD, drug support and surgical corrections of SVASD in early ages increase the long-term survival rate and decrease heart failure incidence. On the other hand, surgical correction of SVASD is a complex procedure and has risks of thrombosis of superior vena cava or pulmonary vein, residual shunt after closure or sinoatrial nodal dysfunction [2,8]. Both of the cases presented had moderate pulmonary hypertension, therefore surgical treatment was not performed.

SVASD can be easily detected by echocardiography but it is often difficult to detect the pulmonary vein confluence or the combined congenital anomaly by echocardiography and catheter angiography. MDCT is a non-invasive technique that may provide useful information on anatomical abnormalities. Furthermore, it may obviate the need for angiography and be an important supplement to echocardiography [3,9]. Magnetic resonance imaging (MRI) is a reliable

and alternative technique without radiation for establishing the diagnosis, defining pulmonary venous anatomy and quantifying the left to right shunt fraction. MDCT with its high spatial resolution (0.4-0.6 mm), fast acquisition speed and wide volume coverage which enables detection of very small anomalous vessels may overcome on standard MRI sequences [10].

It is important to determine the location of ASD, the size of the defect, hemodynamic effects of the left to right shunt, degree of right heart overload, development of pulmonary hypertension to evaluate SVASD as well as other ASD types [3].

5. CONCLUSION

In conclusion, SVASD can cause pulmonary hypertension and dyspnea in adult patients. The diagnosis of SVASD can be made by echocardiography. MSCT is useful for diagnosing both the SVASD and the associated pulmonary venous anomalies.

CONSENT

All of our three patients have given their informed consents for the case report to be published.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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