ORIGINAL RESEARCH PAPER

Radiodiagnosis

SINUS VENOSUS ATRIAL SEPTAL DEFECT: INCIDENTAL DETECTION IN THE SEVENTH DECADE

KEY WORDS: Congenital heart disease, Sinus venosus ASD, CT pulmonary angiography

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ABSTRACT

Atrial septal defects account for 10 to 15 % of congenital heart disease and are classified into four types. Sinus venosus type accounts for only 1 % of all congenital heart disease. It is present high in the interatrial septum in continuity with the superior vena cava hence difficult to diagnose on routine echocardiography. We report a rare case of a sinus venosus atrial septal defect (ASD) diagnosed on CT pulmonary angiography which was missed on a routine 2D echocardiography in a 64-year old female who presented with symptoms of worsening dyspnoea.

INTRODUCTION

Atrial septal defects are the most common congenital heart disease. They are of four types, of which sinus venosus is least common. ASD is generally diagnosed on trans-esophageal echocardiography (TEE) in patients with findings of enlargement of right ventricle and pulmonary hypertension, on 2D echocardiography or electrocardiography. Advent of cross-sectional imaging like CT scan and MRI scans have made the diagnosis simpler.

CASE REPORT

A 64 year old female with no significant past history presented with breathlessness since 2 months NYHA grade 2 which progressed to NYHA grade 4 over a span of 7 days not associated with chest pain, palpitations or syncopal attack. Routine blood investigations revealed no significant abnormality while ECG showed right ventricular strain pattern. A 2D echocardiogram showed enlargement of right ventricle with raised pulmonary arterial pressure upto 110mm of Hg. D-dimer level was also elevated.

CT pulmonary angiography was performed on a 64 slice MDCT SIEMENS SOMATOM machine to rule out pulmonary embolism as the cause for dyspnea and pulmonary hypertension. The scan revealed dilatation of main pulmonary artery and its branches with no evidence of acute thrombus. The pulmonary veins were also dilated (Figure 2). Foci of mural calcifications were seen in the left pulmonary and right lower lobar arteries with eccentric intraluminal filling defects seen in distal left lower lobar and left upper lobar branches suggesting chronic thrombi. In addition, a focal defect was seen in the interatrial septum adjoining the confluence of superior vena cava with the right atrium (Figures 3 and 4) with an intraluminal filling defect seen in the right atrial appendage suggesting a thrombus (Figure 5).

These findings of a high focal interatrial sinus venosus type of septal defect and thrombus in the atrial appendage were confirmed on trans-esophageal echocardiography performed after the CT study.

After discussion with the cardiology and cardiothoracic surgical teams, the patient opted for conservative management and currently has New York Heart Association class II symptoms while receiving moderate-dose diuretic therapy.

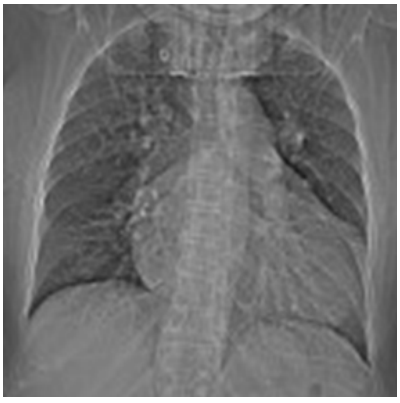

Figure 1: CT Topogram reveals cardiomegaly with prominent hila bilaterally

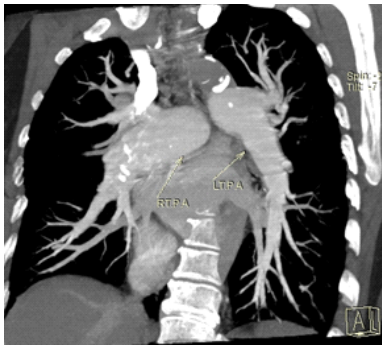

Figure 2: Contrast enhanced coronal images of pulmonary angiography reveal a dilated main pulmonary artery and its branches with no central filling defect

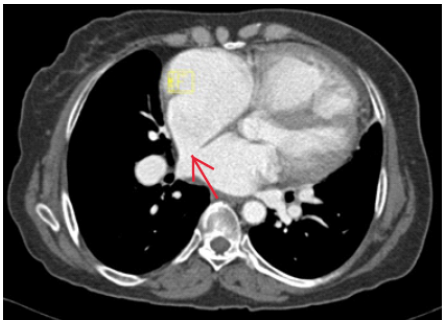

Figure 3: Axial image reveals a defect in the inter atrial septum suggesting an atrial septal defect. Arrow pointing towards the sinus venosus ASD.

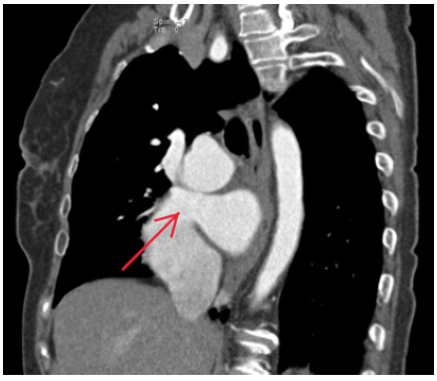

Figure 4: Oblique coronal reconstructed image reveals a defect in the inter atrial septum suggesting an atrial septal defect. Arrow pointing towards the sinus venosus ASD.

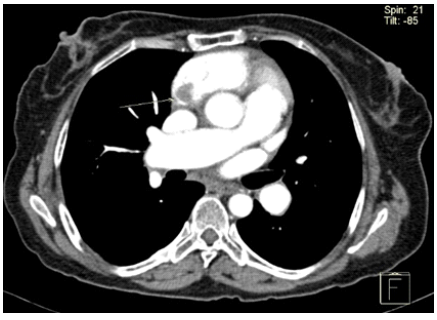

Figure 5: Axial post contrast image reveals a focal filling defect suggesting a thrombus in the right atrial appendage. Arrow points towards the thrombus.

DISCUSSION

The sinus venosus type of ASD is the least common type of ASD encompassing around 4 to 11 percent of all the cases. It was first described in 1858.[9] In atrial septal defect, the malformation of the atrial septum may occur in various positions:

In the lower part–ostium primum, 15% of the cases.

Ostium secundum, in area of fossa ovalis, 75%.

Upper atrial septum, sinus venosus, 10% of the cases.[8]

The exact embryology is controversial, however the typical malformation is an interatrial communication as a result of deficiency of the common wall between the superior vena cava (SVC) and the right-sided pulmonary veins. This is termed as the "unroofing defect". It is commonly associated with an anomalous pulmonary venous connection of some or all of the pulmonary veins, which produces additional left-to-right shunting.[10]

Sinus venosus ASD have nonspecific clinical presentations with age of onset generally before the 4th decade and should always be considered when there is suspicion for right atrial and ventricular enlargement on routine 2D echocardiography.

Transthoracic and trans-esophageal echocardiography, computed tomography and magnetic resonance imaging help in the easy diagnosis of the location of defect and associated vascular abnormality. Due to such imaging facilities, they are usually diagnosed at an early age and amenable for early surgical correction. Our case represents an atypical presentation of significant sinus venosus defect that remained quiescent until over 60 years of age and was comprehensively evaluated using MDCT imaging which was actually done to rule out pulmonary thromboembolism, as patient presented with worsening dyspnea. Very few cases have been described in the literature and one such similar case is cited in the reference.[11]

The malformation often goes unnoticed for decades because symptoms may be absent and signs are subtle. Most children are asymptomatic, though some may present with fatigability and exertional dyspnea. Symptoms usually take 30–40 years to develop. They are the consequences of pulmonary hypertension, atrial arrhythmias and sometimes, associated mitral valve disease.[1] Two-dimensional echocardiography might reveal an ASD with an unusual location raising the suspicion of sinus venosus ASD, but its relationship with nearby structures is not always delineated so in such cases there is need for transesophageal echocardiography.[2] However, the invasiveness and long procedure time are major drawbacks of transesophageal echocardiography. The best view on TEE for recognition of ASD sinus venosus (SVC type) is a bicaval view.4 The echocardiography can establish the size and location of the ASD, the magnitude and hemodynamic impact of the left–to–right shunt, and the presence and degree of pulmonary hypertension [4,5,6.]

Echocardiography and catheter angiography are the primary cardiac imaging modalities but both have their own limitations. Echocardiography is limited by small field of view, over dependence on operator and an acoustic window. Catheter angiography is limited by overlapping of adjacent vascular structures, difficulty in demonstrating pulmonary and systemic vascular structures simultaneously. Computed tomography and Magnetic resonance imaging have important role in overcoming these limitations.(7)

CT has been used in the morphological evaluation of congenital heart disease because of its fast image acquisition time and capacity to obtain volume data. Imaging of heart with CT and MRI depends on technical developments because high spatial and temporal resolution is important for evaluation of congenital heart disease. Multidetector CT scan with its ability for thin reconstructions and multiplanar reconstruction fulfils all the requirements.(7)

CONCLUSION

A rare case of sinus venosus ASD in a female in her seventh decade is presented, which was missed on basic diagnostic investigation like 2D echocardiogram but detected on CT pulmonary angiography, done to rule out chronic pulmonary embolism.

REFERENCES

1. Galie N, Torbicki A, Barst R, et al. Guidelines on diagnosis and treatment of pulmonary arterial hypertension. The Task Force on Diagnosis and Treatment of Pulmonary Arterial Hypertension of the European Society of Cardiology. Eur Heart J. 2004 Dec. 25(24):2243-78.
2. Chen CA, Wang JK, Hsu JY, Hsu HH, Chen SJ, Wu MH. Diagnosis of inferior sinus venosus atrial septal defects using transthoracic three-dimensional echocardiography. J Am Soc Echocardiogr. 2010 Apr. 23(4):457.e4-6.
3. Kivisto S, Hanninen H, Holmstrom M. Partial anomalous pulmonary venous return and atrial septal defect in adult patients detected with 128-slice multidetector computed tomography. J Cardiothorac Surg 2011;6:126.
4. Valdes–Cruz LM, Cayre RO. Echocardiographic diagnosis of congenital heart disease. Philadelphia: 1998.
5. Narula T, Ali MJ, Lopez FA. Clinical case of the month. A 54–year–old man with shortness of breath and irregular pulse. Sinus venosus atrial septal defect . J La State Med Soc. 2010;162(3):129–133.
6. Huang X, Shen J, Huang Y. En face view of atrial septal defect by two–dimensional transthoracic echocardiography: comparison to real–time three–dimensional transesophageal echocardiography . J Am Soc Echocardiography. 2010;23(7):714–721.
7. Roberts WT, Bax JJ, Davies LC. Cardiac CT and CT coronary angiography: technology and application. Heart 2008;94(6):781–792.
8. Miller SW. Cardiac Imaging: the Requisites, 2nd ed. Elsevier Mosby 2005: 36-38;

324-325
9. Jost, Christine Helena Attenhofer et al. "Sinus venosus atrial septal defect: long-term postoperative outcome for 115 patients." Circulation 112 13 (2005): 1953-8.
10. Van Praagh S et al. Sinus venosus defects: unroofing of the right pulmonary veins – anatomic and echocardiographic findings and surgical treatment. Am Heart Journal 1994; 128: 365-79.
11. Never too old for congenital heart disease: sinus venosus atrial septal defect with anomalous pulmonary venous return in an octogenarian Ravi K. Sharma,1 Brian A. Houston,1 Joa˜o A. C. Lima,1 Duke E. Cameron,2 Ryan J. Tedford.