Case Report

Inferior Sinus Venosus Atrial Septal Defect

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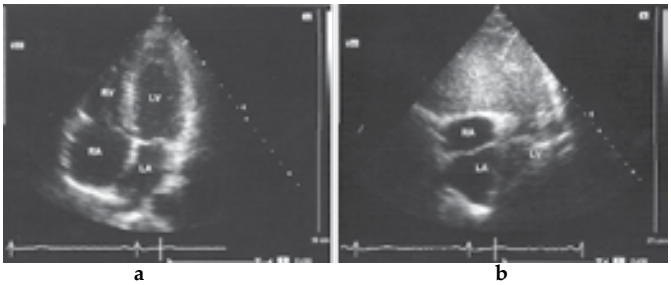
Abstract

One case of inferior vena cava type sinus venosus defect is reported. It is likely to be missed on transthoracic echocardiography. Transesophageal echocardiography is useful. Unlike superior sinus venosus defect which are easily visible on transesophageal echocardiography, a careful search is needed to detect inferior sinus venosus defects. All patients with unexplained right ventricular volume overload on transthoracic echocardiography should have transesophageal echocardiography to see for sinus venosus atrial septal defects.

Case Report

A 26 yrs. asymptomatic male was refered for routine echocardiographic evaluation. Clinical examination and skiagram of chest were normal. Electrocardiogram showed QRS axis around +90° with rsr, pattern in V_1. Transthoracic echocardiography revealed dilatation of RA and RV (Figure 1a). Pulmonary flow acceleration time was 111m.sec. RV systolic pressure was 33.3 mmHg by TR jet. RV systolic functions were normal. RVOT end diastolic dimension was 27.0 mm and end systolic dimension was 10mm. Tricuspid annulus systolic excursion and slope were normal. Lateral tricuspid annulus systolic excursion was 25.6mm. Medial tricuspid systolic excursion was 19.4mm. Tissue doppler imaging of medial and lateral tricuspid annulus were normal. Medial tricuspid annulus –Sa-12.2cm/sec., Ea-22.7cm/sec., Aa-9.7cm/sec. Lateral tricuspid annulus-Sa-15.7cm/sec., Ea-22.7cm/sec., Aa-19.36cm/sec. Inferior vena cava was dilated (expiratory dimension 20.0mm) with diminished inspiratory collapse (inspiratory dimension 12.5mm). Hepatic vein was dilated (16mm) with increased atrial reversal velocity (50cm/sec.). No ASD could be seen from subcostal imaging (Figure 1b). Three pulmonary veins were clearly draining in LA. There was no vertical vein connecting to left innominate vein. Coronary sinus was normal. Cause of RV volume overload was not clear. Transesophageal echocardiography revealed normal SVC-RA junction (Figure 2). There was no defect in this region and region of fossa ovalis. Careful search in the region of junction of IVC with RA revealed an atrial septal defect with left to right shunt (Figure 3). Pulmonary veins were draining in LA. Pulmonary venous flow pattern was normal. Subsequent review from subcostal region with tilting of transducer to focus on the most inferior part of interatrial septum close to IVC confirmed the defect (Figure 4).



a b

Fig. 1 : (a) Apical four chamber view showing dilatation of RA and RV. (b) Showing failure of classical subcostal view to detect inferior sinus venosus defect

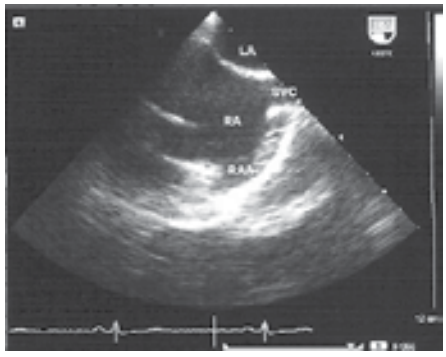


Fig. 2 : Transesophageal echocardiogram showing absence of superior sinus venosus defect

Discussion

Dilatation of RA and RV were clear on transthoracic echocardiography. Cause of volume load of RV was, however, not clear. There was no evidence of pulmonary artery hypertension or pulmonary stenosis. RV systolic functions were normal on 2-D parameters, tricuspid annulus systolic excursion and tissue doppler imaging. Careful evaluation during transesophageal echocardiography revealed inferior vena cava type sinus venosus defect.

Inferior vena cava type of sinus venosus defects are very rare[1,2] and are difficult to image even from trans esophageal echocardiography. Unexplained volume overload of right sided chambers should prompt for careful evaluation for sinus venosus type of ASDs (superior or inferior), coronary sinus type of ASD (absence of roof of coronary sinus) and partial anomalous pulmonary venous drainage. Inferior sinus venosus defects can result in right to left shunt with some degree of systemic oxygen desaturation.[3] Sinus venosus type defects are not managed by device closure and need surgical repair.[4]

Fig. 3 : Showing left to right shunt across inferior sinus venosus defect

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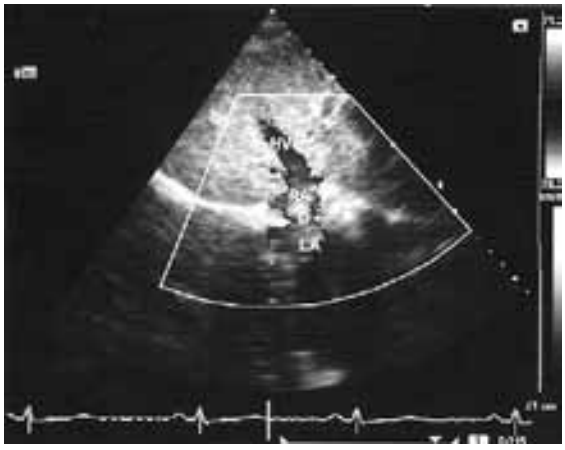

Fig. 4 : Showing inferior sinus venosus defect in tilted subcostal view

Conclusion

Inferior sinus venosus defects are rare. It should be looked for in any patient with unexplained volume overload of right sided cardiac chambers. Modified subcostal imaging and transesophageal imaging are useful in detection and evaluation. Device closure is not possible at present.

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