An Informative Case of Superior Sinus-Venosus Atrial Septal Defect Complicated by Partial Anomalous Pulmonary Venous Connection Successfully Detected by Transthoracic Echocardiography Following Computed Tomography Guidance

Yui Omomo[1], Yasuhide Mochizuki[1], Yukiko Endo[1], Nobuo Oyama[1], Takanari Fujii[1], Hideshi Tomita[1], and Toshiro Shinke[1]

[1]Showa University

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Abstract

We examined a 26-year-old female with a dilated right heart without significant valvular disease. The first transthoracic echocardiography (TTE) did not identify any intracardiac left to right shunt diseases. A contrast-enhanced computed tomography scan showed a sinus-venous atrial septal defect (ASD) and partial anomalous pulmonary venous connection. A superior sinus-venous ASD at the ceiling of atrial septum and two anomalous pulmonary veins connected to the superior vena cava and to the right atrium, respectively, were visualized on the second TTE. Three-dimensional anatomical understanding from multiple imaging modalities may occasionally necessary to delineate rare congenital heart diseases by echocardiography, particularly in adult with a limited echo window.

INTRODUCTION

Sinus-venous atrial septal defect (ASD) is an uncommon type of ASD, accounting for 5-10% of ASD cases [1]. Partial anomalous pulmonary venous connection (PAPVC) is also a rare congenital anomaly that occurs in 0.4-0.7% of the population, as reported by histopathological specimens, and can lead to a volume overload to the right heart as some pulmonary veins return to the venous system. Approximately 10% of ASDs are associated with PAPVC [1, 2]. According to some reports, superior sinus-venosus ASD is difficult to visualize using transthoracic echocardiography (TTE), including in combination with other cardiac imaging modalities[3-5]. There have been few reports about the technical method for delineating superior type of sinus-venosus ASD in TTE. In this case, it was possible to show sinus-venosus ASD complicated by PAPVC clearly. Consequently, we believe, their anatomical characteristics and how it was drawn by TTE is very informative.

CASE

A 26-year-old asymptomatic female was referred to our hospital for a further evaluation of an abnormal shadow on chest X-ray. The first TTE showed the dilated right atrium and ventricle without significant valvular disease, while intraventricular septum was displaced to the left ventricle predominantly in diastole (Figure 1 A - B, Movie Clip S1). The estimated pulmonary to systemic flow ratio (Qp/Qs) was 2.4, suggesting a significant increase in pulmonary blood flow. These results strongly suggested right heart volume overload disease, especially of the pre-tricuspid left to right shunt disease. However, the first TTE could not reveal none of such diseases.

Contrast-enhanced computed tomography (CT) showed a superior sinus-venosus ASD complicated by PAPVC where the upper right and lower pulmonary veins were connected to the superior vena cava (SVC), and to the right atrium (Figure 2 A - D). A right heart catheterization with selective angiography of the right pulmonary artery also showed anomalous connection of the right pulmonary veins to the right heart. Oxygen saturation stepped up in the right upper atrium and Qp/Qs was 2.8. We performed a second TTE with a more complete understanding of the anatomical relationship identified by the CT, transesophageal echocardiography (TEE), and angiography. An atrial septum defect, with a diameter of approximately 7 mm, was observed around the ceiling of the atrial septum. When the probe was gently tilted to the left heart system from the right ventricular inflow view projected at the apical long-axis view (Figure 3 A - C, Movie Clip S2 A - B), we showed the upper and lower right pulmonary veins connected separately to the SVC and to the right atrium (RA), respectively. She successfully underwent surgical intracardiac repair. Intraoperative findings revealed that the upper right pulmonary vein returned to the vicinity of the junction between the SVC and the right atrium. The atrial septal defect was enlarged, while rerouting of the anomalous pulmonary veins to the left atrium was performed with an autologous pericardial baffling.

DISCUSSION and TEACHING POINT

Sinus-venosus ASD complicated by PAPVC is rare, anatomically complex, and difficult to detect by means of TTE alone, particularly in adults with limited echo window. As ASD exists at the ceiling of the atrial septum near the SVC, it is occasionally difficult to evaluate the superior defect type completely. In a report [3] verifying the diagnostic accuracy of TTE and TEE in 41 patients with ASD, 33 cases of them could be identified by TTE, whereas TEE could detect all cases. Meanwhile 6 cases of the 8 ASDs that TTE could not detect were sinus-venosus ASDs, which indicates how difficult it is to identify sinus-venosus ASD with TTE alone. Furthermore, in 7 of these 8 patients with PAPVC coexisting with ASD, PAPVCs were visualized by TEE, but TTE could not visualize the PAPVCs for all. Turner et al reported a case in which the etiology could not be clarified, while dilatation of the right heart became a clue for the final diagnosis as in our case. They reported that re-examination of TTE performed even after CT could visualize neither sinus-venosus ASD nor PAPVC; however, CT scanning clearly showed a sinus-venosus ASD complicated by PAPVC[4]. Inferior sinus-venosus ASD is rarer than the superior type, but technical points have been reported to improve the diagnostic accuracy in TTE. The complete lack of posterior rim in the modified parasternal short axis view is considered to be an effective finding that can distinguish inferior sinus-venosus ASD from ostium secundum type and other ASDs[6].

In this case report, we identified a sinus-venosus ASD complicated by two separately connected anomalous pulmonary veins at the modified right ventricular inflow view projected at the apical long-axis view. Sinus-venosus ASD may also be visualized in the bicaval view in the right thoracic or subcostal approach. Prior study however has indicated that TTE even if with such approach was able to detect only 44% of sinus-venosus ASD [7]. In this case, it was impossible because the patient was obese.

CONCLUSION

We could identify not only the superior sinus-venosus ASD but also PAPVC in a repeated TTE. Echocardiography including anatomical information from other cardiac imaging modalities might be relatively easier to visualize and evaluate more accurately compared to TTE alone, particularly for rare congenital heart disease. It is important to re-evaluate with the help of other modalities in cases that are difficult to evaluate by TTE alone, which will improve our technical skill on TTE screening.

CONFLICT OF INTERESTS

The authors declare no conflict of interest.

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Figure Legends

Figure 1 : Parasternal long-axis view (1-A) and apical 4-chamber view (1-B) of 2-dimensional transthoracic echocardiography during the first examination. Dilatation of the RA and ventricle and intraventricular septum displaced to the left ventricle, predominantly in diastole.

RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle

Figure 2 : Contrast-enhanced CT scan showing PAPVC and sinus-venosus ASD (white arrow). In axial planes, the right upper pulmonary vein (red arrow) connects the SVC (2-A), and the right lower vein (blue arrow) connects the RA (2-B). Three-dimensional CT scan also clearly identifies PAPVC from the back (2-C) and front view (2-D). The translucent vessel shows pulmonary artery.

CT, computed tomography; PAPVC, partial anomalous pulmonary venous connection, sinus-venosus ASD; sinus-venous atrial septal defect; RA, right atrium; RV, right ventricle; LA, left atrium; PA, pulmonary artery; SVC, superior vena cava

Figure 3 : Modified right ventricular inflow view projected at the apical long-axis view of the second transthoracic echocardiography examination. In simultaneous view with color Doppler, Figure 3-A shows superior sinus-venosus ASD (white arrow) and SVC with increased blood flow. The right lower pulmonary vein (yellow arrow) connects to the RA, parallel to PA (3-B). The right upper pulmonary vein (red arrow) connects to the SVC just proximal to the RA (3-C).

RA, right atrium; LA, left atrium; PA, pulmonary artery; SVC, superior vena cava; ASD, atrial septal defect

Electronic Supplementary Material

Movie Clip S1 : Apical 4-chamber view of transthoracic echocardiography showing dilated right-side heart compared to the left-side.

Movie Clip S2 (A, B) : Modified right ventricular inflow view focused on spatial relationship between sinus-venosus ASD, SVC, and anomalous 2 pulmonary veins. Movie clip S2-A clearly detects position of the sinus-venosus ASD, and of the SVC, as well as the right lower pulmonary vein connects to the RA. By adjusting the probe angle to align the axis with the SVC inflow, the spatial relation of the right upper pulmonary vein with the SVC inflow can be drawn (S2-B).

RA, right atrium; SVC, superior vena cava; ASD, atrial septal defect

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Figure\begin{CJK}{UTF8}{gbsn}\end{CJK}\selectlanguage{english}Echocardiography.pptx available at https://authorea.com/users/314897/articles/445260-an-informative-case-of-superior-sinus-venosus-atrial-septal-defect-complicated-by-partial-anomalous-pulmonary-venous-connection-successfully-detected-by-transthoracic-echocardiography-following-computed-tomography-guidance