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Case Report

Cardiac Computed Tomography Angiography Unveils a Concurrent Unroofed Coronary Sinus Atrial Septal Defect with Atrial Septal Defect Secundum

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- Cardiac CTA

Abstract

Unroofed coronary sinus is a rare subtype of atrial septal defect, which is an adult congenital heart disease characterized by communication of the systemic and pulmonary circulations at the atria level. We describe a 25-year-old female with a rare coexistence of a large unrepaired atrial septal defect secundum, and Unroofed coronary sinus type II identified incidentally during a cardiac murmur assessment. Atrial septal defect secundum was diagnosed with transthoracic echocardiography and was confirmed with transesophageal echocardiogram. Unroofed coronary sinus was diagnosed using cardiac computed tomography angiography. The patient underwent a successful reroofing using bovine pericardial patch.

ABBREVIATIONS

UCS: Unroofed Coronary Sinus; PLSVC: Persistent Left Superior Vena Cava; TTE: Transthoracic Echocardiography; TEE: Transesophageal Echocardiography; CCTA: Cardiac Computed Tomography Angiography; ECG: Electrocardiogram; ASD: Atrial Septal Defect

INTRODUCTION

Unroofed coronary sinus (UCS) is a rare subtype of atrial septal defect (ASD), which is an adult congenital heart disease characterized by communication of the systemic and pulmonary circulations at the atria level [1]. Due to partial or complete coronary sinus wall loss, oxygenated blood from the left atrium is carried to the right atrium via the coronary sinus [2].

ASD I is responsible for up to 30% of adult congenital heart disease diagnosis. UCS is the most uncommon type of ASD, usually associated with persistent left superior vena cava (PLSVC). The incidence is estimated to be less than 1% of all atrial septal defects [1]. UCS ASD is anatomically classified by kirklin and Barrat-Boyes into one of four groups, Type I and type II describe completely UCS with or without PLSVC respectively, type II and type IV describe partially unroofed mid or terminal segment respectively [3]. UCS-ASD prevalence and incidence in Saudi Arabia. During childhood and most of adulthood, UCS type ASD are usually asymptomatic. Diagnosis is challenging and it is usually discovered by chance during an examination for another condition, most often prior to coronary bypass surgery [2].

We describe a 25-year-old female with a rare coexistence of a large unrepaired ASD secundum and UCS type II identified incidentally during a cardiac murmur assessment. The electrocardiogram (ECG) revealed sinus rhythm, right axis deviation, and complete right bundle branch block (RBBB). ASD scundum was diagnosed by a transthoracic echocardiography (TTE). Although a transesophageal echocardiography (TEE) confirmed the presence of secundum type ASD, it failed to identify the co-existence of UCS. A big inferiorly situated UCS-ASD type II was diagnosed by cardiac computed tomography angiography (CCTA) that was surgically treated with a bovine pericardial patch.

CASE PRESENTATION

A 25-year-old young female with no history of medical illness had recently developed flu symptoms and had an ejection systolic murmur on auscultation. During her childhood, she had no dyspnea or palpitations. As part of her evaluation, she had a chest X-ray, which revealed cardiomegaly. She then had a TTE, which revealed dilated atria, right ventricle, and an interatrial

shunt. As a result, she was referred to our facility for further evaluation. She had a loud left parasternal systolic murmur at the second intercostal space on auscultation. Her saturation level was 98%. ECG showed a sinus rhythm with right axis deviation and complete RBBB. Repeat TTE at our facility confirmed that her right atrium (RA) and right ventricle (RV) are dilated with RV mid-diameter of 51 mm (Figure 1,2), and at mid interatrial septum (IAS), there is an atrial septal defect (ASD) secondum type measuring ~ 15 mm with QP/QS 3.1 flow from left to right by color Doppler (Figure 3,4) The anterior rim is 8 to 9 mm thick, the posterior rim is 18 mm thick, the distal rim is 17 mm thick, and the proximal rim is 17 mm thick. TEE, showed markedly dilated left atrium (LA). There is a fenestrated defect at the mid IAS with two left-to-right flow jets, the larger of which is 8 mm x 8 mm and the smaller of which is 5 mm x 8 mm, separated by a thin membrane (Figure 5). The total defect diameter was 15 mm x 8 mm, the rim thickness was 19 mm proximally and distally, the anterior rim was 8 mm thick, the posterior rim was 13 mm thick, and the superior and inferior rims were 12 mm and 14 mm thick, respectively. The ASD looks like Swiss cheese at 1 O'clock and a band in the middle, and it measures 8 x 8 mm and 8 x 5 mm, about 15 to 16 mm in diameter. She has significant RV dilatation and RA dilatation, with a Qp/Qs ratio of 3.0.

Patient was taken to the catheterization laboratory for ASD secundum type closure using Amplatzer device under TEE guidance. However, during procedure the multipurpose catheter crossed very inferiorly at the level of the coronary sinus (CS) into the LA. The left upper pulmonary vein identified very easily as well as a small CS vein, which is draining from the great cardiac vein. TEE showed that the multipurpose catheter in the LA, however, it did not go through the previously mentioned secundum ASD, raising the possibility of different co-existing septal defect. Procedure was aborted and patient was scheduled for CCTA to better define the septal defect and rule out the presence of PLSVC. CCTA showed a large ASD inferiorly, representing an UCS. The defect is large, measuring 24 mm in length, and both the superior and inferior vena cava are intact. The pulmonary veins drain in the LA. The right pulmonary veins emerge from a single trunk, while the left pulmonary veins emerge from a very short trunk that splits into two branches early on (Figures 6-9). Patient then referred to cardiac surgery services in our facility. She underwent a successful reroofing using bovine pericardial patch. Echocardiogram post-closure showed no residual interatrial shunt or inferior vena cava obstruction and good ventricular function. She discharged home shortly after.

DISCUSSION

UCS was first described in 1965 by Raghib et al. [4], and is a rare congenital cardiac abnormality characterized by the complete or partial loss of the coronary sinus wall, resulting in an interatrial shunt between the left and right atria [1]. It is often linked with various cardiac abnormalities, the most common of which is PLSVC up to 75% in some series, which drains into the LA if present, causing right to left shunting. Other congenital cardiac malformation correlations have been found, including tetralogy of fallot, tricuspid stenosis, and tricuspid atrisa [5,6].

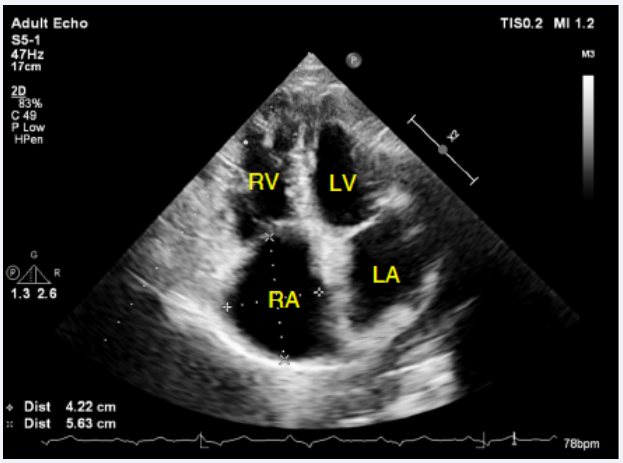

Figure 1 Two-dimensional TTE, apical 4-chamber view, systolic phase, demonstrates a dilated RA. RA, Right Atrium: LA, Left Atrium: RV, Right Ventricle: LV, Left Ventricle.

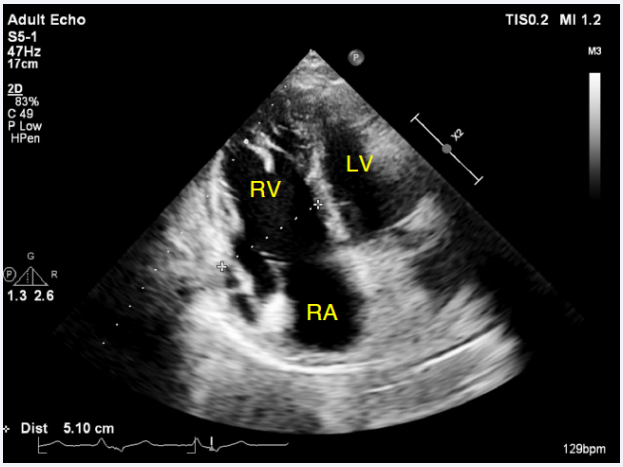

Figure 2 Two-dimensional TTE, apical 4-chamber view, diastolic phase, demonstrates a dilated RV with a mid-segment diameter of 51 mm.

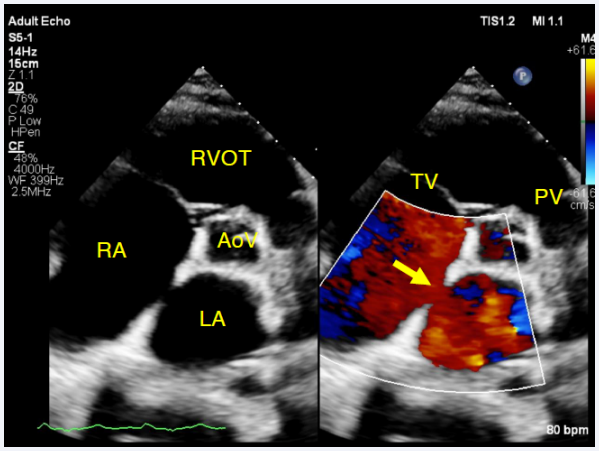

Figure 3 Two-dimensional TTE, parasternal short-axis, aortic valve level, demonstrates a dilated RA, a large ASD secundum type (arrow) with left-to-right flow by color Doppler. RVOT, Right Ventricle Outflow Tract: AoV, Aortic Valve: TV, Tricuspid Valve: PV, Pulmonary Valve.

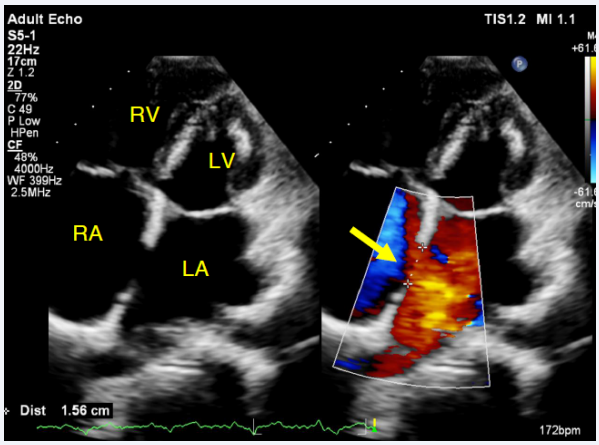

Figure 4 Two-dimensional TTE, apical 4-chamber view with color flow Doppler, systolic phase, demonstrates a large ASD (arrow) with a longitudinal diameter of 15.6 mm.

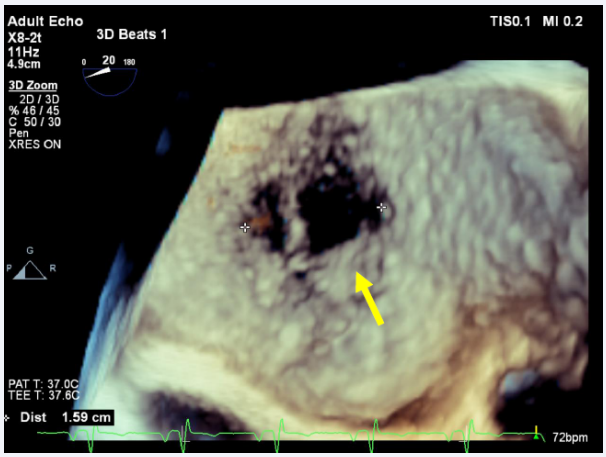

Figure 5 Three-dimensional TEE, Mid-esophageal level, 4-chamber view (20o), demonstrates the large fenestrated ASD Secundum type.

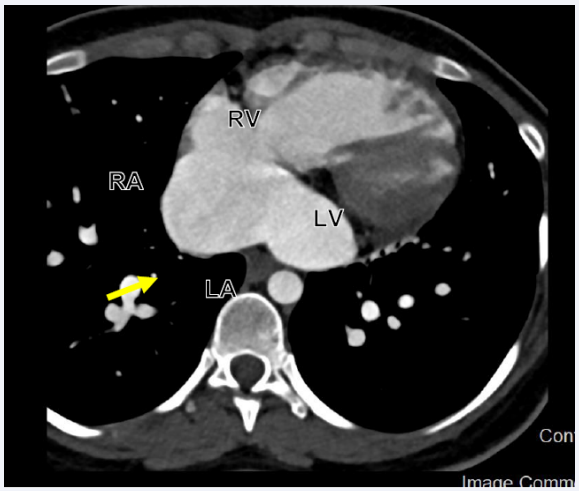

Figure 6 Cardiac computed tomography angiography (CCTA) in axial display showing a communication between the RA and LA representing ASD.

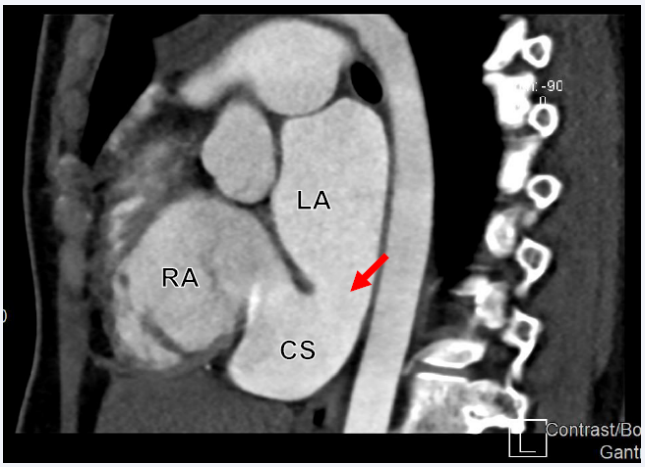

Figure 7 Cardiac computed tomography angiography (CCTA) in sagittal display showing a communication between the RA and LA representing unroofed CS (Arrow). CS, Coronary Sinus.

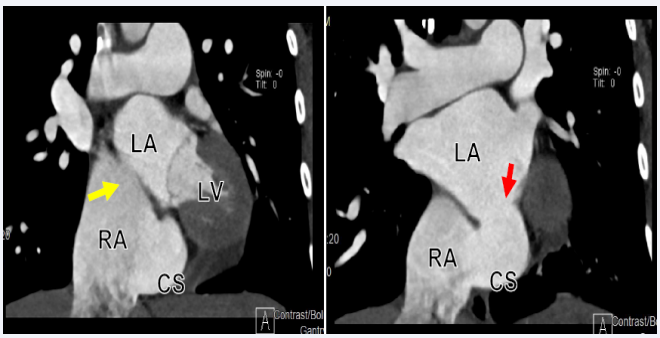

Figure 8 Cardiac computed tomography angiography (CCTA) in sagittal displays showing dilated RA, ASD (Yellow Arrow), and unroofed CS (Red arrow).

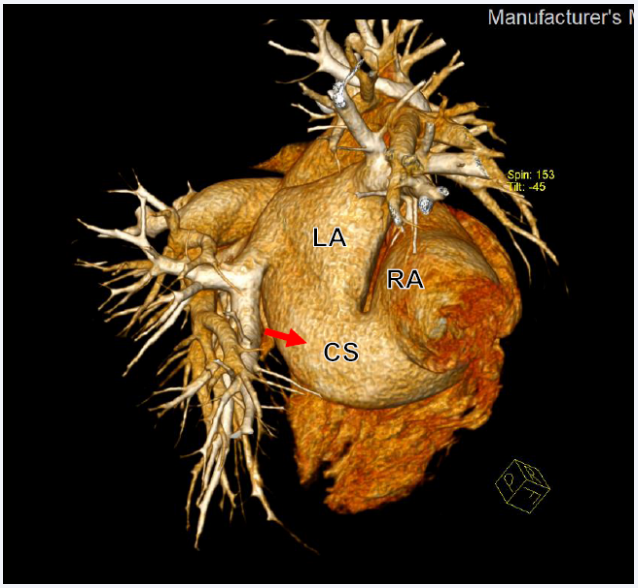

Figure 9 Cardiac computed tomography angiography (CCTA) three-dimensional reconstruction showing the unroofed dilated CS.

Diagnosis of UCS-ASD is challenging since most patients are asymptomatic and, if they do develop symptoms, they are typically non-specific. Evidence of right chambers enlargement and right ventricular failure due to volume overload can occurs with chronic left to right shunt. In contrast, if it is associated with abnormally PLSVC that drains into LA, cyanosis and polycythemia can occur due to right to left shunt in those individuals and hence early detection. TTE can detect an interatrial shunt but is inaccurate in identifying an UCS because it is a deep posterior structure; TEE on the other hand, can accurately identify an UCS in most people. CCTA appears to be superior to both TTE and TEE due to its high spatial and temporal resolutions, multiplanar reconstruction capabilities, and large field of view, CCTA is a powerful tool for defect diagnosis and characterization, as well as the identification of related abnormalities of the heart and pulmonary vasculature [7,8]. The preferred treatment is believed to be surgery. The percutaneous interventions such as covered stenting or implantation of Amplatzer occluding device have shown encouraging outcomes, but further research is required before it can be implemented into routine practice [9,10]. Prognosis after surgical treatment is favorable as showed by Ootaki et al. [3]

CONCLUSION

The extremely rare subtype of the most prevalent adult congenital malformation is UCS-ASD. Although the identification may pose challenges, the utilization of multimodality imaging can effectively facilitate its detection and treatment planning, as exemplified in our patient. Percutaneous interventions such as covered stenting or the implantation of an Amplatzer occluding device hold considerable potential; nevertheless, surgical repair remains the preferred primary treatment option.

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