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OPIS PRZYPADKU
CASE REPORT

From thread to yarn, and yarn to thread: a complex case of persistent left superior vena cava

Od nitki do kłębka, od kłębka do nitki: złożony przypadek przetrwałej lewej żyły głównej górnej

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ABSTRACT

INTRODUCTION: Persistent left superior vena cava (PLSVC) is a rare venous anomaly, occurring in 0.3–0.5% of the general population and up to 4.3% of patients with heart defects. It forms from the junction of the left subclavian and internal jugular veins, passes through the left mediastinum, and drains into the right atrium via the coronary sinus. Usually asymptomatic, it is typically discovered incidentally during imaging and may be associated with an atrial septal defect (ASD).

CASE REPORT: A 52-year-old female patient with persistent atrial fibrillation, a history of ischemic stroke in the left hemisphere of the brain, uncontrolled hypertension, and diagnosed with ASD type 2, was referred for pulmonary vein isolation (PVI) due to symptomatic arrhythmia of European Heart Rhythm Association class IIb and New York Heart Association class II severity. After unsuccessful PVI, pharmacological cardioversion was attempted, followed by electrical cardioversion, which temporarily restored sinus rhythm. Echocardiography revealed moderate tricuspid valve regurgitation and an enlarged coronary sinus. Cardiac computed tomography was ordered, revealing the presence of a PLSVC, into which the left superior pulmonary vein drains, with rightward displacement of the interatrial septum and a patent foramen ovale (PFO). After cardiac surgery consultation, the patient was qualified for defect correction.

CONCLUSIONS: PLSVC may be associated with congenital defects such as ASD type 2/PFO, which is relevant in the treatment of arrhythmias and defect correction. An enlarged coronary sinus on echocardiography should raise suspicion of PLSVC. The presence of PLSVC is significant when placing devices with central venous access.

KEYWORDS

persistent left superior vena cava, atrial septal defect, coronary sinus

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STRESZCZENIE

WPROWADZENIE: Przetrwała lewa żyła główna górna (*persistent left superior vena cava* – PLSVC) jest rzadką anomalią krążenia żylnego, wynikającą z nieprawidłowości w embriogenezie. Występuje u 0,3–0,5% ogólnej populacji oraz nawet u 4,3% pacjentów z wadami serca. Powstaje na skutek połączenia lewej żyły podobojczykowej i żyły szyjnej wewnętrznej, przechodzi przez lewą stronę śródpiersia i zazwyczaj uchodzi do prawego przedsionka przez zatokę wieńcową. Zwykle jest bezobjawowa i często wykrywana przypadkowo podczas badań obrazowych układu krążenia. Może współistnieć z ubytkiem przegrody międzyprzedsionkowej (*atrial septal defect* – ASD).
OPIS PRZYPADKU: 52-letnia pacjentka z utrwalonym migotaniem przedsionków, przebytym udarem niedokrwiennym lewej półkuli mózgu, niekontrolowanym nadciśnieniem tętniczym i rozpoznanym ASD typu 2 została skierowana na izolację żył płucnych (*pulmonary vein isolation* – PVI) z powodu objawowej arytmii o nasileniu IIb według European Heart Rhythm Association i klasy II według New York Heart Association. Po nieudanej PVI podjęto próbę kardiowersji farmakologicznej, a następnie elektrycznej, która jedynie tymczasowo przywróciła rytm zatokowy. Badanie echokardiograficzne wykazało umiarkowaną niedomykalność zastawki trójdzielnej i powiększoną zatokę wieńcową. Zlecono wykonanie tomografii komputerowej serca, ujawniając obecność PLSVC, do której uchodzi lewa żyła płucna górna, z przesunięciem przegrody międzyprzedsionkowej w prawo i drożnym otworem owalnym (*patent foramen ovale* – PFO). Po konsultacji kardiochirurgicznej pacjentka została zakwalifikowana do korekcji wady.
WNIOSKI: PLSVC może towarzyszyć wadom wrodzonym takim jak ASD typu 2/PFO, co ma znaczenie w leczeniu arytmii oraz korekcji wad. Powiększona zatoka wieńcowa w echokardiografii powinna budzić podejrzenie PLSVC. Obecność PLSVC jest istotna przy umieszczaniu urządzeń z centralnym dostępem żylnym, ponieważ może stanowić trudności.

SŁOWA KLUCZOWE

przetrwała lewa żyła główna górna, ubytek przegrody międzyprzedsionkowej, zatoka wieńcowa

INTRODUCTION

Persistent left superior vena cava (PLSVC) is a vascular anomaly in which the vein drains into the right atrium through the coronary sinus at the junction of the left internal jugular and subclavian veins due to abnormal development of the left cardinal vein [1]. It is present in about 0.3–0.5% of the general population and in about 4.3% of patients with heart defects. This congenital anomaly is usually asymptomatic and does not cause any physiological problems. However, it may become a significant problem in several clinical situations. Various complications related to PLSVC are encountered in anesthesia, renal, oncological and cardiovascular procedures. The presence of PLSVC is usually incidentally detected during the placement of a pacemaker (PM), an implantable cardioverter-defibrillator (ICD) and cardiac resynchronization therapy (CRT) leads [2].

CASE REPORT

A 52-year-old woman reported as planned at the end of June 2023 to the 1st Department of Cardiology at the Prof. Leszek Giec Upper Silesian Medical Center of the Medical University of Silesia to restore sinus rhythm by means of pulmonary vein isolation (PVI) ablation and to qualify for the closure of an atrial septal defect type 2 (ASD type 2). The patient was treated chronically for poorly tolerated, persistent atrial fibrillation (AF), treated with anticoagulation using rivaroxaban 1 × 20 mg, diagnosed with ASD type 2 after an ischemic stroke of the left hemisphere of the brain in March 2023, with unregulated arterial hypertension. Additionally, the patient suffers from hypothyroidism – post-strumectomy, depressive disorders and obesity (body mass index – 27.34).

The patient has a family history with cardiovascular disorders. In the clinical picture, the patient had been experiencing heart palpitations with a feeling of arrhythmia of European Heart Rhythm Association (EHRA) class IIb intensity for many years, with no typical symptoms of angina pectoris, reporting non-specific chest pain associated with arrhythmia unrelated to exercise. The patient was assessed as class II on the New York Heart Association (NYHA) scale, 4 points on the CHA2DS2-VASc scale corresponding to the risk of thromboembolic complications, and 2 points on the scale for assessing the risk of bleeding – HAS-BLED scale.

On admission, during the first hospitalization, the state of the patient's cardiovascular and respiratory systems was stable. The resting an electrocardiogram (ECG) showed AF with a ventricular rate of 100/min, and dyselectrolytemia was present in the laboratory tests. The patient underwent transthoracic echocardiography, which demonstrated asynchrony of interventricular septal contraction, normal left ventricular systolic function, left ventricular hypertrophy, severe tricuspid valve regurgitation, mild mitral valve regurgitation, moderate pulmonary valve regurgitation, and a dilated coronary sinus. Transesophageal echocardiography (TEE) confirmed a dilated coronary sinus measuring 32 × 19 mm (Figure 1, 2), an ASD with a permanent left-right shunt measuring 7 × 17 mm (Figure 3), severe/moderate tricuspid valve regurgitation (Figure 4), and mild mitral valve regurgitation.

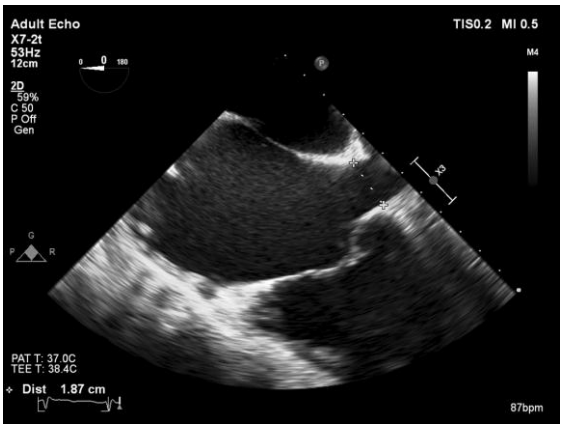

Fig. 1. Transesophageal echocardiography showing enlarged coronary sinus – 32 × 19 mm.

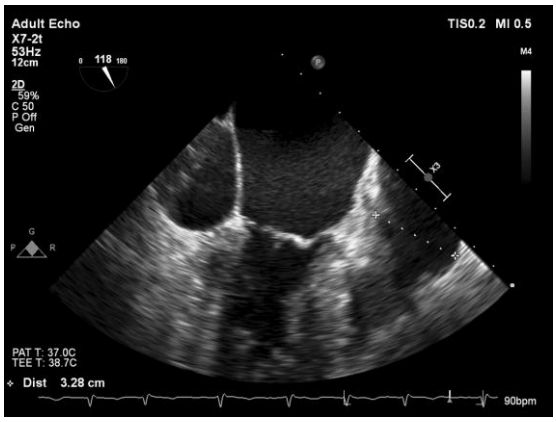

Fig. 2. Transesophageal echocardiography showing enlarged coronary sinus – 32 × 19 mm.

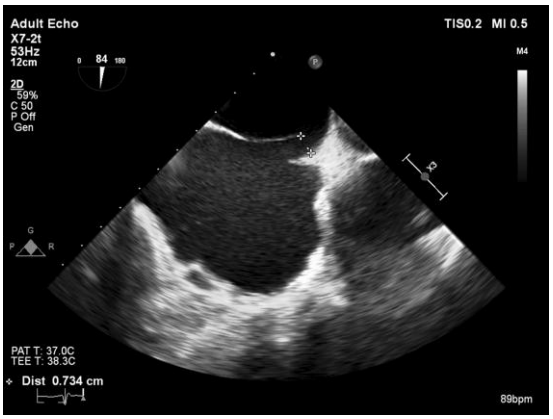

Fig. 3. Transesophageal echocardiography – visible patent foramen ovale canal – 7 mm.

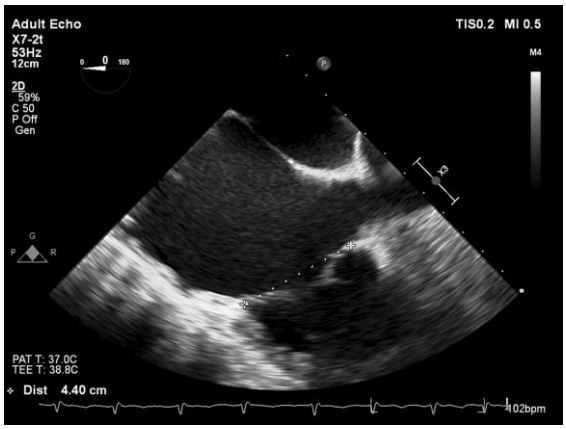

Fig. 4. Transesophageal echocardiography revealing tricuspid ring measuring 44 mm.

On the 27th of June 2023 the patient underwent a planned circumferential ablation of the pulmonary veins, achieving their electrical isolation. Using a transseptal approach, a mapping electrode (Lasso) and an ablation electrode (SmartTouch) were introduced into the left atrium. Mapping was performed using the CARTO 3 electroanatomical system. It showed the presence of the left common pulmonary vein. Then the procedure was extended to include an ablation line on the roof and posterior wall, creating BOX, and an oblique line on the anterior wall from the mitral annulus to the right pulmonary veins. During the procedure, pharmacological cardioversion was performed using 300 mg of antazoline and 600 mg of amiodarone, which was unsuccessful.

Subsequently, electrical cardioversion (CVE) was performed, successfully restoring sinus rhythm, but after 24 hours there was a recurrence of typical atrial flutter with a QRS rate of 65/min, which was well tolerated by the patient. Antiarrhythmic therapy with amiodarone was administered until the patient's admission to the hospital for the performance of CVE.

After the optimization of treatment and scheduling of the percutaneous closure of ASD type 2, the patient was discharged with recommendations. In October 2023, the patient underwent an outpatient CT scan of the heart, which revealed the presence of a congenital heart defect in the form of PLSVC, with draining of the left upper pulmonary vein, with a right shift of the interatrial septum and a visible, wide 1.9 × 0.4 cm, patent foramen ovale (PFO; Figure 5, 6).

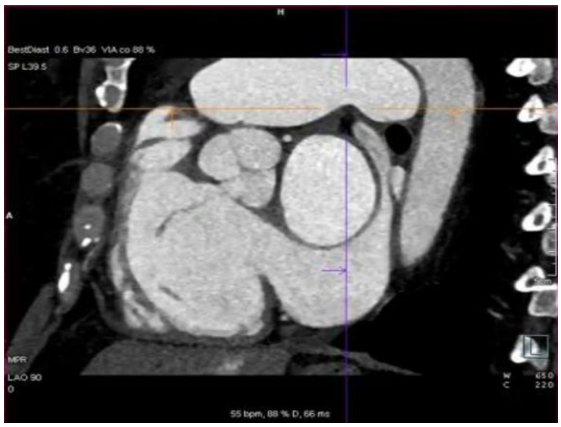

Fig. 5. Cardiac computed tomography showing congenital heart defect in form of PLSVC.

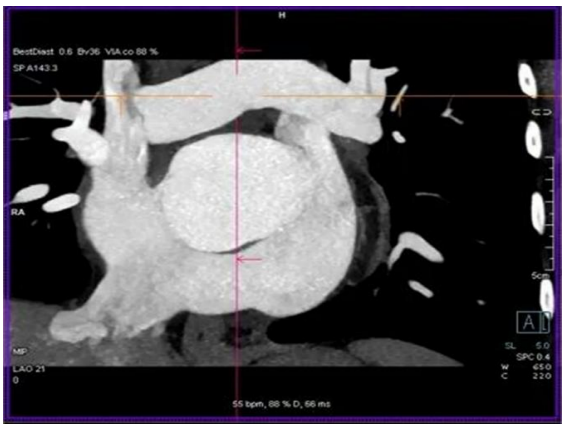

Fig. 6. Cardiac computed tomography showing congenital heart defect in form of PLSVC, into which left superior pulmonary vein drains, with rightward displacement of atrial septum.

During the second hospitalization in November 2023 for the purpose of undergoing CVE, the procedure was abandoned in favor of pharmacological cardioversion with restoration of sinus rhythm, which was performed without complications. After echocardiography, which showed no changes compared to the previous one, the patient underwent cardiac surgery consultation and was qualified for surgical correction of the congenital defect.

DISCUSSION

A comprehensive knowledge of the standard anatomical structures of the major vessels, especially in the thorax, is crucial for any clinician. PLSVC is the most common systemic venous anomaly in the thorax, with a reported prevalence of up to 0.5% in the general population and up to 10% in patients with congenital heart disease (CHD) [3]. It is worth noting that individuals with PLSVC typically do not show symptoms, and as many as 80% of those affected have normal vasculature on the right side [4]. Our patient's case, in which PLSVC was diagnosed at the age of 54, exemplifies such an incidental finding. In cases of PLSVC, commonly associated congenital cardiac disorders include an ASD, similar to the case of our patient, a ventricular septal defect sequenced with coarctation of the aorta, transposition of the great vessels, tetralogy of Fallot, and abnormal pulmonary vein connections [5].

Knowledge of embryology aids in understanding congenital malformations of the superior vena cava (SVC). In the 5th week of fetal life, three pairs of cardinal veins drain the embryo's body: the anterior cardinal veins drain the cephalic portion, and the posterior cardinal veins drain the caudal portion. Both empty into the short paired common cardinal veins, which eventually drain into the sinus venosus. The formation of the vena cava system is marked by the development of anastomoses between the left and right sides, redirecting blood from the left side to the right side. The development of the vena cava and associated veins involves the differentiation and regression of specific embryonic veins to form the mature structures found in the adult cardiovascular system. Specifically, the right anterior and common cardinal veins as well as the right horn of the sinus venosus form the right SVC and part of the azygos vein. On the left side, the anterior cardinal vein contributes to the left superior intercostal vein and the left brachiocephalic vein, while its regression forms the ligament of Marshall, and the left horn of the sinus venosus forms the coronary sinus [6]. The awareness of a PLSVC is crucial for patients undergoing invasive procedures such as CRT, PM implantation, or central venous catheterization as this anatomical variant can complicate these interventions [7]. Therefore, pre-procedural identification and understanding of PLSVC are essential for successfully planning and executing these invasive interventions, ensuring patient safety and optimal outcomes. The awareness of our patient's PLSVC would have enhanced the effectiveness of cardiac ablation by allowing tailored planning according to their unique anatomy. PLSVC diagnosis typically relies on contrast echocardiography, revealing coronary sinus dilation and opacification [8].

In summary, based on the gathered information and the case of our patient, it is essential to consider the diagnosis of PLSVC whenever symptoms such as a dilated coronary sinus or ASD type 2/PFO are present. Early identification of this anomaly allows effective treatment, improves the patient's quality of life, and prevents the late complications that may arise from a missed diagnosis.

CONCLUSIONS

Asymptomatic isolated PLSVC is a benign pathology but it can be associated with other heart defects. The

presence of an enlarged coronary sinus detected during echocardiography should always raise the suspicion of an additional anomaly, such as a PLSVC, as it may contribute to complications of electrophysiology procedures.

This anomaly is often accompanied by other congenital heart defects including a PFO or ASD type 2, which is significant for planning and conducting invasive treatments, which may require cardiac surgery, as in this case.

Authors' contribution

Study design – J. Dołęga, K. Krzywiecka, N. Lekston, K. Mizia-Stec
Manuscript preparation – J. Dołęga, A. Machnik, K. Krzywiecka, N. Lekston
Literature research – J. Dołęga, A. Machnik
Final approval of the version to be published – K. Mizia-Stec

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