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Case Report

Sinus Venosus Interatrial Communication, Anomalous Pulmonary Venous Return, Pulmonary Artery Aneurysm With Left Main Compression: Complex Case With Complex Management

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ABSTRACT

A young female patient was referred for pulmonary arterial hypertension (PAH). Catheterization revealed a large sinus venosus interatrial communication (SVIAC), partial anomalous pulmonary venous return (PAPVR), pulmonary vascular resistance (PVR) 15 Wood units, and bidirectional shunting. She was then put on target medication for PAH. Two years later, she had angina and underwent computed tomography examination, which showed pulmonary arterial aneurysm compressing the left main coronary. Coronary stenting was performed, which successfully relieved the compression and angina. Meanwhile, PVR lowered to 3.5 Wood units after medical therapy. Surgical correction for SVIAC and PAPVR was done successfully 5 years after diagnosis.

RÉSUMÉ

Une jeune patiente a été envoyée en consultation en raison d'hypertension artérielle pulmonaire (HAP). Le cathétérisme a révélé une grande communication interauriculaire (CIA) de type *sinus venosus*, un retour veineux pulmonaire anormal partiel (RVPAP), une résistance vasculaire pulmonaire (RVP) de 15 unités Wood et un *shunt* bidirectionnel. On lui a donc prescrit un traitement ciblé contre l'HAP. Deux ans plus tard, elle a eu une angine et a subi une tomodensitométrie, qui a montré un anévrisme de l'artère pulmonaire compressant l'artère coronaire principale gauche. L'implantation d'une endoprothèse coronarienne a permis de soulager efficacement la compression et l'angine. Entre-temps, la RVP est passée à 3,5 unités Wood après le traitement médical. La correction chirurgicale de la CIA de type *sinus venosus* et du RVPAP a été effectuée avec succès 5 ans après le diagnostic.

Closing a large interatrial communication might worsen right heart failure if pulmonary vascular resistance (PVR) remains high after device or surgical closure. The latest guidelines are strict with respect to setting up the acceptable threshold of pulmonary arterial pressure (PAP) and PVR for closing the interatrial communication with PAH.[1] Herein, we present a case of a young female patient with a large sinus venosus interatrial communication (SVIAC), partial anomalous pulmonary venous return (PAPVR), pulmonary artery aneurysm (PAA) with initial presentation of high mean PAP and PVR, and bidirectional shunting.

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See page 544.e9 for disclosure information.

Case

A 27-year-old woman was referred to our hospital for progressive dyspnea with diagnosis of PAH. The electrocardiogram showed a severe pressure-overload–type right ventricular hypertrophy (Supplemental Fig. S1). The chest radiograph revealed a dilated pulmonary artery and right ventricle (Supplemental Fig. S2). Cardiac catheterization showed a large SVIAC, PAPVR (Fig. 1A), PAH with mean PAP 48 mm Hg, PVR 15 Wood units, and bidirectional shunting (predominantly left to right shunting), and pulmonary to systemic blood flow ratio of 1.7 in November 2013. She was then put on target medications for PAH.

Two years later, she had angina pectoris and underwent multidetector computed tomography (MDCT) examination, which showed a SVIAC (Fig. 1B), PAPVR (Fig. 1B, 1C, Supplemental Fig. S3) and PAA (Supplemental Fig. S4, S5) compressing the LMCA ostium and its proximal shaft (Fig. 2A). Coronary angiogram (Fig. 2B) and intravascular ultrasound (IVUS) (Supplemental Fig. S6) confirmed the diagnosis of extrinsic compression of LMCA ostium and shaft. Surgery for

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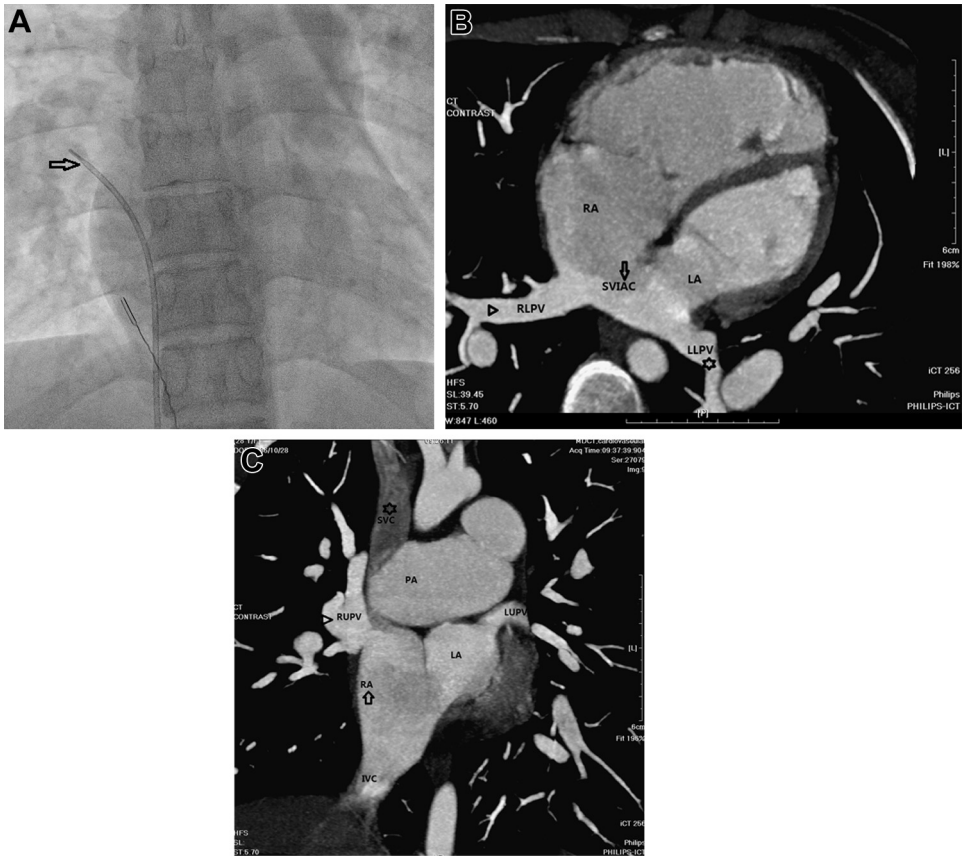

Figure 1. (**A**) Catheter in the anomalous right upper pulmonary vein (RUPV) (**arrow**) connecting with the right atrium (RA). (**B**) Multidetector computed tomography (MDCT) showed a large sinus venosus interatrial communication (SVIAC) (**arrow**) and the anomalous drainage of right lower pulmonary vein (RLPV) (**arrowhead**) into the RA as well as the normal drainage of left lower pulmonary vein (LLPV) (**asterisk**) to left atrium (LA). (**C**) MDCT disclosed anomalous RUPV (**arrowhead**) returning to RA (**arrow**) as well as normal connection of left upper pulmonary vein (LUPV) with the LA. It also depicted the proximity of anomalous RUPV, RA, and the superior vena cava (SVC) (**asterisk**). IVC, inferior vena cava; PA, pulmonary artery.

SVIAC, PAPVR, and PAA was planned but then cancelled because of her reluctance at that moment. A bare metal stent was placed in the LMCA (Fig. 2C), and IVUS showed relief of extrinsic compression after coronary stenting (Supplemental Fig. S7) in October 2015. Meanwhile, after 2 years of targeted medication therapy for PAH, mean PAP decreased to 38 mm Hg; PVR decreased to 3.5 Wood unit; and shunting was solely left to right, with a pulmonary to systemic flow ratio of 2.3.

In July 2018, follow-up angiogram revealed patency of the LMCA stent (Supplemental Fig. S8). Five years after the initial diagnosis, she finally agreed to undergo surgery, and thus surgical closure for SVIAC and baffling of PAPVR to the left atrium were performed in August 2018. She recovered well after surgery with functional class II symptoms and is kept on targeted medication for PAH.

Discussion

The latest 2018 American guideline for adult congenital heart disease recommends not closing interatrial communication if systolic PAP is more than half of the systolic aortic pressure or if PVR is more than one third of the systemic vascular resistance.[1] Nevertheless, 2 strategies have been proposed: namely, treat-and-repair[2] and repair-and-treat, using PAH medication before or after closure in interatrial communication with severe PAH. Our case had bidirectional shunting, high PVR and PAP initially, which contraindicated SVIAC closure. After receiving PAH medications for 2 years, shunt status was solely left-to-right, and PVR and mean PAP were lower. Our case is a good demonstration of the treat-repair-treat strategy for interatrial communication with severe PAH.

Our case is unique in showing a congenital heart PAH-related PAA with a diameter of only 3.75 cm (Supplemental Fig. S4, S5) that resulted in extrinsic compression of LMCA ostium (Fig. 2A). Coronary angiogram follow-up 3 years after placement of bare metal stent in the LMCA demonstrated good patency without restenosis or recoil (Supplemental Fig. S8). Our case results are in line with those in the current literature showing good efficacy and safety of stenting for LMCA extrinsic compression in PAH-related PAA.[3]

Conclusion

Our case demonstrates that a treat-repair-treat strategy can be successfully applied to close a large interatrial communication with severe PAH. Clinicians should check for the possible existence of PAPVR in cases with interatrial communication or unexplained PAH. Moreover, in cases with severe PAH and angina, LMCA extrinsic compression by a

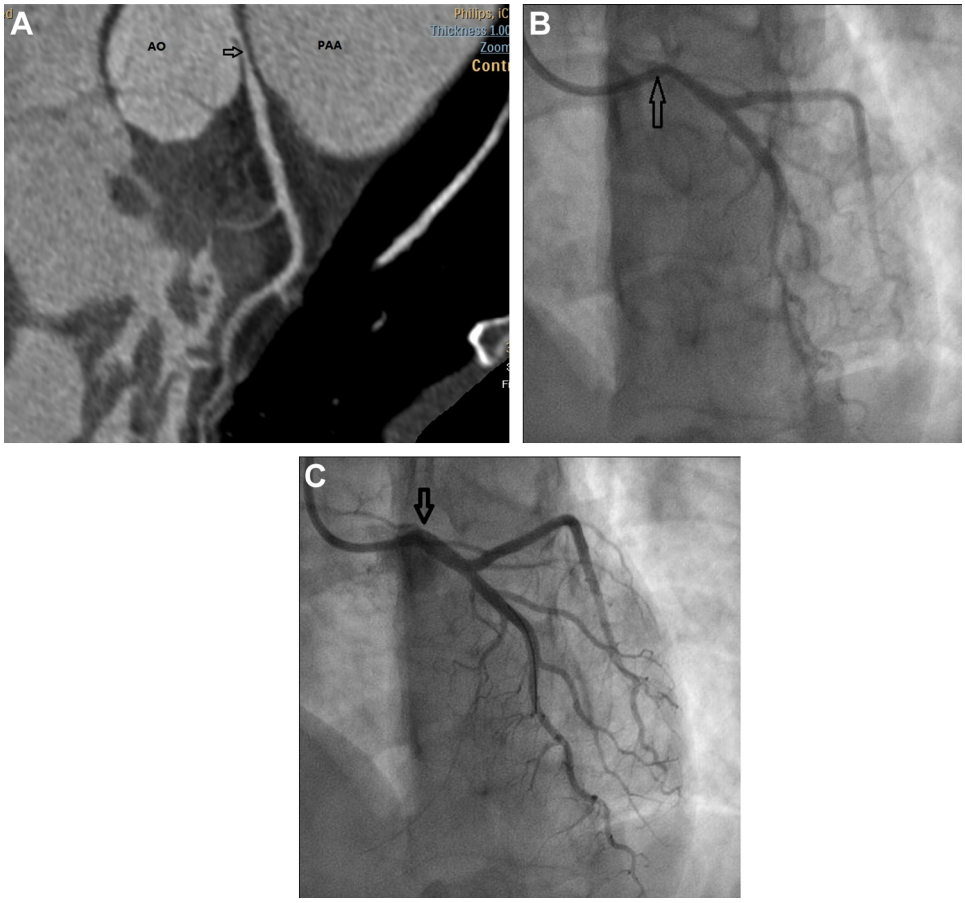

Figure 2. **A**) Multidetector computed tomography showed pulmonary artery aneurysm (PAA) compressing the ostium and proximal shaft (**arrow**) of left main coronary artery (LMCA). **B**) Left coronary angiogram showed a beak-shaped ostium of LMCA (**arrow**) due to extrinsic compression. **C**) After stenting, angiogram showed good expansion of LMCA ostium (**arrow**). AO, aorta.

PAA should be included in the differential diagnosis. LMCA stenting is a safe and effective solution for relieving PAA-related LMCA ostial compression.

Disclosures

The authors have no conflicts of interest to disclose.

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Supplementary Material

To access the supplementary material accompanying this article, visit the online version of the *Canadian Journal of Cardiology* at www.onlinecjc.ca and at https://doi.org.org/10.1016/j.cjca.2018.12.030.