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Case Report

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Extremely rare combination of diseases: Partial venous return anomaly and aortic dissection

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ABSTRACT
Anomalous pulmonary venous drainage (APVD) is a rare congenital anomaly in which one or more pulmonary veins connect to the right atrium, inferior or superior vena cava, azygous vein, or coronary sinus instead of the left atrium. APVD can be divided into two different types: total (TAPVD) and partial anomalous pulmonary venous drainage (PAPVD). We present an incidentally diagnosed PAPVD in a type one aortic dissection patient.
Keywords: Aorta, PAPVD, Type A dissection, Catheter

1. INTRODUCTION

Partial anomalous pulmonary venous drainage (PAPVD) is an uncommon congenital anomaly in which some of the pulmonary veins drain to the right heart. The prevalence of PAPVD is 0.1% in adulthood [1,2].

While, total (TAPVD) is usually symptomatic in childhood, PAPVD can remain asymptomatic and sometimes diagnosed incidentally, as in our case [3]. On the other hand, if the patient is symptomatic, chronic overloading of the right heart cavities can be the reason for the symptoms. Pulmonary hypertension and right heart failure are usually the reason behind the symptoms in the long term. Symptoms can be variable, such as dizziness by intense exertion, dyspnea, peripheral edema secondary to pulmonary hypertension, and right heart failure [4-6].

Acute aortic dissection (AAD) is a tear of the intimal layer [7]. AAD, especially type A, is a vascular emergency and is associated with high mortality and morbidity [8,9].

Concomitance of aortic dissection and pulmonary venous return anomalies are extremely rare. In this case, we will present a patient who was operated on for type 1 dissection and incidentally got diagnosed with PAPVD postoperatively.

2. CASE REPORT

A 48-year-old male patient was admitted to our emergency department with sharp substernal pain. There was no medical history. He was a nonsmoker. In the admission, his physical examination was normal. His blood pressure was 110/60 mmHg on both upper extremities. His distal pulses were palpable. His blood samples were normal. After thoracoabdominal computed tomography angiography (CTA) scanning, the diagnosis was type 1 aortic dissection. The echocardiography was performed at the bedside for aortic valve dysfunction. There was mild aortic regurgitation, and ejection fraction was normal as well.

He was emergently transferred to the operation room. Under general anesthesia, through the infraclavicular incision, we explored the right axillary artery, and arterial cannulation was performed. Following the median sternotomy, we opened the pericardium. The dissection went from the aortic root (starting from the right coronary artery orifice) and extending to the distal aorta. We performed supra coronary ascending aorta replacement with a right coronary artery (RCA) bypass since the RCA ostium was damaged. There was no complication. The patient was transferred to the intensive care unit (ICU). During the ICU follow-up, the chest x-ray showed that the left

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thorax was filled with hematoma. We decided to reoperate for the hematoma drainage and lung expansion. Three different surgical procedures were performed because of the bleeding problem in the left thorax. During these procedures, anesthesia removed the previous central venous catheter (CVC) from the right internal jugular vein due to catheter obstruction. They replaced the CVC from the left internal jugular vein. This lung complication prolonged the intubation time and ICU stay. After the extubation, intensive lung physiotherapy was necessary. The patient was occasionally taken to ICU from service for continuous positive airway pressure (CPAP). Blood gas samples were taken from the left CVC for patient comfort at the ICU CPAP treatment. The results were abnormal. The partial oxygen pressure (pO2) was 250 mmHg, unsuitable for venous blood samples. After repeated abnormal results of venous blood samples, arterial and venous blood samples were taken and compared simultaneously. It was seen that arterial pO2 was 86 mmHg while venous pO2 was 275 mmHg.

We figured that the tip of the CVC was not in the right atrium. We examined chest graphs, and the left CVC was out of the right atrium (Figure 1). In the thoracic CT scanning, we detected that the CVC went from the internal jugular vein to the superior vena cava to the pulmonary vein via the right atrium (Figure 2). Previous CTA was scanned, and detailed echocardiography was performed. The diagnosis was PAPVD. After the definitive diagnosis, the CVC was pulled back to two cm to the right atrium (Figure 3), and a control blood gas sample was taken. The result was suitable with venous blood gas results. The partial oxygen pressure was 67 mmHg, and oxygen saturation was 89%.

In our case, there was no time to do a detailed ECHO because of the emergent situation. It was performed at the bedside for valve dysfunctions and main cardiac functions. We diagnosed PAPVD postoperatively.

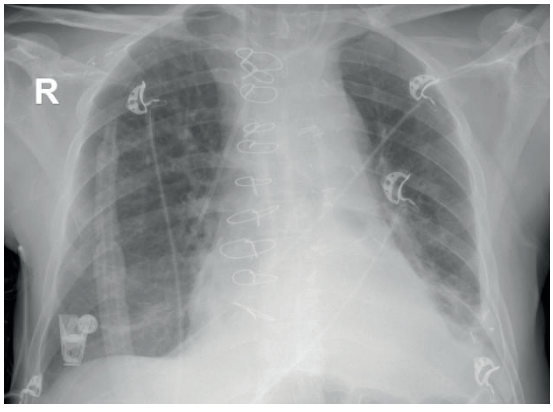

Figure 1. *Misplaced catheter (Chest X-ray)*

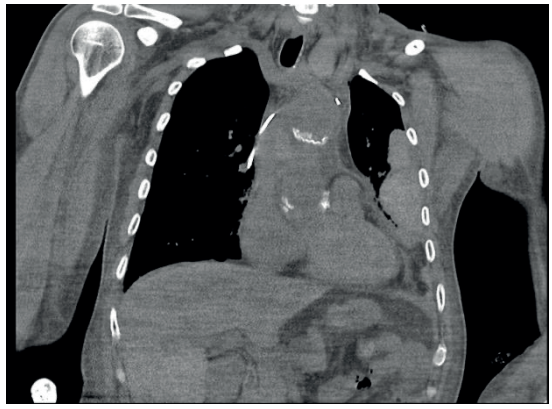

Figure 2. *Misplaced catheter (Thorax CT)*

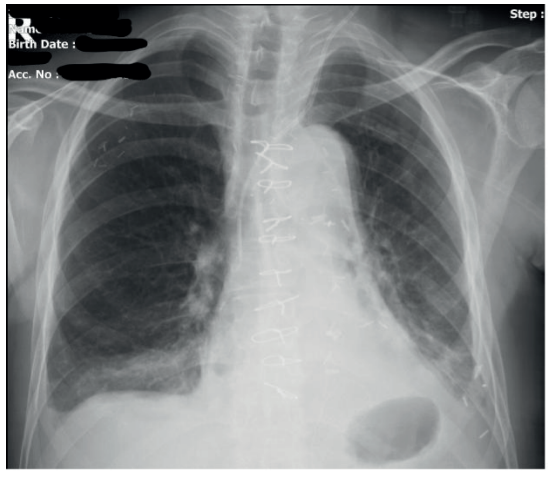

Figure 3. *Correctly placed catheter (Chest X-ray)*

3. DISCUSSION

Partial anomalous pulmonary venous drainage is a congenital disease. In childhood, it can be an isolated pathology or sometimes associated with other congenital heart diseases, particularly atrial septal defect (ASD) [10]. It can be described as supra-cardiac, infra-cardiac, cardiac, or mixed type. As in our patient, the upper right pulmonary vein draining to VCS is the most common type [11,12].

The isolated PAPVD may remain asymptomatic through adulthood. ECHO and CT scans are the main techniques used to diagnose PAPVD. ECHO and CT images are also essential to determine the type or how many or which pulmonary vein drains into the right atrium [13].

If the diagnosis is incidental, as in our patient, it should be kept in mind that if any venous blood gas sample results with higher pO2 levels and malposition of CVC on chest x-rays, it might be a sign of PAPVD. CTA is also diagnostic for the disease and perfectly describes the anatomical variation [3,14].

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The treatment options can be variable depending on the symptoms and severity of the shunt, in a study by Lewis et al. [15]. Patients with pulmonary hypertension were evaluated for PAPVD, and ninety patients were detected. Seventy-nine percent of these patients underwent CTA previously, but it was overlooked [2]. If the blood gas samples did not incidentally lead us to diagnose, we would also overlook CTA. In the same study, 15 patients were suitable for correction surgery (11 were associated with ASD, and four were isolated), and 13 underwent surgery. Other patients received medical therapy for pulmonary hypertension. In follow-up, 1 year after surgery, mild pulmonary hypertension was detected in our patient without symptoms. Medical follow-up was recommended.

Acute aortic dissection is a life-threatening condition with high mortality and morbidity rates [16]. The prevalence of AAD is approximately 0.2-0.8% in autopsy studies [17]. Patients usually present with sharp chest or back pain, as in our patient, or sometimes with malperfusion symptoms. A majority of patients benefit from emergency surgery. Surgery time, cardiopulmonary bypass, and cross-clamping time have a significant impact on prolonged ICU stay, hospitalization, and mortality [18,19].

Conclusion

There is no time to perform surgery for concomitant pathologies. Since, our patient underwent emergent surgery for aortic dissection, which means prolonged cardiopulmonary or cross-clamp time equal to higher mortality and morbidity, it was not essential to diagnose PAPVD preoperatively. However, before elective cardiac surgical procedures, it is important to detect PAPVD and decide on treatment options.

Compliance with Ethical Standards

This manuscript was conducted ethically in accordance with the principles of Helsinki World Medical Association Declaration.

Patient Consent: The patient gave his written consent for clinical information related to him to be reported in a medical publication.

Conflict of interest: The authors declare that there is no conflict of interest.

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Authors contributions: OD and ED: Writing the manuscript, ED, AG, FO and KA: Supervision, OT and KA: Methodology, OT and KA: Investigation, OT, FO, AG and ED: Formal analysis, All authors approved the final manuscript.

REFERENCES

[1] Ho M L, Bhalla S, Bierhals A, Gutierrez F. MDCT of partial anomalous pulmonary venous return (PAPVR) in adults. J Thorac Imaging 2009;24:89-95. doi: 10.1097/RTI.0b013e318194c942.

[2] Lewis RA, Billings CG, Bolger A, et al. Partial anomalous pulmonary venous drainage in patients presenting with suspected pulmonary hypertension: A series of 90 patients from the ASPIRE registry. Respirology 2020;25:1066-72. doi: 10.1111/resp.13815.

[3] Verma A, Sethi S, Kohli N. Partial anomalous pulmonary venous connection: state-of-the-art review with assessment using a multidetector computed tomography angiography. Pol J Radiol 2022; 87:549-56. doi: 10.5114/pjr.2022.120513.

[4] Ludeña-Martín-Tesorero R, Martín-Fernández M, Calvo-Blanco J, Álvarez-Velasco R. Partial anomalous pulmonary venous return: a casual finding in many cases. Arch Cardiol Méx 2023;93:504-05. doi: 10.24875/ACM.22000242.

[5] Sears E H, Aliotta J M, Klinger J R. Partial anomalous pulmonary venous return presenting with adult-onset pulmonary hypertension. Pulm Circ 2012;2: 250-5. doi: 10.4103/2045-8932.97637.

[6] Galiè N, Humbert M, Vachiery J L, et al. 2015 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension: the joint task force for the diagnosis and treatment of pulmonary hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT). Eur Heart J 2016;37:67-119. doi: 10.1093/eurheartj/ehv317.

[7] Pape LA, Awais M, Woznicki EM, et al. Presentation, diagnosis, and outcomes of acute aortic dissection: 17-year trends from the International Registry of Acute Aortic Dissection. J Am Coll Cardiol 2015;66:350-8. doi: 10.1016/j.jacc.2015.05.029.

[8] Prisant LM, Nalamolu VR. Aortic dissection. J Clin Hypertens (Greenwich) 2005;7:367-71. doi: 10.1111/j.1524-6175.2005.04116.x.

[9] Czerny M, Grabenwöger M, Berger T, et al. EACTS/STS Guidelines for diagnosing and treating acute and chronic syndromes of the aortic organ. Ann Thorac Surg 2024;118:5-115. doi: 10.1016/j.athoracsur.2024.01.021

[10] Singhal K, Newton AD, Corbett C, Predina JD. Management of partial anomalous pulmonary venous connections in patients requiring pulmonary resection: a case report and systematic review. J Thorac Dis 2017;9:5434-9. doi: 10.21037/jtd.2017.11.68.

[11] Demos TC, Posniak HV, Pierce KL, et al.. Venous anomalies of the thorax. AJR Am J Roentgenol 2004; 182: 1139-50. doi: 10.2214/ajr.182.5.1821139.

[12] Snellen HA, van Ingen HC, Hoefsmit EC. Patterns of anomalous pulmonary venous drainage. Circulation 1968; 38: 45-63. doi:10.1161/01.cir.38.1.45

[13] Hsu HM, Chang YT, Su WJ, Chu JJ, Chang YS, Hwang MS. The morphogenesis and associated anomalous pulmonary venous drainage in sinus venosus defect. Pediatr Neonatol 2020;61:92-9. doi: 10.1016/j.pedneo.2019.06.013.

[14] Razek A A K A, Al-Marsafawy H, Elmansy M, Abd El-Latif M, Sobh D. Computed tomography angiography and magnetic resonance angiography of congenital anomalies of pulmonary

veins. J Comput Assist Tomogr 2019;43:399-405. doi: 10.1097/RCT.000.000.0000000857.

[15] El-Kersh K, Homsy E, Daniels C J, Smith J S. Partial anomalous pulmonary venous return: A case series with management approach. Respir Med Case Rep 2019;27:100833. doi: 10.1016/j.rmcr.2019.100833.

[16] Malaisrie S C, Szeto W Y, Halas M, et al. 2021 The American Association for Thoracic Surgery expert consensus document: surgical treatment of acute type A aortic dissection. J Thoracic Cardiovasc Surg 1021;162: 735-58. doi: 10.1016/j.jtcvs.2021.04.053.

[17] Pacini D, Di Marco L, Fortuna D, et al. Acute aortic dissection: epidemiology and outcomes. Int J Cardiol 2013;167:2806-12. doi: 10.1016/j.ijcard.2012.07.008.

[18] Wen M, Han Y, Ye J, et al. Peri-operative risk factors for in-hospital mortality in acute type A aortic dissection. J Thorac Dis 2019;11:3887-95. doi: 10.21037/jtd.2019.09.11

[19] Schwartz J P, Bakhos M, Patel A, Botkin S, Neragi-Miandoab S. Impact of pre-existing conditions, age, and the length of cardiopulmonary bypass on postoperative outcome after repair of the ascending aorta and aortic arch for aortic aneurysms and dissections. Interact Cardiovasc Thorac Surg 2008;7:850-4. doi: 10.1510/icvts.2008.182303.