OXFORD

Oxford Medical Case Reports, 2025, **4**, 184–188
https://doi.org/10.1093/omcr/omae214
Case Report

Diagnostic evaluation and management of pulmonary hypertension with concomitant incidental partial anomalous pulmonary venous return

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Abstract

Partial anomalous pulmonary venous return (PAPVR) is a rare congenital heart condition which is often asymptomatic and hence remains undiagnosed, but could occasionally be detected on thoracic imaging as an incidental finding. For rare cases of newly diagnosed pulmonary hypertension with concurrent PAPVRs, the diagnostic workup and subsequent management are more complicated, requiring a thorough evaluation of secondary causes of pulmonary hypertension, and assessing relative PAPVR shunt contribution to the pulmonary hypertension. We herein report a case of a 74-year-old Chinese male patient, a chronic smoker of 50 pack-years, and past medical history of hypertension and diabetes mellitus, who was admitted to our intensive care unit with acute decompensated type 2 respiratory failure, and subsequently found to have newly diagnosed pulmonary hypertension with right heart failure and an incidental PAPVR identified on inadvertent central venous cannulation (CVC) of the anomalous pulmonary vein draining into the left internal jugular vein. There are a few key learning points from this case study: firstly, we profile the characteristics and clinical outcomes of cases of incidental CVC cannulation of undiagnosed PAPVR from a literature review; and secondly, we discuss the diagnostic and management approach to newly diagnosed pulmonary hypertension with concomitant, incidental PAPVR that may be useful for internists and critical care physicians.

Keywords: critical care medicine

Case presentation

A 74-year-old Chinese male, a smoker of 50 pack-years, with background of diabetes mellitus, and hypertension, presented with a 3-day duration of breathlessness, dry cough and lower limb edema. On examination, he was drowsy, with bilateral lung crepitations on chest auscultations. Initial investigations (Table 1) revealed acute decompensated type 2 respiratory failure (T2RF) on arterial blood gas, sinus tachycardia with right heart strain patterns on electrocardiogram (ECG) (Fig. 1), and patchy bilateral lung field airspace opacities on chest X-ray (CXR) (Fig. 2A). A point-of-care-ultrasound (POCUS) showed a hypokinetic and dilated right ventricle, with a D-shaped septum. The clinical differentials included infective exacerbation of undiagnosed chronic obstructive pulmonary disease (COPD), acute decompensated heart failure, and pulmonary embolism (PE).

The patient was admitted to the intensive care unit, and initiated on non-invasive ventilation (inspiratory positive airway pressure 16 mmHg, expiratory positive airway pressure 6 mmHg), nebulised bronchodilators, antibiotics, and diuretics therapy. A left internal jugular vein (IJV) central venous catheter (CVC) was inserted under ultrasound guidance, with repeat CXR showing the central line abnormally sited in the left hemithorax (Figure 2A). A computed tomography pulmonary angiogram (CTPA) was performed to rule out PE, which showed bilateral consolidation, extensive emphysema and an incidental anomalous left superior pulmonary vein (PV) draining into the left brachiocephalic vein (Figure 2B). As the CVC had inadvertently cannulated the anomalous PV, the central line was removed and re-sited in the right femoral vein.

The patient made a good recovery within a few days, and was transferred to the general ward. During the same admission, a transthoracic echocardiogram was done which showed severe right ventricular systolic dysfunction, elevated estimated pulmonary artery systemic pressure of 68 mmHg, with preserved left ventricular ejection fraction. Hence, he was discharged with cardiology follow-up, and an elective right heart catheterisation was performed, demonstrating mixed pre-capillary pulmonary hypertension (PH). As the PAPVR shunt fraction was relatively low at 1.35, the patient's PH was predominantly contributed by undiagnosed COPD (group 3), with minor contribution from the PAPVR (group 1). He was treated with regular bronchodilator therapy, long-term oxygen therapy (LTOT), sildenafil and spironolactone.

Received: July 17, 2024. **Revised:** December 14, 2024. **Accepted:** December 25, 2024

Table 1. Pertinent investigations in this patient with first-onset pulmonary hypertension with concomitant incidental PAPVR.

Laboratory Tests	**Result(s) (Reference Range(s))**
High sensitivity Troponin I (serial trending of 3 sets of troponins)	52.6 ng/l, 54.3 ng/l, 61.7 ng/l (0–26.2 ng/l)
NT-proBNP	7042 ng/l (≤ 125 ng/l)
Arterial blood gas (ABG)	pH: 7.22 (7.35–7.45) pO_2 (taken on *non-rebreather mask*): 401 mmHg (75–105 mmHg) pCO_2: 74.8 mmHg (35–45 mmHg) Bicarbonate: 30.4 mmol/l (22–28 mmol/l)
Autoimmune markers	RF: < 20 IU/ml (≤ 30 IU/ml) Anti-CCP: < 0.5 U/ml (≤ 5 U/ml) ANA: < 1:80 (negative titre: < 1:80) Anti-dsDNA: < 10 (<100 IU/ml) C3: 93 mg/dl (82–185 mg/dl) C4: 33 mg/dl (15–53 mg/dl) Anti-ENA panel (anti-Ro, anti-La, anti-RNP, anti-Sm, anti-Jo1, anti-Scl 70): negative
Electrocardiogram (ECG)	Sinus rhythm, no ischaemic ST/T changes, presence of right bundle branch block with strain pattern, right axis deviation, and S1Q3T3 pattern
Radiological investigations	
CT pulmonary angiogram	• Dilated pulmonary arteries and right ventricle suggestive of pulmonary arterial hypertension. • Nodular consolidative changes in bilateral lung fields, with mildly enlarged mediastinal lymph nodes. • Moderate centrilobular emphysema present. • No pulmonary embolus. • No radiological features suggestive of parenchymal lung disease.
Transthoracic echocardiogram	• Normal left ventricular ejection fraction (LVEF; 55%) • Severe right ventricular systolic dysfunction with dilated right heart chambers • Moderately severe pulmonary hypertension (PASP 68 mmHg) • Mild tricuspid regurgitation • No atrial septal defect
Procedures	
Right heart catheterisation	• Mean pulmonary arterial pressure (PAPm): 37 mmHg (main pulmonary artery), 38 mmHg (right pulmonary artery) • Mean pulmonary arterial wedge pressure (PAWP): 11 mmHg (with large variations as expected in COPD patients) • Pulmonary vascular resistance (PVR) 5.8 Wood units • Shunt fraction of partial anomalous pulmonary vein drainage (left upper pulmonary vein to left brachiocephalic vein) Qp:Qs 1.35

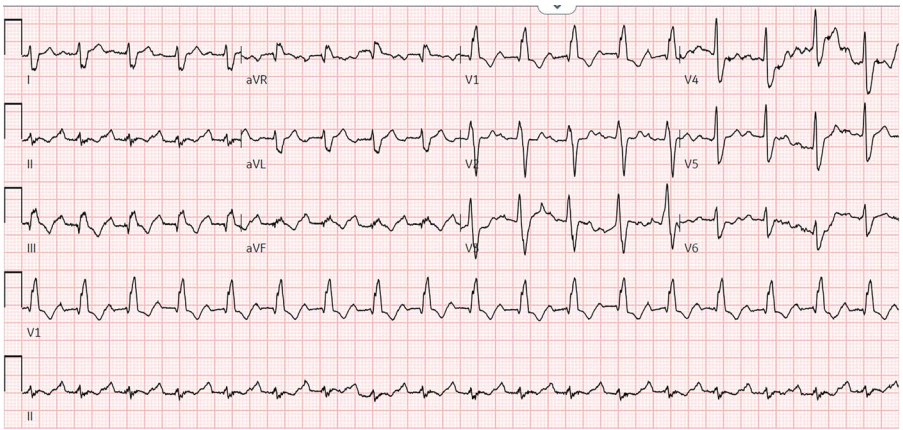

Figure 1. ECG showing sinus tachycardia, and features of right heart strain (right bundle branch block, right Axis deviation and S1Q3T3 pattern).

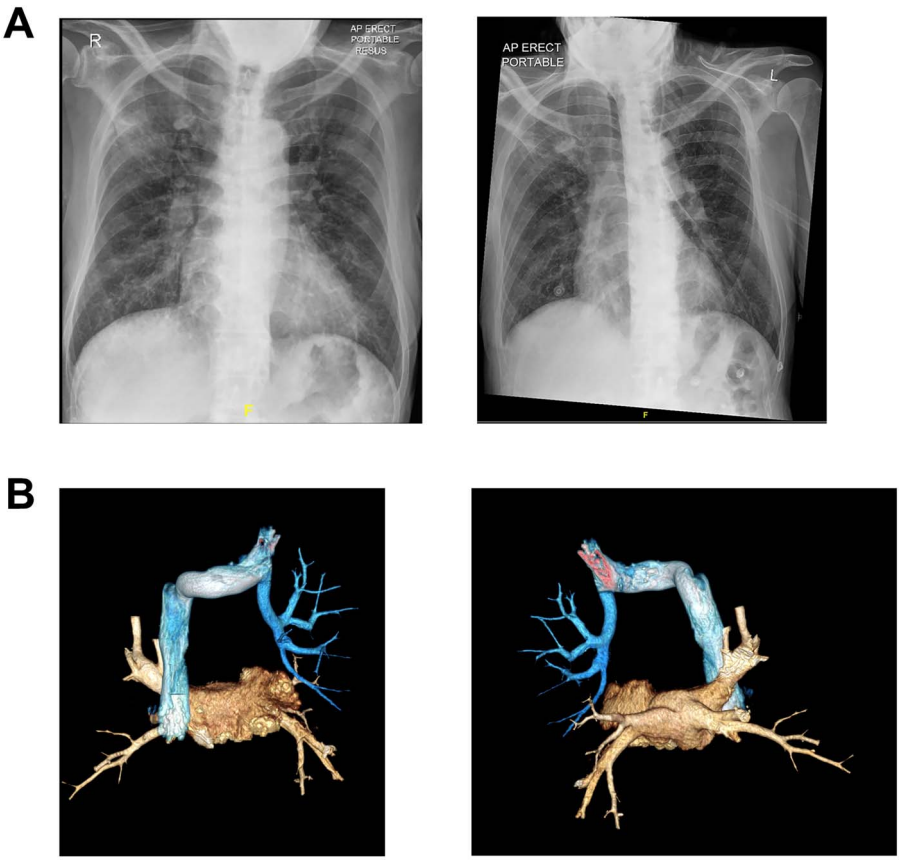

Figure 2. Incidental PAPVR identified on central venous cannulation. (A) Initial CXR (left); Post-CVC placement CXR showing inadvertent cannulation of anomalous left pulmonary vein (right). (B) CTPA (with CT 3D reconstruction) showing left-sided partial anomalous pulmonary venous return anterior view (left); posterior view (right).

While awaiting for an outpatient lung function testing to confirm the diagnosis of COPD, the patient was re-admitted with severe pneumonia, and developed cardiac arrest during hospitalisation leading to demise.

Discussion

We herein discuss two major learning points from this unique case study pertaining to inadvertent CVC cannulation of an anomalous pulmonary vein, and evaluation/management of newly diagnosed PH in context of incidental PAPVR.

Firstly, PAPVR is a rare (0.4–0.7%), congenital heart condition, characterised by aberrant drainage of ≥1 PVs into the right atrium (RA) or its venous tributaries [1]. Anomalous PVs can be either unilateral or bilateral, but are more commonly right-sided (80%–90%) [2, 3], and can drain into RA, superior vena cava (SVC), inferior vena cava (IVC), brachiocephalic (innominate) vein, azygos vein and or coronary sinus [4]. Typically, left-sided PAPVR drains into the left brachiocephalic vein through an anomalous vertical vein [3]. Isolated PAPVRs usually remain asymptomatic, do not cause clinically significant pulmonary hypertension [5], and are often incidental findings on thoracic imaging, post-mortem studies or even rarely, inadvertent central venous cannulation of an anomalous PV [6].

Till date, less than 20 cases of CVC cannulation of anomalous PVs have been reported in literature, with most cases involving left IJV CVC placement into left-sided PV that drains into left brachiocephalic vein (Appendix S1). In all reported cases, CVC blood gases had paradoxically higher PaO_2 than peripheral ABG and on pressure transduction, nearly all showed a 'venous' waveform, except for a case of pulsatile 'arterial' waveform that was attributed to catheter tip wedging into a pulmonary venous branch which led to pressure tracing of upstream pulmonary artery being captured [7]. Interestingly, three cases documented successful usage of the malpositioned CVC in an anomalous PV with no adverse complications. Further studies are needed to assess if CVC placement in anomalous PV is safe for usage in critical care settings, where re-siting may not always be practical in hemodynamically unstable patients or those with difficult vascular access.

However, it is important to recognise that CVC malpositioning occurs in 6.7% of cases [8], most commonly involving the left IJV or subclavian vein cannulation [9]. Besides cannulation of undiagnosed left-sided PAPVR, other differentials for a malpositioned left IJV include cannulation of left carotid artery, persistent left SVC and azygos vein [6], which can be distinguished through clinical/biochemical/radiological features (Appendix S2). Definitive identification/characterisation of a PAPVR can be done through contrasted CT, cardiac magnetic resonance imaging (cMRI) or transoesophageal echocardiography [10, 11].

Secondly, management of newly diagnosed PH hinges on identifying the predominant contributor(s) of PH and assessing PAPVR shunt significance. Although a PAPVR acts as a partial left-to-right shunt that contributes to pulmonary hypertension, it is known that each PV only contributes 20%–25% of total PV drainage

and therefore isolated PAPVRs without concurrent ASD or other left-to-right cardiac shunts would not typically cause cardiac decompensation or warrant surgical intervention [11]. However, surgical repair may be considered in symptomatic disease, right ventricular enlargement, significant shunt fraction (Qp/Qs > 1.5) or in planned lobectomy for lung cancer, which has generally excellent outcomes with low complication rates [3, 12]. On the other hand, a recent study found that unrepaired cases of PAPVR with intact atrial septum had no temporal deterioration in clinical or right heart indices at 3- to 5-year follow-up [13].

Right heart catheterisation remains the gold standard modality to diagnose and classify PH (pre-capillary, post-capillary and mixed) based on predefined hemodynamic criteria [14]. PAPVR shunt fraction (Qp/Qs) is assessed through the modified Fick's equation [15]. Further evaluation of secondary causes of PH includes autoimmune workup for connective tissue disease-related PH, TTE/coronary evaluation for left heart disease, high-resolution CT thorax/lung function testing for airway/parenchymal lung disease, and ventilation-perfusion scan for chronic thromboembolic pulmonary hypertension.

Management of PH with an isolated PAPVR without clinically significant shunt fraction largely hinges on addressing the predominant aetiology. In our patient's case, he likely had advanced COPD causing severe PH with cor pulmonale. He was advised for smoking cessation, commenced on LTOT based on the British Thoracic Society's clinical indication (if resting $PaO_2 \leq 55$ mmHg or 55–60 mmHg with pulmonary hypertension, cor pulmonale or polycythemia) [16] and bronchodilator therapy. He was given diuretic therapy during volume overloaded states to reduce right ventricular preload, and improve its contractility/function. The decision to initiate vasodilator therapy requires balancing its hemodynamic benefits in alleviating pulmonary arterial pressures and consequent right heart failure (by reducing afterload), with the theoretical risk of worsening hypoxaemia through exacerbating V/Q mismatch by altering the physiologic hypoxic pulmonary vasoconstriction in COPD patients [17]. In particular, sildenafil, a phosphodiesterase-5 inhibitor, appears to be both efficacious in treating pulmonary hypertension, and well-tolerated [17]. The recent COMPERA trial showed that COPD patients with PH were functionally more impaired with poorer outcomes than those with idiopathic pulmonary arterial hypertension, and found that those who received PH medical therapy (primarily phosphodiesterase-5 inhibitors) had improved function (in terms of 6-minutes walking distance, WHO functional class) and better outcomes [18]. Further studies are required to evaluate the clinical/functional outcomes of using vasodilator therapies in the unique subgroup of patients with PH caused by COPD with coincident PAPVR.

Supplementary material

Supplementary material is available at the *Journal of Surgical Case Reports* online.

Conflict of interest

None declared.

Funding

None.

Guarantor

Adj A/Prof Adrian CL Kee is the guarantor of this article.

Consent

Written consent was obtained from patient.

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https://doi.org/10.1093/omcr/omae214
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