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Partial anomalous pulmonary venous return in a patient undergoing left upper lobectomy for adenocarcinoma of the lung: A case report

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ABSTRACT

INTRODUCTION: Partial anomalous pulmonary venous return (PAPVR) is a rare congenital heart disease that complicates surgical management of pulmonary pathology.
CASE PRESENTATION: This case describes the successful management of a 73-year-old female with a left upper lobe adenocarcinoma and pre-operative discovery of left superior anomalous pulmonary venous return into the innominate vein. This patient presented to our clinic for evaluation regarding her newly discovered adenocarcinoma of the lung. Here, we also discuss findings in the literature for management of these patients regarding the importance of preoperative evaluation to determine the extent to which a lobectomy will alter pulmonary function with special emphasis on identifying patients at risk of increased shunting leading to cardiopulmonary failure.
CONCLUSION: Consideration should focus on the extent of the shunting, the presence of symptoms, and underlying right heart strain or pulmonary hypertension.

1. Introduction

Partial anomalous pulmonary venous return (PAPVR) is a rare congenital heart disease that occurs in 0.4–0.7% of the population [1]. When this occurs, pulmonary veins connect partially to right circulation or with an incomplete connection with the left atrium. Most commonly, this condition afflicts adults by presenting with the left superior pulmonary vein draining to the innominate vein or the right superior pulmonary vein draining to the superior vena cava. Often times, PAPVR presents with a concurrent atrial septal defect (ASD) found in up to 80% of patients [2].

Patients with PAPVR requiring lung resection, pose a unique challenge in today's world of medicine, as there is no standardized treatment protocol given the rarity of this type of case presentation [1]. Although not protocoled, consideration of PAPVR patient anatomy and physiology remains imperative for treatment planning during thoracic surgery as alterations in blood flow during surgery could result in serious complications including pulmonary artery hypertension and subsequently right ventricular failure [1,3,4]. The following case presents a patient with PAPVR and adenocarcinoma of the left lung undergoing left upper lobectomy in an academic hospital. This case report has been reported in line with the SCARE criteria [5].

2. Presentation of case

We present the case of a 73-year-old female who presented to Thoracic Surgery clinic having had a left upper lobe pulmonary nodule incidentally found on CT scan of her chest, performed during an episode of pneumonia. Imaging revealed a four by six-millimeter solid nodule within a two-centimeter area of nodular ground glass opacity in the anterior left upper lobe. Additionally, there was a medial posterior left upper lobe seven-millimeter nodule near the oblique fissure. While the medial posterior nodule remained stable, the anterior nodule increased to five by nine millimeters over the next year. A subsequent CT guided core biopsy was notable for atypical adenomatous hyperplasia. With increased monitoring, the area of hyperplasia increased to ten by fourteen millimeters over the next year, prompting surgical evaluation. She had been completely asymptomatic from this nodule.

Her past medical history includes hypertension, anxiety, hypothyroidism, dyslipidemia, and osteoporosis. She takes aspirin, metoprolol, pravastatin, paroxetine, levothyroxine, and a calcium containing multivitamin on a regular basis. She has a ten pack-year smoking history. She has no family history of anatomic abnormalities. Pulmonary function was evaluated, her FEV1 was 2.26 L and DLCO 91.3% of predicted. Following discussion at our institutional thoracic oncology review board, a recommendation was for wedge resection with conversion to left upper lobectomy if intra-operative pathologic evaluation were to indicate malignancy. Fig. 1 shows the left upper lobe anomalous pulmonary venous return.

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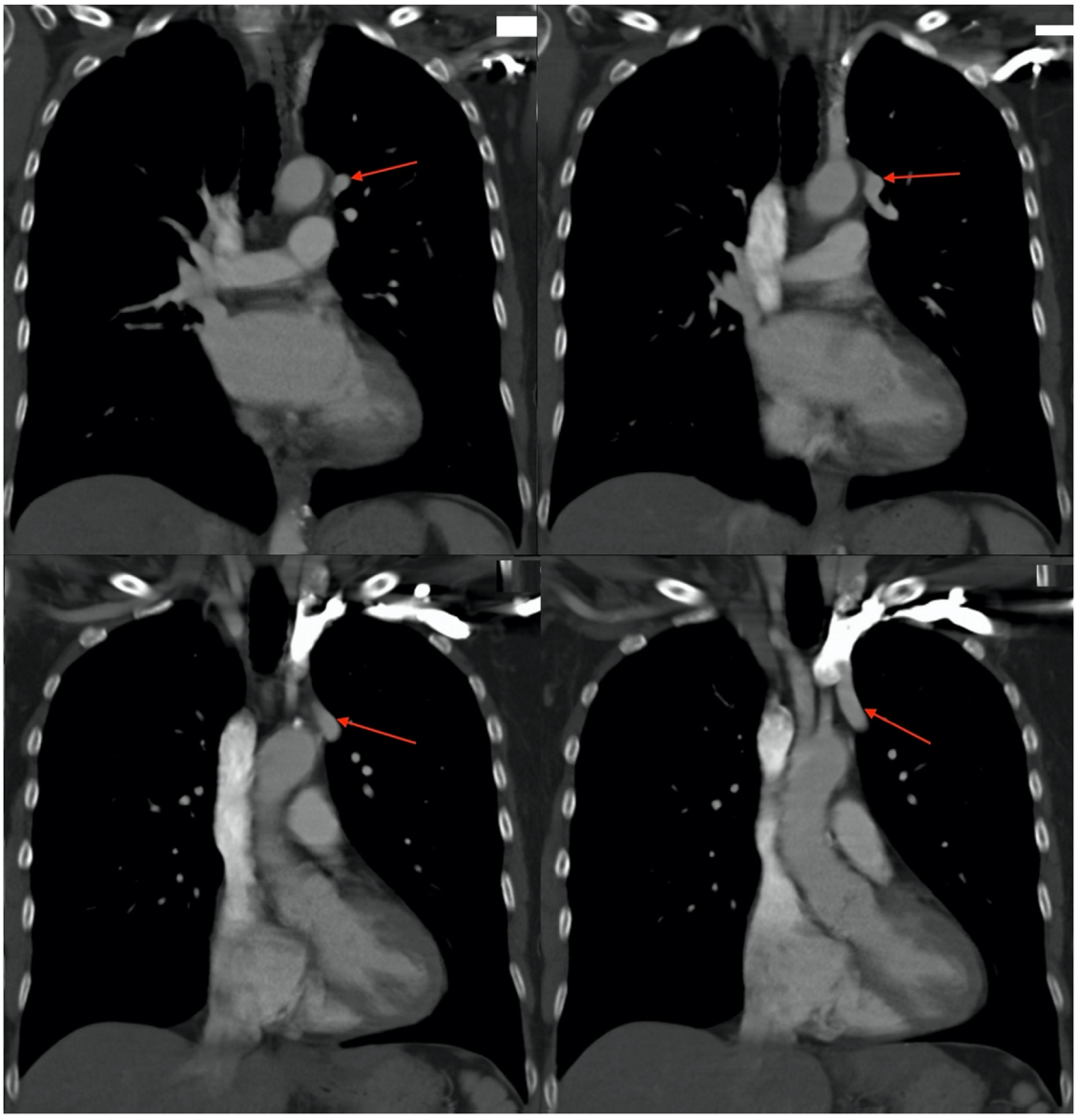

Fig. 1. CT scan of patient's chest with red arrow demonstrating the left upper venous return directly to the left innominate vein.

During her pre-operative work-up, imaging was consistent with left superior anomalous pulmonary venous return. Her surgery was conducted by an experienced Cardiothoracic Surgeon with the assistance of a General Surgery Resident. She subsequently underwent video assisted thoracoscopy which confirmed left superior pulmonary venous return directly into the left innominate vein. During video-assisted thoracoscopy for an intended wedge resection, PAPVR was confirmed, which, along with other intra-operative factors, warranted the need for conversion to an open thoracotomy and left upper lobectomy. Given her lack of symptoms due to the PAPVR, her favorable PFTs, and the location of the PAPVR, the anomalous connection was confirmed to be directly from the left superior pulmonary vein and then suture ligated near its insertion into the innominate vein. The phrenic nerve was draped over the vertically oriented anomalous vein and preserved throughout the operation. The lingular pulmonary vein was confirmed to be entering the left atrium. She concurrently underwent left hemithorax lymphadenectomy with nodes identified at levels L4, 5, 6, 7, L9, L10, and L11 lymph nodes. There were no complications during the procedure.

The patient's final pathology showed two foci of moderately-differentiated adenocarcinoma, acinar type. The largest being 2.0 cm in size with visceral pleural invasion. Two level 4 and one level 11 nodes were positive with extra capsular extension and lymphovascular invasion. Changing the patient's clinically cT1bN0M0 Stage IA2 disease state to a pathologic pT3pN2cM0 stage IIIB.

Follow-up consisted of post-operative clinical visits every few weeks to months. She tolerated the procedure well and had no adverse events, specifically no reported dyspnea. Since, she has recently completed pemetrexed and carboplatin-based chemotherapy and is currently undergoing radiation therapy with curative intent.

3. Discussion

Less than 1% of adults were found to have PAPVR during autopsy, and those without an ASD were found to be asymptomatic [6]. This demonstrates the low impact this aberrancy has on the lives of those adults with PAPVR. This less efficient circulation places additional strain on the circulatory system, becoming important when

considering lobectomy or pneumonectomy, by necessitating the need to also consider the effect of the pulmonary resection on the patient's circulation. Removal of the pulmonary tissue may result in right heart failure due to the increased proportion of blood traveling through the anomalous connection [1,3,4]. Therefore, patients presenting for lobectomy or pneumonectomy for nodule resection require additional intra- and post-operative precautions as a result of their presenting condition.

Our patient presented with the anomalous connection arising from the left upper lobe. There are no set guidelines for the management of this rare anomaly. However, there are themes and general recommendations noted within case reports that serve to guide management of this congenital anomaly when faced in the setting of pulmonary resection. Performing a thorough preoperative work-up for lobectomies to identify these rare anomalies and concomitant structural anomalies and to assess for surgical correction of PAPVR based on symptomatology and the extent of right to left shunting remains of utmost importance and will help to prevent potentially serious postoperative complications.

Despite low prevalence of PAPVR, careful and complete preoperative imaging of pulmonary venous blood flow is essential to determine the extent a lobectomy will affect lung function [4,7,8]. The imaging modality of choice has been described as both transthoracic echocardiography (TTE) and dual-source CT [9,10]. A feared complication is increasing shunt flow after lobectomy with a co-existing PAPVR in a different lobe. There have also been described cases in which the PAPVR presented on the left but the intended resection on the right, leading to increased shunt flow and subsequent right heart failure [4]. Roughly 80–90% of cases of PAPVR have an associated ASD [2,11]. Special care should be given to this relationship as this could affect surgical management [12].

Guidelines for a primary surgical repair of all PAPVR have not been well defined on a global scale. Recommendations based on general consensus are based on the extent of left to right shunting and the presence of symptoms. Patients who have these anomalies are usually asymptomatic and, therefore, these connections in general are not clinically significant [6]. Surgery is not recommended for asymptomatic patients when the risks outweigh the benefits [13]. Should a wedge resection or lobectomy be necessary for another reason and the PAPVR is not going to be included in the resection, careful evaluation of lung function pre-operatively, calculation of the extent of shunting represented by the shunt fraction, Qp/Qs, and close follow up are essential to management [1]. This is due to the significant left to right shunting that may result in acute right heart failure in the presence of a contralateral anomalous connection [4]. These cases are evaluated on an individual basis.

Primary repair of PAPVR should be undertaken in patients who are symptomatic and may need to undergo life-saving repair [13]. For these cases, there are some general guidelines in the form of physiologic parameters, which, when met, suggest the patient will benefit from definitive surgical repair. For patients with large left to right shunts, typically in the presence of multiple pulmonary veins, there can be significant right ventricular overload and pulmonary hypertension for which surgical correction of the anomaly can often reverse the failure [14]. Current recommendations for surgery include a shunt fraction, Qp/Qs, greater than 1.5 with or without symptoms [1]. A Qp/Qs less than 1.5 but in a patient with symptoms should warrant a more thorough evaluation and work-up assessing risks and benefits of correction [1]. In cases where surgery should be delayed in high risk patients despite evidence of severe right heart failure, ongoing follow-up and risk reassessment provides a reasonable option with the hope for future intervention [11].

4. Conclusion

Although PAPVR is rare, knowledge of its existence is essential for the avoidance of potential adverse outcomes during thoracic surgery. Management of PAPVR depends on preoperative imaging to determine extent and location of the anomalies. Primary repair of these connections is generally not indicated without evidence of right heart strain either due to reported symptom severity or calculation of a clinically significant shunt fraction. The lack of research on the role extensive diagnostic studies with the utility for a guideline-based approach remain an area of study that could prove beneficial when faced with such a clinical dilemma.

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Ethical approval

As a Case Report, this study is exempt from ethical approval.

Consent

Written informed consent was obtained, all identifying information has been omitted to protect anonymity.

Author contribution

Ankit Verma, MD – research and writing the paper.
Xander Jacobson – research and writing the paper.
Katherine Nordick – researching and writing the paper.
Vincent Nicchi – researching and writing the paper.
Marcus Balters, MD – design, interpretation, and analysis.

Registration of research studies

This case report is not registered due to being a case report that is not a first-in-man or animal study, per Research Registry guidelines (https://www.researchregistry.com/help-and-support/faqs).

Guarantor

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Declaration of Competing Interest

The authors report no declarations of interest.

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