Unilateral pulmonary vein atresia presenting with recurrent hydrothorax in an adult: A case report

Yan Wang, Shuangshuang Jiang, Jiasheng Yang, Ruilin Sun *
Guangdong Second Provincial General Hospital, China

A B S T R A C T

UPVA (Unilateral pulmonary vein atresia) is the failure of connection between the common pulmonary vein and the left atrium. UPVA is a rare malformation of common pulmonary vein caused by embryonic development defects. Isolated UPVA is uncommon, the diagnosis commonly occurs during early childhood because of asthma, recurrent pneumonia or hemothysis, but diagnosis in adults is unusual. Some patients can be asymptomatic until adulthood. In this report, we describe a case about UPVA presenting with recurrent hydrothorax in an adult. We gradually carried out routine diagnostic methods and eventually confirmed the rare UPVA according to the two common clinical manifestations of repeated pleural effusion and hilar soft tissue shadow.

1. Background

UPVA (Unilateral pulmonary vein atresia) is the failure of connection between the common pulmonary vein and the left atrium [1]. UPVA is a rare malformation of common pulmonary vein caused by embryonic development defects. About 50% of the patients had the complications-the other cardiac malformations or ectopic pulmonary venous drainage [2]. Isolated UPVA is uncommon, the diagnosis commonly occurs during early childhood because of asthma, recurrent pneumonia or hemothysis, but diagnosis in adults is unusual. Some patients can be asymptomatic until adulthood.

2. Case presentation

A 30-years-old woman was referred to our hospital for recurrent hydrothorax presented with 3 months dysnea. In June 2021, the patient was admitted to another hospital for pleural effusion. The surgery of pleural biopsy and closed drainage had been done for her, revealing chronic inflammatory and transudate respectively. However, her hydrothorax was still unexplained. Hence, a contrast-enhanced CT was performed, showing soft tissue mass-like lesions in right mediastinum and the right hilum of the lung, for further diagnosis and treatment the woman was transferred to our hospital at July 2021.

Physical examination showed normal vital signs for age. Pulmonary signs showed remarkable diminished breath sounds over the right lower lung field. Oxygen saturation was 98% when breathing air. Contrast-enhanced chest CT performed at our hospital confirmed a soft tissue mass-like lesions in the right mediastinal and hilum of the lung with multiple tortuous vessel shadows; the right pulmonary vein was not clearly shown; and the trunk of right pulmonary artery was obviously narrowed; the left pulmonary vein shows compensatory thickening; pleural effusion on the right side (Fig. 1).

To further investigate the unexplained symptom and the mass, we systematically performed more advanced examinations following the enhanced CT scan. Bronchoscopy reveals diffuse erythema of the mucosa at the entrance to the bronchus basal branch of the right lower lobe and endobronchial ultrasound finds abundant blood vessels in the right hilar mass (Fig. 2).

* Corresponding author.
E-mail address: sunruilin213@126.com (R. Sun).

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Fig. 1. Contrast-enhanced CT showed that soft tissue mass-like lesions in right mediastinal and hilum of the lung with multiple tortuous vessel shadows (A); narrowed right pulmonary trunk (B); smooth left atrial wall with the absence of the right pulmonary vein (C); normal pulmonary venous drainage on the left side (D).

Fig. 2. Bronchoscopy revealed mucosal normal on the trachea and left side (A + B), while diffuse erythema of the mucosa at the entrance to the bronchus basal branch of the right lower lobe (C), and bled on contact with the bronchoscope (D).
Trans-thoracic echocardiography revealed mild pulmonary hypertension with no signs. Through the literature search, combined with right pulmonary vascular abnormality, pulmonary hypertension, repeated pleural effusion, and hilar mass, it was speculated the patient may have UAPV. So, we further improved the diagnostic significance of the test - Digital Subtraction Angiography, and the outcome confirms the absence of right pulmonary veins (Fig. 3).

After the diagnosis was finally clear, a multidisciplinary team consultation was held to discuss the best treatment strategy. A conservative management with close follow up was adopted based on her mild symptoms.

3. Outcome and follow-up

The patient is currently on conservative management and follow-up.

4. Discussion

The presence of unilateral, isolated pulmonary vein atresia is an extremely rare congenital anomaly. It could occur in either lung, with no right- or left-sided predominance, and usually being diagnosed in infants [10]. The anomaly likely relates to the abnormal embryologic development of the pulmonary veins [3]. Prior to the development of pulmonary veins, the primordial lung buds, surrounded by the pulmonary vein plexus, drain through splanchnic venous plexus to the cardinal veins or vitelline veins, which will develop to systemic veins. Coinciding with development, the primitive connections between the pulmonary vein plexus and the systemic veins degenerated [4]. At birth, the common pulmonary vein has been absorbed into the posterior wall of the left atrium with the growth of atrium. It is considered that UPVA is due to the disconnection of the connection between one side of the common pulmonary vein and the left atrium after the degradation of the connection between the pulmonary vein and other venous systems.

The symptoms and signs of UPVA lack specificity. The main clinical manifestations of UPVA are recurrent pneumonia and hemoptysis in children and infants [5,10]. In our case, the patient presented 3 months of dyspnea because of repeated pleural effusion, without congenital heart disease or other previous history similar to classic symptoms of UPVA. Bronchoscopy finds mucosal hyperemia in the right bronchial tree and bled on contact with the bronchoscope (Fig. 4) which is likely to be related to the high capillary blood pressure and congestion due to UPVA. This is consistent with the result of bronchoscopy reported by Tissot C et al. [6].

The pathology of UPVA was the compensatory opening of mediastinal collateral veins, leading to the high pressure in peripheral vein and venule. Because of the increased pressure, excessive tissue fluid was formed and squeezed into lymphatic vessels, resulting in lymphatic dilatation and interstitial edema and cell infiltration. With the infiltration of fibroblasts, the pulmonary stroma is gradually transformed into fibrous tissue, which destroys the original structure and its elasticity. The small bronchi in the affected area are stretched and twisted, resulting in lumen dilatation or stenosis, while distal terminal bronchioles are altered as bronchiectasis or partial compensatory dilatation, or fusing into larger vesicles [4]. MRI reveals that the mediastinal soft tissue mass is the substitute vascular system in patients with UPVA [10]. In our case, the contrast-enhanced CT also reveals that the mediastinal soft tissue was significantly enhanced in the venous phase, and the degree of enhancement was similar to that of the vein (Fig. 1); ultrasonic bronchoscopy showed that many blood vessels present in right hilar soft tissue.

Pleural effusion is a common symptom in adult patients, which commonly caused by tuberculosis, bronchogenic carcinoma or pneumonia. Diagnostic tests for pleural effusions frequently includes chest radiograph, chest CT, bronchoscopy and thoracoscopy. Although pleural effusion caused by UPVA is rare, such a possibility should not be ignored. Confirmation of UPVA requires CT angiography or digital subtraction angiography [7]. In this case, the Digital Subtraction Angiography revealed that right bronchial artery-pulmonary artery fistula, narrow right pulmonary artery and the occluded right pulmonary vein. And the fistula was treated by transcatheter right bronchial artery embolism.
The management of UPVA remains controversial. Early diagnosis during infancy allows anatomical surgical correction to prevent all complications at the beginning. It would be very important to identify these patients before pulmonary hypertension or massive hemoptysis that greatly reduces patient prognosis ensues. Unfortunately, when the diagnosis occurs in adulthood, acquired problems make the anatomical correction impossible. Pneumonectomy is one of best choice when recurrent infections or significant hemoptysis become frequent and life-threatening [8,9]. On the other hand, as far as we considered, for adult patient with mild symptoms, like in this case, a conservative treatment may be enough or even be better systemically speaking.

In conclusion, the treatment regimen for UPVA depends on the age at diagnosis and the severity of the clinical manifestations. The number of pulmonary veins affected is positively correlated with the severity of the clinical manifestations and this correlation can be reflected in chestCT imaging. Severely progressive pulmonary hypertension is more appropriate for lung transplantation than for anatomical surgical correction, which may be the first choice.

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