CASE REPORT

Unilateral pulmonary vein atresia: diagnostic dilemma unfolded on imaging

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SUMMARY

Unilateral pulmonary vein atresia is a rare entity, usually congenital in origin. It is thought to result from failure of incorporation of common pulmonary vein to left atrium. Patients often present with recurrent chest infections and haemoptysis during infancy or early childhood. Associated anomalies are commonly present in these cases. Pulmonary angiography is generally used for definitive diagnosis; however, characteristic imaging findings on latest multislice CT can be virtually diagnostic.

BACKGROUND

Unilateral pulmonary vein atresia (UPVA) is a fatal condition of early childhood and is rarely diagnosed in adults. It is characterised by complete unilateral obliteration of the luminal continuity of pulmonary venous circulation. It can be isolated or may be seen associated with other congenital heart defects in approximately 50% of patients.¹ It is usually congenital in origin and should be differentiated from acquired causes of pulmonary venous obstruction such as fibrosing mediastinitis, mediastinal tumours and constrictive pericarditis.

CASE PRESENTATION

A 2-year-old male child presented with a history of recurrent pneumonia since 2–3 months of age. On admission, the child was afebrile and had

age-appropriate vital signs. There was no cyanosis with an arterial oxygen tension of 97%.

INVESTIGATIONS

Chest radiograph (figure 1) showed reduced volume of the left lung with ipsilateral mediastinal shift and prominent pulmonary vascular markings on the right side. There were no obvious signs of pulmonary infection on radiograph. Echocardiography revealed atrial septal defect and small left pulmonary artery.

CT angiography (figure 2) images showed small left hemithorax, hypoplastic left pulmonary artery with ground-glass opacities and interlobular septal thickening in the left lung.

No bronchial obstruction was seen. Complete absence of pulmonary veins was seen on the left side with a smooth lateral border of the left atrium (figure 2). Normal pulmonary venous drainage into the left atrium was seen on the right side. Associated cardiovascular lesions including an atrial septal defect, unroofed coronary sinus and double superior vena cava were also seen.

Outcome and follow-up

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

DISCUSSION

UPVA is a rare abnormality which is postulated to result from a failure of incorporation of the common pulmonary vein into the left atrium.² It may occur in either lung, with no predilection for right or left lung. It is associated with other congenital cardiac anomalies in 50% of the cases, most common being ventricular septal defect and double superior vena cava.¹ Histologically, the affected veins show medial hypertrophy with significant intimal fibrosis and little or absence of inflammatory reaction.³ Consequently, collateral extrapulmonary vessels develop to partially drain the affected lung.

These cases generally present in infancy or childhood with only few cases reported in adulthood.² Recurrent respiratory tract infection and haemoptysis are the most common presenting symptoms.² The striking radiological abnormalities on CT include reduced volume of the affected lung, ipsilateral hypoplastic pulmonary artery which occurs due to inappropriate gaseous exchanges caused by alterations between ventilation and perfusion and eventually presents flow reversal towards the contralateral pulmonary artery.^{3 4} Other parenchymal

Figure 1 Chest radiograph shows small left hemithorax with increased right lung vascularity and prominent right hilum.

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Figure 2 (A–D) CT angiography images show small left hemithorax with small left pulmonary artery (arrow in A) and dilated main and right pulmonary artery. Ground-glass opacity with interlobular septal thickening is seen on the lung window images (B). Pulmonary veins were absent on the left side with a smooth lateral aspect of the left atrium (arrow in D) and normal pulmonary venous drainage (*) on the right side. Note is also made of the left superior vena cava (arrow in C) and the left atrial appendage (arrowhead in D).

findings include ground-glass haze or consolidation, septal thickening which occurs likely secondary to dilatation of lymphatics and bronchial veins. Later chronic pulmonary oedema along with pulmonary venous infarction may result in fibrosis of the lung. Preferential pulmonary artery perfusion to the contralateral side due to impaired growth of the affected pulmonary artery accounts for development of systemic to pulmonary artery collaterals in these cases.^{2,5} Hypertrophy of these collaterals is the postulated cause of haemoptysis in these patients.⁶

This entity in adults may present with diagnostic dilemma due to the presence of confluent mediastinal soft tissue which may suggest a fibrosing mediastinitis or a malignancy involving the hilar vessels causing small ipsilateral pulmonary artery and pulmonary vein obstruction. However, the presence of patent bronchus in small hemithorax should suggest a congenital anomaly.² The confluent soft tissue mass may represent to contain venous collaterals. Therapeutic options include superselective embolisation of the hypertrophied collateral vessels in a patient presenting with acute haemoptysis. Pourmoghadam *et al* indicate surgery as a first therapeutic option with pneumonectomy being the preferred option because it alleviates the nidus of infection and also relieves significant left-to-right shunt and the dead space causing exercise intolerance.³ Undiagnosed cases presenting later in the life are usually associated with pulmonary

hypertension.⁷ Unrelenting progression and development of severe pulmonary hypertension preclude surgery for which lung transplantation is preferred.

CONCLUSION

UPVA is a rare condition with a diagnostic challenge. Findings of small ipsilateral pulmonary artery, absence of corresponding pulmonary venous opacification and reduced ipsilateral lung volume on CT angiography images are virtually diagnostic in patients with recurrent unilateral pneumonias and haemoptysis.

Learning points

- ▶ The diagnosis and treatment of unilateral pulmonary vein atresia is clinically challenging.
- ▶ Pulmonary angiography provides a definite diagnosis, however CT angiography findings of a small ipsilateral pulmonary artery with the absence of venous opacification on venous phase imaging are virtually diagnostic.
- ▶ Pneumonectomy is the preferred treatment option as it alleviates the nidus of recurrent pneumonias or haemoptysis. Moreover, it will also relieve significant left-to-right shunt and the dead space causing exercise intolerance.

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