In Thorax

Lack of a pulmonary vein causing unilateral interlobular septal thickening

Hanping Wang,1 Li Zhang,1 Wei Liu,2 Kai-Feng Xu,1 Juhong Shi1

DESCRIPTION

A 24-year-old woman presented with an 8-year history of chest tightness and shortness of breath during intense exercise. Physical examination showed decreased expansion of the right rib cage and respiratory sounds in the right lung were quiet. A routine examination was unremarkable. There were no systemic symptoms, including fever, skin rash and arthralgia. The erythrocyte sedimentation rate and high sensitivity C-reactive protein levels were normal. Autoantibodies, including antinuclear antibodies, antinuclear antibodies and antineutrophil cytoplasmic antibodies, were negative.

Chest high-resolution CT showed a low right lung volume, markedly thickened interlobular septae (mainly in the right lower lung) and some ground-glass opacity. Bronchoscopy showed submucosal varices of the right bronchial tree, with markedly increased surface secretions and mucosal hypertrophy (figure 2). Echocardiography showed normal inner atrial and ventricular diameters. Additionally, ostia of the right superior pulmonary vein were unclear, and pulmonary arterial systolic pressure was normal. CT pulmonary angiography (axial image, figure 1B; coronal maximum intensity projection image, figure 1C) showed a thin right pulmonary artery (PA), with fewer and thinner branches than normal. Right pulmonary venous drainage was not visible. There were multiple, slender, tortuous collateral veins with reflux into the superior vena cava in the right hilum. Selective right PA angiography showed reversed contrast medium flow to the left PA and a decreased diameter of the right PA compared with the left PA (figure 3A). The right pulmonary vein was not visible during the venous phase (figure 3B).

A diagnosis of congenital unilateral pulmonary vein atresia (CUPVA) was made. The patient experienced shortness of breath only after intense exercise and there was no pulmonary infection or recurrent haemoptysis. She was closely monitored, and light-intensity physical activity was permitted.

Unilateral diffuse pulmonary parenchymal lesions are clinically rare. The causes of these lesions include...
focal chronic inflammatory diseases, such as bronchiectasis and gastro-oesophageal reflux, radiation pneumonitis and mechanical ventilation of a unilateral lung. Unilateral diffuse pulmonary parenchymal lesions have also been reported in diseases, such as Sjogren’s syndrome and systemic sclerosis. Additional rare causes of unilateral lesions include pulmonary circulatory diseases, such as proximal PA blockage (eg, PA sarcoma), venous thrombosis and unilateral pulmonary vein stenosis due to mediastinal fibrosis. In adult patients with CUPVA, long-term pulmonary circulation abnormalities can cause typical unilateral diffuse lung lesions. However, diffuse thickening of interlobular septae, decreased lung volume and a slender PA on the affected side can differentiate CUPVA from other causes of unilateral diffuse lung disease.

Bronchial varices is a rare bronchoscopic finding, but it is clinically important for establishing the diagnosis of CUPVA. Rupture of these dilated bronchial veins can cause mild to massive haemoptysis. Therefore, transbronchial biopsy should be avoided in this situation.

Pulmonary angiography is the gold standard for investigation of CUPVA. CT angiography is also helpful. Magnetic resonance can be useful because it can define anatomic hypoplasia of the right PA and the absence of right pulmonary veins, as well as a lack of any forward flow to the right lung. CUPVA should be considered in the differential diagnosis of unilateral diffuse thickening of interlobular septae.

The presence of concurrent bronchial varices as visualised by bronchoscopy could strongly suggest diagnosis of this condition.

Contributors HW and JS were involved in designing and drafting of the manuscript. All the authors equally contributed to the collection and interpretation of the data and approval of the final manuscript.

Competing interests None declared.

Patient consent Obtained

Provenance and peer review Not commissioned; externally peer reviewed.

Open Access This is an Open Access article distributed in accordance with the Creative Commons Attribution Non Commercial (CC BY-NC 4.0) license, which permits others to distribute, remix, adapt, build upon this work non-commercially, and license their derivative works on different terms, provided the original work is properly cited and the use is non-commercial. See: http://creativecommons.org/licenses/by-nc/4.0/

Open Access This is an Open Access article distributed in accordance with the Creative Commons Attribution Non Commercial license, which permits others to distribute, remix, adapt, build upon this work non-commercially, and license their derivative works on different terms, provided the original work is properly cited and the use is non-commercial. See: http://creativecommons.org/licenses/by-nc/4.0/

CUPVA should be considered in the differential diagnosis of unilateral diffuse thickening of interlobular septae.

REFERENCES
Lack of a pulmonary vein causing unilateral interlobular septal thickening

Hanping Wang, Li Zhang, Wei Liu, Kai-Feng Xu and Juhong Shi

Thorax 2018 73: 199-200 originally published online July 29, 2017
doi: 10.1136/thoraxjnl-2017-210338

Updated information and services can be found at:
http://thorax.bmj.com/content/73/2/199

These include:

References
This article cites 3 articles, 0 of which you can access for free at:
http://thorax.bmj.com/content/73/2/199#ref-list-1

Open Access
This is an Open Access article distributed in accordance with the Creative Commons Attribution Non Commercial (CC BY-NC 4.0) license, which permits others to distribute, remix, adapt, build upon this work non-commercially, and license their derivative works on different terms, provided the original work is properly cited and the use is non-commercial. See: http://creativecommons.org/licenses/by-nc/4.0/

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Topic Collections
Articles on similar topics can be found in the following collections
- Open access (272)
- Thorax Images in Thorax (166)

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/