CASE OF THE MONTH

Something's missing in the chest

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Received 21 July 2009
Revised 25 November 2009
Accepted 26 November 2009

DOI: 10.1259/bjr/58400844

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A 44-year old man presented with symptoms of a productive cough, night sweats and worsening breathlessness on exertion. This was associated with intermittent episodes of left-sided chest pain over several years, together with recurrent chest infections. He was a smoker of at least 10–15 pack years. No haemoptysis or significant weight loss was reported.

On clinical examination, the findings were grossly normal. Spirometry revealed a mild obstructive function but no overwhelming evidence to support a diagnosis of bronchial asthma. The patient's inflammatory markers were normal. Microbiology cultures were negative for infective aetiology, including tuberculosis. What did the chest radiograph (Figure 1) show?

The chest radiograph illustrated reduced volume of the right hemithorax with compensatory hyperinflation of the left lung. The left pulmonary artery was enlarged. Even when allowing for the slight rotational asymmetry, the right hilum was poorly identified. There was a generalised increase in vascular markings in the right lung, resulting in a hyperlucent left hemithorax. There was shift of the mediastinum to the right side.

The differential diagnosis at this stage for a unilateral hyperlucent lung could be categorised by various aetiologies:

- (1) increased pulmonary air space with decreased blood flow
 - large airway obstruction with air trapping or endobronchial obstruction
 - small airway obstruction such as Swyer–James–Macleod syndrome
 - pneumothorax in a supine patient;

Figure 1. Plain chest radiograph.

- (2) pulmonary vascular cause such as pulmonary artery agenesis, congenital lobar emphysema or central pulmonary embolism;
- (3) poor radiographic technique;
- (4) overlying chest wall defect.

Diagnosis

A definitive diagnosis at this stage was not forthcoming, hence the patient was referred to the lung multidisciplinary meeting where the resulting decision was to perform a staging CT scan of the thorax to help elucidate the cause. What did the CT (Figure 2) demonstrate?

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Figure 2. (a) Axial CT image of the thorax lung window. (b–d) Axial CT image of thorax mediastinal window.

CT confirmed volume loss of the right hemithorax with compensatory hyperinflation of the left (Figure 2a). No central mass lesion was identified. Unusually, there was complete absence of the right main pulmonary artery (Figure 2b). This was associated with hypertrophy of the right-sided intercostal vessels leading to inferior rib notching (Figure 2c,d). No cardiac abnormalities were seen and this was later confirmed by echocardiography.

Discussion

Isolated unilateral pulmonary artery agenesis (UPAA) has a non-specific presentation in adulthood [1]. Congenital absence of one pulmonary artery is rare, with an estimated prevalence of 1 in 200 000 [2]. It can occur as an isolated lesion or in combination with other cardiac or vascular lesions, *e.g.* tetralogy of Fallot or septal defects. Most patients with isolated UPAA may have delayed diagnosis into adulthood because of the benign nature of this anomaly.

The most common symptoms include recurrent pulmonary infections, reduced exercise tolerance and mild dyspnoea. Haemoptysis may arise from the presence of extensive collateral circulation; however, it is unusual for it to be the only symptom.

Right-sided UPAA is twice as common as left-sided UPAA [3]. The blood supply to the affected lung is maintained by the formation of collaterals arising from the bronchial arteries, major aortopulmonary collateral arteries, intercostals and transpleural vessels.

Cross-sectional imaging can help to clinch the diagnosis. In this case, a spiral CT scan through the entire thorax was the investigation of choice because the major concern was to exclude a central endobronchial lesion. However, if all the pertinent chest radiograph findings were identified, one could proceed to performing a limited CT scan involving acquisition of images around the carina only, thereby reducing the ionising radiation dose to the patient.

Conclusion

UPAA is a rare condition with typical non-specific symptoms. If the radiologist is aware of the entity of UPAA and correctly interprets the chest radiograph findings, the diagnosis can be suggested. This can then be subsequently confirmed by a limited CT scan and ultimately a lower ionising radiation dose for the patient.

References

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