PULMONARY PUZZLES

An unusual cause of haemoptysis in a smoker

M J McDonnell,1 J Garvey, D G Lohan,1,2 G J O’Sullivan,2 K Redmond,3 J E Jackson,4 R M Rutherford1

CASE DESCRIPTION

A 33-year-old Caucasian male smoker presented to our rapid access lung cancer clinic with a single episode of moderate haemoptysis and no other respiratory or systemic symptoms. There was no relevant past medical history. As an adopted child he was unaware of any significant family history. The only finding on clinical examination was decreased air entry on the right side. Laboratory tests were all normal. Chest x-ray demonstrated a small right hemithorax with marked mediastinal displacement to the right, increased soft tissue opacity adjacent to the right side of the mediastinum causing widening of the right paratracheal stripe and loss of silhouette of the right heart border, and increased interstitial opacity throughout the right lung (figure 1A). A CT scan of the thorax confirmed the presence of a large amount of abnormal low attenuation soft tissue around the right side of the mediastinum, which also involved the azygo-esophageal space causing considerable thickening of the esophageal wall (figure 1B), a small right hemithorax, bronchial and non-bronchial systemic artery hypertrophy and widespread interlobular septal thickening (figure 1C). No endobronchial lesion was noted on bronchoscopy but multiple superficial vessels were present in the trachea and entrance to the right main bronchus (figure 1D). Bronchial and systemic artery hypertrophy and interlobular septal thickening is consistent with a diagnosis of isolated congenital unilateral pulmonary vein atresia. Diagnosis is usually made in

DISCUSSION

The combination of a small right hemithorax, abnormal right paramediastinal soft tissue, bronchial and non-bronchial systemic artery hypertrophy and interlobular septal thickening is consistent with a diagnosis of isolated congenital unilateral pulmonary vein atresia.

This condition, without associated cardiac malformation, is very rare with fewer than 50 reported cases worldwide.1–3 Diagnosis is usually made in

Figure 1 (A) Chest x-ray showing a small right hemithorax with right paramediastinal soft tissue and interstitial shadowing in the right lung. (B) Axial CT image at the level of the right hemidiaphragm showing abnormal right paracardiac and paraesophageal soft tissue. Note also the enlarged right inferior phrenic artery coursing over the top of the liver. (C) Axial CT image through the lung bases showing widespread right-sided interlobular septal thickening which was present throughout the right lung. (D) Bronchoscopy shows multiple superficial vessels in the trachea and origin of the right main bronchus.

To cite: McDonnell MJ, Garvey J, Lohan DG, et al. Thorax Published Online First: [please include Day Month Year] doi:10.1136/thoraxjnl-2013-203476

Copyright Article author (or their employer) 2013. Produced by BMJ Publishing Group Ltd (& BTS) under licence.
infancy. To our knowledge, only five adult cases have been described (table 1).1–4 Recurrent pulmonary infections (affected side), exertional dyspnoea and haemoptysis are the predominant presenting complaints.23

Imaging features include:

▸ Bronchial and non-bronchial systemic artery hypertrophy which, through normal systemic artery to pulmonary artery (PA) anastomoses, causes reversal of blood flow within the ipsilateral PA. Figure 2A shows a hypertrophied right bronchial artery (arrowhead) and increased enhancement of the right PA (arrow) compared with the left. The reversal of flow is further confirmed by the complete absence of activity in the right lung on a technetium-99m macroaggregate albumin perfusion lung scan (figure 2B).

▸ Complete absence of ipsilateral central pulmonary veins with a smooth contour to the right side of the left atrium (figure 3).

▸ Ipsilateral interlobular septal thickening due to severe pulmonary venous outflow obstruction (figure 1C).

▸ Abnormal paramediastinal soft tissue which is largely made up of venous collaterals.2

Pulmonary vein atresia is caused by late failure of incorporation of the common pulmonary vein into the left atrium.4 Mortality approaches 50% if untreated.1 3 Management may be conservative. Outcome data from attempted restorative surgery are poor.3 4 Pneumonectomy, if possible, is the preferred treatment choice to avoid life-threatening haemoptysis.1–3

Contributors All authors have made substantial contributions to the conception and design of the study or acquisition and interpretation of imaging data; drafting the article or revising it critically for important intellectual content; and final approval of the version to be submitted. RMR is responsible for the overall content as guarantor.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES
An unusual cause of haemoptysis in a smoker

M J McDonnell, J Garvey, D G Lohan, G J O'Sullivan, K Redmond, J E Jackson and R M Rutherford

Thorax published online May 24, 2013

Updated information and services can be found at:
http://thorax.bmj.com/content/early/2013/05/23/thoraxjnl-2013-203476

These include:

References
This article cites 5 articles, 0 of which you can access for free at:
http://thorax.bmj.com/content/early/2013/05/23/thoraxjnl-2013-203476#BIBL

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

Hemoptysis (80)
Radiology (diagnostics) (812)
Screening (oncology) (407)
Cardiothoracic surgery (676)
Health education (1223)
Smoking (1037)
Tobacco use (1039)
Health effects of tobacco use (211)
Lung infection (97)
Lung cancer (oncology) (670)
Lung cancer (respiratory medicine) (670)
Lung neoplasms (608)
TB and other respiratory infections (1273)
Child health (843)
Tobacco use (youth) (191)

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/