

Case report

Fatal haemorrhage from Dieulafoy's disease of the bronchus

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Abstract

A 70 year old woman with a previous history of healed tuberculosis and suspected chronic obstructive pulmonary disease presented with recurrent haemoptysis and respiratory failure from a lobar pneumonia. Massive bleeding occurred when biopsy specimens were taken during bronchoscopy which was managed conservatively, but later there was a fatal rebleed from the same site. Two different Dieulafoy's vascular malformations were found in the bronchial tree at necropsy, one of which was the biopsied lesion in the left upper lobe. This report confirms the possibility that vascular lesions occur in the bronchial tree. It is suggested that, if such lesions are suspected at bronchoscopy, bronchial and pulmonary arteriography with possible embolotherapy should be performed.

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Keywords: Dieulafoy's disease; bronchial vascular lesion; fatal haemorrhage

A 70 year old woman presented with respiratory failure, apparently due to acute lobar pneumonia of the right upper lobe, and was admitted to the intensive care unit after conservative

management had failed. She required intubation and mechanical ventilation. She had a 60 pack year history of smoking and had suffered tuberculosis 45 years earlier but information about treatment given at the time was not available. She had had at least two significant episodes of haemoptysis during the last few months which recurred after admission. On examination there were signs of rhonchi and reduced breath sounds, with few crepitations over the right upper lung field, but no bronchial breathing was noticed. The chest radiograph showed consolidation over the right upper lobe but was otherwise normal. Bronchoscopic examination showed changes compatible with chronic obstructive pulmonary disease; there were hypertrophic mucosal glands with increased collapsibility of the posterior wall of the tracheobronchial tree during coughing. There was no endobronchial abnormality in the right upper lobe ostium, and bronchoalveolar lavage did not show pathogenic micro-organisms other than *Streptococcus pneumoniae*. Ziehl-Neelsen staining and polymerase chain reaction for IS 6110 were negative. In the upper division of the left lobe at the lateral wall a smooth elevated non-pulsating lesion, approximately 8 mm in length, with normal overlying mucosa was noticed. The lesion was believed to be compatible with submucosal tumour and therefore a biopsy sample was taken. Massive bleeding occurred and the tracheal tube was therefore positioned in the right main bronchus and guided over the flexible bronchoscope. No dramatic fall in haemoglobin or blood pressure was noticed and two days later the patient was weaned from the ventilator. Residual atelectasis and blood clots were seen in the distal left main bronchus at repeat bronchoscopy with no evidence of active bleeding at that time. She was transferred to the ward for vascular imaging with possible embolisation and further assessment of pulmonary function. Histopathological examination revealed that the biopsy specimen indeed contained vascular wall, probably a branch of the pulmonary artery. Twelve days after the first bronchoscopy, before the planned tests could be completed, and after a mild rebleed two days earlier from the left upper lobe, she had a massive fatal haemoptysis and attempts at resuscitation failed.

Necropsic examination of the left upper lobe showed a mucosal thickening 5 mm in

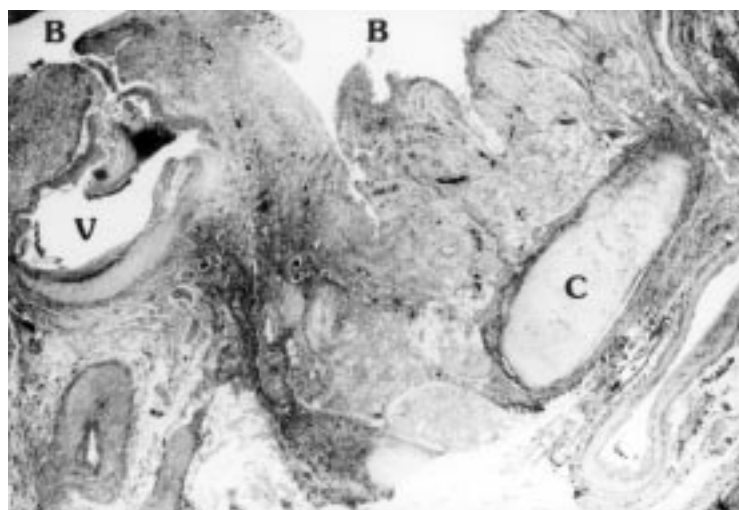


Figure 1 Overview of necropsy specimen of left main bronchus showing bronchial cartilage on the right (C) with a large vascular structure on the left (V) ending in the lumen of the bronchus (B). Magnification $\times 16$.

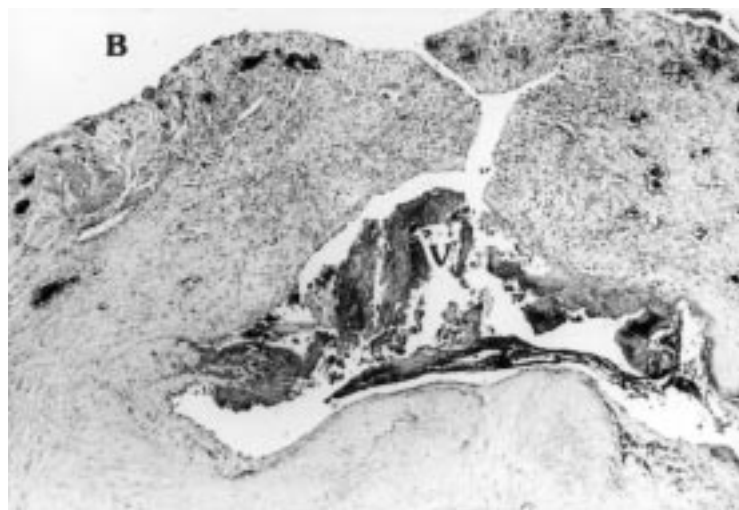


Figure 2 The vessel (V) appears to be in continuity with the bronchial lumen (B). Magnification $\times 40$.

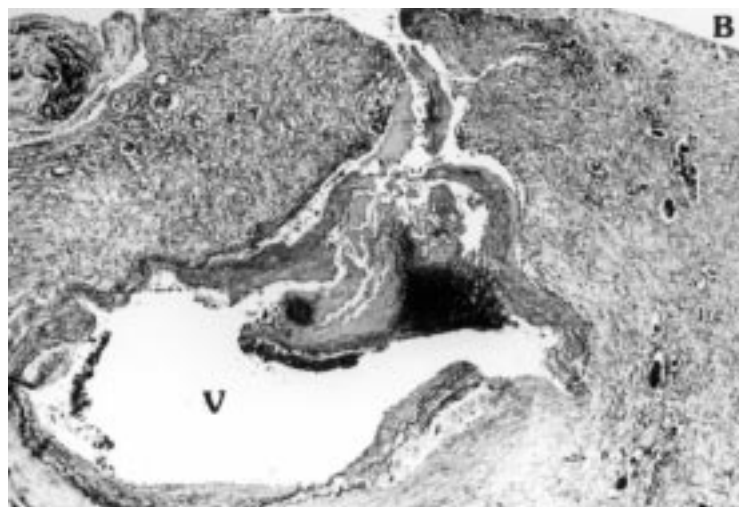


Figure 3 Biopsy specimen taken from the site where the vessel appears to be in continuity with the bronchial lumen showing detail of the blood vessel (presumably the pulmonary artery branch) just beneath the bronchial mucosa. Magnification $\times 40$.

cross section with a vessel of 1 mm cross section visible in the centre. Microscopic examination showed that a branch of the pulmonary artery appeared to pass between the bronchial cartilage rings, just below the bronchial mucosa, ending in the bronchial lumen (figs 1–3). A second similar vascular lesion was found in bronchial tissue blindly sampled from the right lower lobe. There was a varying degree of intimal fibrosis and mucoid degeneration of the medial layer, suggesting some degree of pulmonary hypertension. Bronchial changes suggestive of chronic obstructive pulmonary disease were present but there were no features to suggest emphysema. Both the left and right ventricular wall showed features of hypertrophy. Slight residual changes in the right upper lobe compatible with healing pneumonia were seen. There were no features consistent with tuberculosis or malignancy.

Discussion

This report, which follows an earlier report of two cases,¹ confirms that Dieulafoy's disease may occur in the bronchial tree. Dieulafoy's

disease was first described as a lesion in the stomach,² but it appears to occur throughout the digestive tract.³ Although this is the second report of Dieulafoy's vascular anomaly in the bronchial tree, we suspect that its occurrence is underdiagnosed rather than being very rare.

As this report shows, fatal haemorrhage cannot be precluded even in intubated patients in the ICU if biopsy samples of endobronchial vascular lesions are taken. Initial salvage by advancing the endotracheal tube into the contralateral lung, thus controlling bleeding by inflating the cuff over the bleeding main bronchus and securing a patent airway to the contralateral lung for gas exchange, could not prevent a fatal outcome. We therefore stress the potential danger of taking a biopsy sample of a vascular lesion at bronchoscopy. If such a lesion is either recognised or suspected, pulmonary and bronchial angiography should first be undertaken.

Our patient presented with respiratory failure because of pneumonia in the right upper lobe and, presumably, pre-existing chronic obstructive pulmonary disease; resection of the left upper lobe was clearly not an option at the time of bleeding. The planned treatment was embolisation if the source of bleeding had been the bronchial arterial system. However, histopathological examination of the biopsy specimen and the necroscopic specimen suggested an anomaly of a branch of the pulmonary artery. Sweerts *et al* do not state whether the anomalous artery reported in their patient was bronchial or pulmonary¹ but, interestingly, initial treatment of bronchial artery embolisation failed. The origin of the anomalous vessel may not therefore be a branch of the bronchial artery but, as in the Rendu-Osler-Weber syndrome, bleeds may have occurred from the pulmonary artery system. Embolotherapy has recently been recommended for these lesions.⁴ Although Sweerts *et al* argue in favour of embolotherapy,¹ both patients were only cured after resection of the affected lobe(s). The presence of pulmonary hypertension was not documented during their stay in intensive care, nor had systemic hypertension ever been noticed during life.

We therefore recommend vascular imaging if vascular lesions are suspected in the bronchial tree as taking biopsy specimens may result in severe and, indeed, fatal haemorrhage. If vascular lesions are diagnosed, embolotherapy or resection can then be planned, thus avoiding hazardous and unnecessary biopsy procedures.

1 Sweerts M, Nicholson AG, Goldstraw P, *et al*. Dieulafoy's disease of the bronchus. *Thorax* 1995;50:697–8.

2 Dieulafoy G. Exulceratio simplex. L'intervention chirurgicale dans les hématemèses foudroyantes consécutives à l'exulcération simple de l'estomac. *Bull Acad Med* 1898;49:49–84.

3 Dy NM, Gostaut CJ, Balm RK. Bleeding from the endoscopically identified Dieulafoy lesion of the proximal small intestine and colon. *Am J Gastroenterol* 1995;90:108–11.

4 Guttmacher AE, Marchuk DA, White Jr RI. Hereditary hemorrhagic telangiectasia. *N Engl J Med* 1995;333:918–24.