Thorax 1988;**43**:214–215

Bronchoscopic findings in hemitruncus

U ROSA, K C WADE

From the Department of Medicine, Pulmonary Section, University of Missouri, Kansas City, School of Medicine, Kansas City, Missouri, USA

A right pulmonary artery originating from the ascending aorta was first reported by Fraentzel in 1868.[1] Including our own, only six cases of this anomaly have been described in adults.[1-5] The condition is more often recognised early in life, because the large left to right shunt rapidly leads to biventricular failure.[3 6 9] Patients living beyond infancy experience frequent respiratory complaints, such as dyspnoea on exertion, episodic wheezing, and frequent airway infections.[1-4] Later haemoptysis will overshadow these symptoms. Haemoptysis first occurred in the age range 15–23 years in the reported cases.[2-4] It characteristically follows exertion and the expectorated blood varies from minor amounts to as much as 200 ml. The true nature of the cause of bleeding is usually not apparent from physical examination or review of standard chest radiographs. Adults with this malformation are thus likely to undergo bronchoscopy for recurrent haemoptysis. The importance of this is well illustrated by the following case.

Case report

The patient, a 26 year old man, underwent fibreoptic bronchoscopy after coughing up about 200 ml of bright red blood after exertion. He had had recurrent haemoptysis since the age of 23, usually associated with heavy activity. At the age of 24 he underwent bronchoscopy for haemoptysis. Bleeding appeared to originate from the right upper lobe and no apparent cause was identified. His childhood has been marked by frequent respiratory infections and intermittent wheezing. His parents were told that he had asthma. He underwent repair of a patent ductus arteriosus at 9 years of age. Orthopnoea and dyspnoea during moderate to heavy exertion became apparent after the age 21 and he had an episode of heart failure during service in the Army.

At fibreoptic bronchoscopy a tracheal diverticulum 0·8 cm wide and 0·5 cm deep was found, originating from the distal right lateral wall of the trachea (fig 1). Several centimetres lower there was extrinsic pulsatile compression of the anterior walls of the trachea and the right main stem bronchus to a slit like orifice (fig 2). Pulsations were strong and synchronous with the heart beat. Mucosal blood vessels appeared engorged and were most prominent in the right upper lobe, where clotted blood protruded from the posterior segment.

Address for correspondence: Dr Ute Rosa, UMKC School of Medicine, 4 Green Unit, 2411 Holmes, Kansas City, Missouri 64108, USA.

Accepted 4 May 1987

The bronchoscopy findings strongly suggested a congenital vascular malformation and a diagnosis of pulmonary artery sling was initially suspected. Subsequent studies included computed tomography of the chest as well as pulmonary and aortic angiography, all of which established the diagnosis of hemitruncus. Haemodynamic measurements indicated mild to moderate hypertension in the main and left pulmonary arteries. Pressures in the right pulmonary artery equalled those in the aorta. The patient later had open lung biopsies and a successful surgical repair.

Discussion

The bronchoscopic findings in vascular anomalies of the large mediastinal vessels are scarcely documented in published reports. Most descriptions concern pulmonary artery sling, a malformation in which the left pulmonary artery originates from the right pulmonary artery and wraps itself around the right lateral and posterior walls of the trachea, anterior to the oesophagus. It causes pulsatile

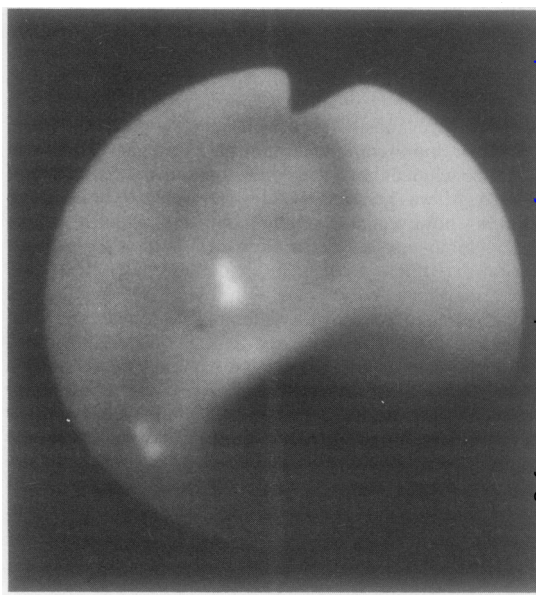

Fig 1 *Close up view of a 0·8 cm wide and 0·5 cm deep diverticulum of the right lateral tracheal wall. The carina it forms with the trachea strongly suggests an aborted bronchus. The arrow points to the aborted bronchus; the bottom of the print shows the anterolateral wall of the trachea.*

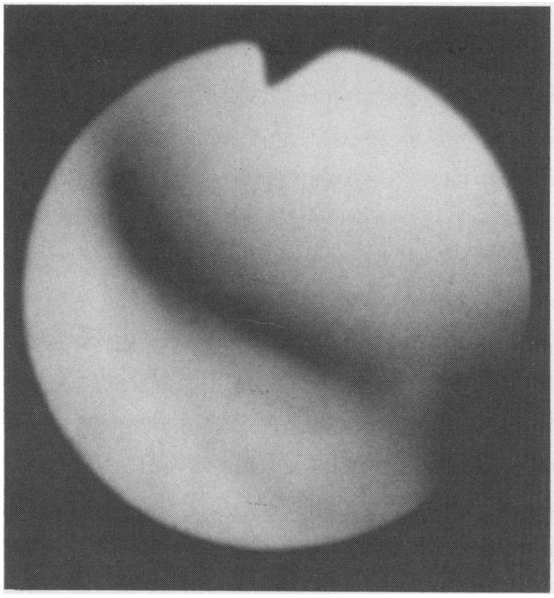

Fig 2 *View of the right main stem bronchus orifice at the level of the carina. The right side of the distal trachea and the right main stem bronchus are reduced to a slit like opening by a strongly pulsatile structure exerting extrinsic compression on the anterior wall of the airway. The arrow points to the right side of the posterior wall of the trachea; the anterior wall of the trachea faces the bottom of the picture.*

compression of the superior wall of the right main stem bronchus and of the right lateral and posterior walls of the trachea near the carina. The indentation of the anterior wall of the oesophagus by the anomalous artery is pathognomonic of this condition.[7 8]

In anomalies of the innominate artery the vessel compresses the anterior wall of the intrathoracic trachea above the aortic arch. Stenosis may be severe, the tracheal lumen being reduced to a pulsatile transverse slit.[9] The left anterior arch of a double aortic arch will considerably compress the distal third of the trachea, imparting a characteristic triangular shape to its residual lumen. The base of this triangle is formed posteriorly by the pars membranacea, while the strongly pulsating cartilaginous walls converge to form the anterior apex of the triangle.[9]

Occasionally tracheal compression is caused by the left subclavian artery or by a left ligamentum arteriosum associated with a right aortic arch. In the former there is pulsatile displacement of the anterior wall of the trachea similar to that accompanying an anomalous innominate artery. The left ligamentum arteriosum causes no pulsations. It indents the left tracheal and oesophageal walls at variable levels of the aortic arch.

Ben-Shachar *et al* described the bronchoscopic findings in a 2 month old girl with hemitruncus.[10] The trachea was compressed anterolaterally just above the carina, reducing the luminal diameter to a quarter of normal. The findings in our patient were essentially similar. Strongly pulsatile extrinsic compression of the anterior walls of the distal trachea and right main stem bronchus reduced the lumen of these structures to about 15% of normal. While congenital malformations of the airways, such as complete cartilage ring and tracheal bronchi, frequently coexist with pulmonary vascular sling, such anomalies have not been reported with hemitruncus.[8 10] The right tracheal diverticulum found in our case strongly suggests an aborted tracheal bronchus. An association of a tracheal diverticulum with hemitruncus has not to our knowledge been described hitherto.

In conclusion, anomalies of the large mediastinal vessels can result in airway obstruction or haemoptysis. The pulsatile nature of the extrinsic compression and its location in the airway are important diagnostic clues.

References

1 Fraentzel O. Ein Fall von abnormer Communication der Aorta mit der Arteria pulmonalis. *Arch Pathol Anat* 1868;**43**:320–6.
2 Caro C, Lermanda VC, Lyons HA. Aortic origin of the right pulmonary artery. *Br Heart J* 1957;**19**:345–52.
3 Keane JF, Maltz D, Bernhard WF, Corwin RD, Nadas AS. Origin of one pulmonary artery from the ascending aorta. Diagnostic, physiological and surgical considerations. *Circulation* 1974; **50**:588–94.
4 Jucá ER, Carvalho W Jr, Ribeiro de Sousa J, *et al*. Origem anômala da artéria pulmonar direita na aorta ascendente. *Arg Bras Cardiol* 1979;**33**:348–50.
5 Huang T-Y. Sudden death secondary to anomalous origin of the right pulmonary artery from the aorta. *Indiana Med* 1985;**78**:1018–20.
6 Penkoske PA, Castañeda AR, Fyler DC, Van Praagh R. Origin of pulmonary artery branch from ascending aorta. Primary surgical repair in infancy. *J Thorac Cardiovasc Surg* 1983;**85**: 537–45.
7 Grover FL, Norton JB, Webb GE, Trinkle JK. Pulmonary sling. Case report and collective review. *J Thorac Cardiovasc Surg* 1975;**69**:295–300.
8 Bardon WE, Baker DH, Wung J-T, *et al*. Complete cartilage-ring tracheal stenosis associated with anomalous left pulmonary artery: the ring-sling complex. *Radiology* 1984;**152**:57–64.
9 Benjamin B. Endoscopy in congenital tracheal anomalies. *J Pediatr Surg* 1980;**15**:164–71.
10 Ben-Shachar G, Beder SD, Liebman J, Van Heekeren D. Hemitruncal sling: a newly recognised anomaly and its surgical correction. *J Thorac Cardiovasc Surg* 1985;**90**:146–8.