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Short reports

Ectopic right upper lobe bronchus as a cause of breathlessness

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Patients with upper airways obstruction may present with symptoms suggestive of asthma. An extrathoracic obstruction is usually made obvious by the presence of inspiratory stridor, but diagnostic difficulty may arise if the obstruction is longstanding or is intrathoracic. Additional clues may include failure of conventional treatment for asthma and characteristic abnormalities of pulmonary function. We report on a patient with noisy breathing from birth whose upper airways obstruction was not diagnosed until she was 13.

Case report

A 13 year old schoolgirl was referred with a three year history of "asthma" unresponsive to inhaled bronchodilators, sodium cromoglycate, beclomethasone, and oral ketotifen. There was no cough or sputum but she was alleged to have wheezed on numerous occasions and recently had been refusing to participate in games because of effort breathlessness. She was causing some concern to her school teachers and parents, who had begun to believe that she was faking her symptoms to avoid games. Her parents were unconvinced that she had asthma and maintained that her breathing had been noisy since birth. There was no atopy or family history of asthma.

At examination the girl's breathing was noisy at rest and increased with exercise owing to loud inspiratory breath sounds, best heard at the mouth. There was no wheeze when one listened to the chest, and expansion of the chest, percussion, and breath sounds were normal. The plain chest radiograph was normal and a Vitalograph showed an FEV₁ of 1.5 litres and a forced vital capacity (FVC) of 2.21 (predicted values 1.91 and 2.31). The spirogram trace was suggestive of main airways obstruction (fig 1). At bronchoscopy there appeared to be a widened carina at the distal end of the trachea with a right bronchial orifice opening at an acute angle and the left bronchus continuing in a vertical direction. A tracheobronchogram revealed abnormal anatomy with an ectopic right upper lobe bronchus coming off the trachea. The trachea below the right upper lobe bronchus was narrowed

to half the diameter of the main trachea and this narrow segment then divided into the right bronchus intermedius and the left main bronchus (fig 2).

Surgical reconstruction of the narrowed segment of tra-

Surgical reconstruction of the narrowed segment of trachea was considered, but the patient's symptoms were not thought to be severe enough to warrant this major operation.

Discussion

Breathlessness and wheeze are common features of upper airways obstruction.¹ The two symptoms are often more pronounced during exertion and may be made temporarily worse by the presence of secretions or mucosal oedema from bronchial infection. It is well recognised that upper airways obstruction may masquerade as asthma. Pulmonary function tests and in particular the flow-volume loop may be useful in detecting both variable and fixed upper airways obstruction.¹ The Vitalograph tracing may be helpful when there is fixed or intrathoracic main airways obstruction, the tracing typically showing a near straight line because of reduction to flow at the site of obstruction.²

Congenital anomalies of the right upper lobe are rare. An anomalous bronchus is classified as displaced or ectopic where a normal bronchus arises from an abnormal site, and supernumerary when the anomalous bronchus is additional to a normal right upper lobe bronchus. The case reported here is that of an ectopic right upper lobe bronchus. Sometimes the tracheal bronchus represents an anomalous origin

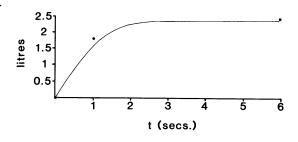


Fig 1 Spirogram showing that the initial expiratory phase is straighter than usual and suggestive of main airways obstruction. The closed dots refer to predicted figures.

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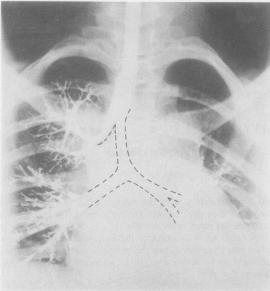


Fig 2 Bronchogram showing an ectopic right upper lobe bronchus arising from the trachea. The distal trachea is narrowed and divides into a left main and a right intermediate bronchus.

for a superior segmental bronchus of the right upper lobe.³

The condition is usually an unsuspected finding at bronchoscopy or bronchography carried out for the investigation of respiratory symptoms, the only clue on a plain radiograph being the presence of consolidation or bronchiectasis in the region of the right upper lobe. Ritsema⁴ reviewed about 800 bronchograms performed for the investigation of chronic bronchitis. He found eight patients with either ectopic or supernumerary right upper lobe bronchi, but only one patient had an ectopic lobar bronchus arising from the trachea. Two of the four supernumerary bronchi were bronchiectatic and one had surrounding consolidation. McLaughlin⁵ regard ported a series of 412 children with tracheobronchial abnormalities who had undergone bronchoscopy for the evaluation of stridor, recurrent pneumonia, suspected for eign body aspiration, and other reasons. Eighteen children were found to have a tracheal bronchus, of whom nine had an ectopic lobar bronchus. Five of the nine children had presented with recurrent pneumonia and had had recurrent infiltrates or persistent atelectasis of the right upper lobe. Only one of the nine had presented with stridor, but this was not a specific symptom as 47% of all the children studied had had bronchoscopy because of stridor.

In many cases the tracheal bronchus is morphologically normal and does not give rise to symptoms, but a narrowed anomalous bronchus may give rise to bronchiectasis or obe structive pneumonia. Distal narrowing of the trachea has not been commented on by previous authors and the absence of data on pulmonary function makes it difficult tooknow whether there was coexisting upper airways obstruction. In our patient drainage from the right upper lobe bronchus appeared to be satisfactory in that there was not evidence of recurrent infection. The breathlessness and stridor seem likely to have been due to the distal narrowing of the trachea rather than the tracheal bronchus itself.

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