PostScript

LETTERS TO THE EDITOR

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Exertional haemoptysis: LAM and TSC

Tuberous sclerosis (TSC) is characterised by the occurrence of hamartomas in different organs. It is autosomal dominant with complete penetrance and variable expression. TSC is associated with epilepsy, learning difficulties, behavioural problems, and renal and dermatological pathology. Lymphangioleiomyomatosis (LAM) is principally a pulmonary condition characterised by smooth muscle (*leiomyo*) proliferation around lymphatics (*lymph*), blood vessels (*angio*), and alveolar airways. Cystic destruction of lung parenchyma results in the development of pneumothoraces. 50% of patients with LAM have renal angiomyolipomas which are also the most common renal lesion in TSC.¹ LAM occurs in association with TSC or, less commonly, as a sporadic single entity.² It is almost exclusive to woman, usually of child-bearing age. The most common presentations are dyspnoea, pneumothorax, or chylothorax.^{1,2} LAM may be asymptomatic.

A 47 year old woman presented with a 9 month history of haemoptysis on exertion. Dyspnoea was not a feature. Haemoptysis was occurring with increasing regularity following

swimming and sexual intercourse, precluding both activities. The volume of blood was usually 10–20 ml. She had stopped smoking a year previously and had a 25 pack year history. There was a history of childhood epilepsy. There was no medical family history.

On examination the patient had a rash on the chin which reportedly bled following viral infections. There was a single subungual fibroma. Cardiovascular and respiratory examinations were normal. Pulmonary function tests showed normal lung volumes: FEV₁ 2.72 l, FVC 3.43 l, TLC 5.21 l, and RV 1.96 l with a corrected transfer factor of 73% predicted. Bronchoscopic examination revealed no source of bleeding. A high resolution CT scan of the thorax showed multiple cystic spaces with well defined walls and normal intervening lung (fig 1). A contrast CT scan of the head showed a single densely calcified subependymal nodule related to the right lateral ventricle. An abdominal CT scan identified multiple renal lesions bilaterally and a single hepatic lesion. Renal biopsy confirmed the presence of angiomyolipomas.

The above findings fulfil the criteria for a diagnosis of LAM and TSC.¹ In view of the diverse clinical course of LAM and the questionable value of hormone therapy, the patient was not commenced on treatment but referred for genetic screening.¹ This case underscores the need to consider such a diagnosis in female patients presenting with solitary exertional haemoptysis.

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References

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- 2 Hancock E, Tomkins S, Sampson J, et al. Lymphangioleiomyomatosis and tuberous sclerosis. *Respir Med* 2002;96:7–13.

Diaphragm plication following phrenic nerve injury

We read with great interest the paper by Simansky *et al*¹ describing the good results of plication of the diaphragm following phrenic nerve injury. The authors conclude that pulmonary function tests (PFTs) in combination with quantitative perfusion scans are helpful in selecting patients for this procedure. In table 4 they present the PFTs they were using and, in addition, they suggest that more sophisticated tests such as ultrasonography or fluoroscopy can also be useful in assessing diaphragmatic paralysis. Although we agree that all these tests are very helpful, assessment of vital capacity (VC) in both sitting and supine positions was omitted. This is a very simple test that gives important information about the function of the diaphragm, with a decrease in VC of >30% from the sitting to the supine position suggesting diaphragmatic paralysis.

The practical value of this test is clearly shown in the following patient in whom we

initiated non-invasive positive pressure ventilation (NIPPV) because of a right sided diaphragmatic paralysis due to a coronary bypass. At the start of NIPPV there was a gap between the VC in the sitting and supine positions of 0.8 l (30%; VC sitting 2.7 l, VC supine 1.9 l). We started NIPPV and the patient became less dyspnoeic and less tired. After 18 months the clinical situation was still improving, with an increase in VC both in the sitting and supine positions to 3.5 l and 2.8 l, respectively. After 36 months the gap between VC in the two positions had almost disappeared (3.6 l and 3.5 l, respectively). In addition, the radiograph of the thorax showed a downward shift and normalisation of the position of the right diaphragm. We therefore stopped NIPPV and after several weeks the patient slept well without ventilatory support. This case illustrates that the assessment of VC in both the sitting and supine positions can be very helpful in the diagnosis and follow up of patients with diaphragmatic paralysis.

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Reference

- 1 Simansky DA, Paley M, Refaely Y, et al. Diaphragm plication following phrenic nerve injury: a comparison of paediatric and adult patients. *Thorax* 2002;57:613–6.

Dysfunctional breathing in COPD

I was interested to read Dr Morgan's review of dysfunctional breathing in asthma in the 2002 Year in Review,¹ but the problem may be even greater in COPD.

Dr Morgan suggests that the problem may have serious consequences in terms of morbidity, but we have published indirect evidence of an association with mortality. In the 10 year follow up of the Darlington and Northallerton Asthma Study the odds ratio for the risk of dying in those who had no best function recorded was 2.5, equivalent to a risk of best function of 60% predicted.² Although failure to obtain best function was sometimes associated with steroid phobia, by far the most frequent cause was an inability to complete spirometric tests which is a sensitive indicator of dysfunctional breathing.

In non-clinical practice one sees large numbers of patients managed in primary care who have breathlessness attributed to COPD which may or may not exist objectively. By the time they are seen the subjects usually are genuinely breathless because of deconditioning. There is an urgent need to correct this under recognition of the problem. Perhaps a change in the approach to history taking might be helpful. Breathlessness is usually regarded not only as a symptom of COPD—which it may be—but also as a measure of disability due to physiological limitation—which it certainly is not in moderate airway obstruction. The prime measure of disability in chronic cardiorespiratory dysfunction is exercise limitation. If this is physiologically

Figure 1 Section from pulmonary high resolution CT scan showing multiple cystic regions with normal intervening lung parenchyma.