Idiopathic diaphragmatic weakness

A disorder that is easily overlooked

Paralysis of one hemidiaphragm is a relatively common clinical finding and can result from dysfunction of the phrenic nerve due to malignant infiltration, trauma at surgery, and neuropathies.^{1,2} Often, however, the finding is unexplained. Symptoms are not usually marked, unless the onset is acute. Bilateral diaphragm paralysis is much less common, but the symptoms can be severe.

Case history

A 38 year old man was referred to the Brompton Hospital with suspected bilateral paralysis of the diaphragm. His hobbies were scuba diving and water-skiing, and he had first noticed breathlessness while walking into water on holiday. He was too breathless to swim and had voluntarily stopped all water sports. He reported severe orthopnoea on lying flat and had started to use three pillows. He described no other respiratory symptoms and in particular no cough, sputum, or wheeze. He had smoked 20 cigarettes daily until five years previously. A fortnight before noticing breathlessness, he had had a pain in his left shoulder for a week, which had interfered with his sleep and had been only partially relieved by anti-inflammatory drugs. He recalled a similar pain affecting his right shoulder a year earlier.

Full physical examination showed no abnormality apart from severe orthopnoea associated with inward movement of the abdominal wall with inspiration when supine. Plain chest radiography showed small lungs with a little linear band shadowing at the left base, suggestive of subsegmental collapse above the hemidiaphragm, which was high (fig 1). The right hemidiaphragm was also raised, at the level of the fifth rib in the midclavicular line anteriorly. Electrocardiography gave normal results, and total white cell count; serum haemoglobin, urea, and electrolyte concentrations; and results of liver function tests were within normal limits. Radiological screening of the diaphragms at the referring hospital was reported to give normal results.

Full lung function tests showed that his lung volumes and carbon monoxide transfer factor were slightly reduced whereas his specific gas transfer coefficient (transfer factor corrected for lung volume) was at the upper limit of normal (table). His forced vital capacity fell by 39% on lying flat: from 3.8 l to 2.3 l (normal fall <20%). Measurement of blood gas tensions while he was sitting at rest and breathing air

FIG 1—Chest radiograph of man with diaphragmatic weakness. Some linear shadowing is visible at left base, suggesting subsegmental collapse. Hemidiaphragms are raised, but otherwise lung fields are clear

showed a mild reduction of partial pressure of oxygen at 10.8 kPa, with a partial pressure of carbon dioxide at 4.8 kPa and pH of 7.5.

Global respiratory muscle strength was assessed by measuring the pressure generated in the mouth during maximal inspiratory and expiratory efforts at residual volume and total lung capacity respectively.³ His maximal expiratory effort was normal at 21 kPa (144% of normal value) but his inspiratory effort was reduced at 5.7 kPa (56%). Global inspiratory strength measured with an oesophageal balloon catheter during a sniff was also low (4.7 kPa; 45%).⁴

Diaphragm strength was assessed by measuring transdiaphragmatic pressure (the difference in pressure measured between an oesophageal and a gastric balloon) during a sniff (1.0 kPa; 7% normal) (fig 2), when inhaling to total lung capacity (0 kPa, normal >2.5 kPa), and during a maximum inspiratory effort from residual volume (2.2 kPa, 20% normal).⁵ Finally, phrenic nerve and diaphragmatic function were assessed by transcutaneous electrical stimulation of these nerves in the neck.⁶ The latency between applying the stimulus and the appearance of a signal on a electromyogram (recorded from the costal diaphragm

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Results of lung function tests in man with idiopathic diaphragm weakness

	Normal range	Actual value measured	% Of mean normal value
Forced expiratory volume in one second (FEV ₁) (l)	3.14-4.81	3.07	77
Forced vital capacity (FVC) (l)	3.81-5.81	3.82	79
FEV ₁ /FVC (%)	69-92	80	
Peak expiratory flow (l/min)	445-680	529	94
Peak inspiratory flow (l/min)	380-490	325	75
Total lung capacity (TLC) (l)	5.83-8.13	4.99	71
Residual volume (RV) (l)	1.24-2.58	1.24	65
RV/TLC (%)	20-38	25	
Carbon monoxide transfer factor (mmol/min/kPa)	8.70-13.33	9.00	82
Lung volume accessible to single breath of helium (l)	5.3-7.39	4.36	69
Gas transfer coefficient (mmol/min/kPa/l)	1.26-2.15	2.06	121

FIG 2—Oesophageal, gastric, and transdiaphragmatic pressures measured during short, sharp voluntary sniffs in a normal subject (left) and in a patient with severe diaphragm weakness due to neuralgic amyotrophy (right)

with surface electrodes placed over the lower rib cage) is a measure of the nerve conduction time (normal <9.5 ms). Transdiaphragmatic pressure during stimulation (twitch) is a measure of the strength of the diaphragm that is independent of effort. In this patient, however, a skilled operator found no signal on electromyography or transdiaphragmatic twitch responses, indicating that there was no phrenic activity.

The effects of immersion were quantified by conducting spirometry in a swimming pool. His forced vital capacity was 3.6 l when the water was below his waist, 2.8 l when it was 7.5 cm below nipples, 2.2 when 5 cm above nipples, and 1.8 l when up to his neck. With water up to his neck he was extremely dyspnoeic with vigorous movements of the upper rib cage and accessory neck muscles.

The investigations confirmed severe weakness or paralysis of both hemidiaphragms, which we believe was caused by neuralgic amyotrophy. We were able to tell the patient the diagnosis and reassure him that the condition was unlikely to progress. The mechanical consequences of the weakness were explained so that he understood why he became breathless. He was advised to sleep in a semirecumbent position and not to resume water sports. Before referral he had been off work, mainly because of anxiety about his breathlessness. Since his condition was diagnosed he has been working full time and has come to terms with his disability.

Comment

Neuralgic amyotrophy was first described in 1943,⁷ and in 1948 it was reported in 136 young service staff, who developed severe shoulder pain of sudden onset, usually lasting several days or weeks, followed by long term weakness and wasting of the shoulder girdle or arm muscles, or both.⁸ Most of these patients were affected unilaterally, although in those affected bilaterally the condition tended to develop asymmetrically. Dysfunction of the phrenic nerve was not described. The aetiology of the syndrome is unknown, and although a viral infection of the cervical nerve roots is suspected,⁹⁻¹⁰ no candidate virus has been identified. Allergic mechanisms have been suggested as the shoulder girdle has been affected after injections with serum or vaccines.^{8,10} Fever and constitutional symptoms were absent in the 1948 series at the onset of muscle weakness.

More recently neuralgic amyotrophy has been reported in association with paralysis of the diaphragm, presumably due to the phrenic nerve being affected.^{1,2,9-11} Of the 13 patients affected, nine had unilateral paralysis and four bilateral paralysis. In seven cases the condition developed shortly after an upper respiratory tract infection. Our patient did not recall symptoms of a prodromal upper respiratory tract infection and there was no evidence of shoulder girdle weakness, but he did describe the typical shoulder pains and his condition seems to be a variant of this poorly defined syndrome.

In the past three years 12 men have been referred to the respiratory muscle laboratory with a similar clinical picture and weakness of the diaphragm. Each volunteered a history of shoulder pain that was closely followed by the onset of shortness of breath. In all patients the results of lung function tests showed a restrictive pattern with normal or raised specific transfer factor and normal daytime arterial blood gas concentrations. Other features of these 12 patients were weakness of shoulder or arm muscles (five patients), breathlessness during water sports (four); two separate prodromal episodes (three), and bilateral diaphragm paralysis (seven); all had a reduction in global inspiratory muscle and diaphragm strength but normal expiratory muscle strength. One patient had a

history compatible with neuralgic amyotrophy over 10 years' previously, when a plain chest radiograph taken shortly after he had developed right forearm weakness, showed a raised right hemidiaphragm. An elevated left hemidiaphragm was found during preoperative assessment for cardiac surgery. When studied in our laboratory, both his hemidiaphragms were severely weak and no electromyographic signals or twitches were obtained from the diaphragm.

The association of neuralgic amyotrophy with weakness of the diaphragm is rare. In a review of 800 000 admissions to a New York hospital over 25 years,¹² only 25 cases of flaccid paralysis of the diaphragm were identified. In eight cases the cause of the paralysis was unexplained. In four of these eight cases the condition was preceded by symptoms suggestive of an upper respiratory tract infection, and none of the subjects developed progressive neuromuscular problems when followed for up to eight years.

Discussion

MG: This patient was fortunate as he was referred promptly to his local respiratory physician, who was aware of the salient features of the condition. I have been referred several patients with diaphragm weakness in whom diagnosis has been delayed for long periods, 12 years in one case, and who have undergone many unhelpful invasive investigations.

TC: Observing paradoxical movement of the anterior abdominal wall with respiration when lying supine was important in this case. Is it sensitive and specific for the diagnosis?

MG: Patients with severe bilateral diaphragmatic paralysis (sniff transdiaphragmatic pressure less than 3 kPa) show this sign, together with a fall in forced vital capacity from standing to supine greater than 20%. Patients with lesser degrees of weakness, however, usually have normal abdominal wall movement, as do patients with hemidiaphragm paralysis.

KFC: How much can radiology help in diagnosing this condition?

MG: Plain chest radiographs in patients with bilateral diaphragm paralysis may be misinterpreted as normal or showing poor inspiration. Radiological screening of the diaphragm movement is usually characteristic in patients with unilateral diaphragm weakness but in bilateral paralysis screening may appear to give normal results for several reasons. As patients are reluctant to lie flat screening may be performed in an upright position, when the diaphragm moves passively downwards due to relaxation of abdominal muscles and the raised rib cage leads to a false impression of active descent. If the patient does agree to lie flat screening time may be limited by the induced breathlessness, the inspiratory efforts increase, and the rapid passive motion of the diaphragm may be misinterpreted as normal.

AD: I have noticed that an isolated raised hemidiaphragm is not uncommon on mass miniature radiography. Do you consider that neuralgic amyotrophy is the diagnosis in these patients?

MG: It is certainly possible, although the aetiology of neuralgic amyotrophy is unknown and the condition is not well defined. A history of shoulder symptoms after an upper respiratory tract infection without breathlessness might well be missed unless asked for directly.

DMD: The results of the lung function tests in this patient are typical of those found in patients with a chest wall or diaphragm problem, showing a restrictive ventilatory defect with slightly reduced lung volume and reduced transfer factor but increased specific transfer factor. His residual volume is reduced to a greater extent than his other lung volumes, which

could not be achieved voluntarily and fits with the secondary plate atelectasis on the chest radiograph. The reduction in forced vital capacity on immersing the legs and abdomen in water is interesting. It is primarily explained by upward movement and splinting of the diaphragm by the incompressible abdominal contents, which are pushed inwards and upwards by the pressure of water, presumably 3-4 kPa (fig 3).

Diaphragm paralysis in water

FIG 3—Effect of immersion in water on patient with severe diaphragm weakness. Hydrostatic pressure of water causes inward motion of the anterior abdominal wall and upward displacement of the diaphragm from the abdomen into the thorax. In healthy subjects diaphragm develops tone preventing upward displacement, and contraction during inspiration overcomes the hydrostatic pressure of water, lifting rib cage and displacing anterior abdominal wall outwards

However about 0.5 l blood would be displaced from the veins in the lower half of the body into the thoracic cage during the manoeuvre, leading to reduction in the vital capacity by this volume even in a normal subject.

MG: I agree, but redistribution of blood would account for less than 30% of the fall in this man's vital capacity on immersion, and presumably at least some of the extra blood would be distributed through the body by a healthy heart. We have reported breathlessness in water as a presenting symptom of this condition,¹³ but to my knowledge the fall in vital capacity on immersion has not previously been recorded in such patients. The fall is substantial and would be compatible with the severe breathlessness.

TC: What are the chances of recovery in patients with diaphragm weakness?

MG: The prognosis depends on the aetiology. If the phrenic nerve is sectioned or irreversibly damaged then no recovery is possible, but patients can learn to live remarkably normal lifestyles, adapting to their disability. If the cause is traumatic, for example, after surgery, substantial recovery may occur, although this is slow, taking one to two years or more. Most of the patients I have seen with neuralgic amyotrophy seem to recover only a little if at all, but in fact this man has noticed some improvement in his symptoms and his transdiaphragmatic pressures had improved by about 30% at follow up nine months later.

DG: Would you summarise the salient clinical features of a patient with severe diaphragm weakness?

MG: Such patients are breathless on exertion, and complain of orthopnoea if the weakness is severe. As with this patient, they may complain of breathlessness on going into water. The chest radiograph may show small lungs, but is often otherwise normal, but a fall in vital capacity when lying down from standing is characteristic. The diagnosis needs to be confirmed by measuring inspiratory and expiratory pressures and diaphragmatic strength in a specialised laboratory. Though diaphragm weakness is not common, it is important to recognise the condition in order to give specific advice to the patient, who can otherwise find the symptoms frightening and exacerbated by lack of a clear diagnosis.

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ANY QUESTIONS

What are the risks of addiction to opiates after patient controlled analgesia?

The term addiction has been replaced by dependence and was defined by the World Health Organisation in 1969 as: "A state of psychic and sometimes physical resulting from the interaction between person and drug characterised by behavioural and other responses that always include compulsion to take the drug on a continuous or periodic basis in order to experience its psychic effects and sometimes to avoid the discomfort of its absence."

It has been said that dependence is more likely with access and self administration,¹ a view supported by the relatively high percentage of doctors, dentists, and nurses who become dependent on opiates. But personality and social and environmental factors are also important.

Dependence is rare when opiates are used to treat acute pain.² Among nearly 12 000 patients followed up after inpatient treatment with strong opiates four cases of

dependence were found. Only one patient was considered to have major dependence. There is no evidence to suggest that the position is any different with patient controlled analgesia despite widespread use of this technique in the past few years, particularly in the United States. But continued vigilance is required.

Patient controlled analgesia is, in fact, a useful means of treating painful conditions in those dependent on opiates, with or without tolerance. Care must be taken to balance use and patients' statements and demeanour with reasonable estimates of usual requirements.³ It is also essential to secure the pump against tampering by the patient and visitors. —T H MADEJ, consultant anaesthetist, York

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