

An unusual case of diaphragm paralysis

Our patient was a 62-year-old retired accountant of previous good health. He was not taking any medications and had no family history of neurological disease. He was involved in a minor road traffic accident in which his car was shunted from behind. No immediate injuries were sustained; however, he presented to his general practitioner 4 days later with lumbar spine discomfort. He was prescribed a codeine–paracetamol combination pill and diclofenac. Immediately after taking the first dose of these medications he developed severe dizziness and marked vomiting, which culminated in mild haematemesis, indicative of a Mallory–Weiss tear. Subsequently he discovered that he was dyspnoeic lying flat. He presented to his local emergency department with symptoms including marked orthopnoea, and dyspnoea on water immersion past his costal margin and on bending forward.

Physical examination showed gross paradoxical abdominal motion and mild breathlessness when recumbent. Respiratory system, some left basal crackles; cardiovascular system, normal; abdomen, normal; neurology, normal.

Arterial blood gas measurements showed a P_{aO_2} of 11.1 kPa and a P_{aCO_2} of 5.1 kPa. A chest radiograph showed an elevated left hemidiaphragm with some atelectasis above it. A CT pulmonary angiogram excluded any diaphragmatic rupture or pulmonary embolus as an underlying cause of orthopnoea. His sniff nasal inspiratory pressure (SNIP) was measured at 22 cm H_2O . Detailed testing showed no response to bilateral anterior magnetic phrenic nerve stimulation or to right unilateral phrenic nerve stimulation, with a greatly reduced response to left-sided stimulation (twitch transdiaphragmatic pressure 1.3 cm H_2O ; normal >8 cm H_2O). Pulmonary function tests (performed in the erect position) were consistent with diaphragm paralysis,

showing a restrictive pattern with mildly reduced carbon monoxide gas transfer (TL_{CO} 82% predicted) which became supernormal when corrected for alveolar volume (K_{CO} 116% predicted). Forced vital capacity (FVC) was 64% predicted, with a forced expiratory volume in 1 s (FEV_1)/FVC ratio of 81% and stable over the course of 7 months follow-up. Clinic FVC testing in the erect position was 2.8 and 1.5 litres when supine (54% reduction). Upright cervical spine MRI revealed minor degenerative changes at C5/6 only. Nerve conduction studies (arms and legs) and needle electromyograph (EMG) examination of the upper limbs were normal.

QUESTIONS

What was the cause of this patient's diaphragm paralysis and what are the therapeutic options?

See page below for the answers

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ANSWER

From the questions on page above

The investigations demonstrated bilateral diaphragm paralysis following prolonged vomiting. The patient had been previously well, thus it seems most likely, given the temporal relationship, that the hyperemesis was causal to the subsequent diaphragmatic weakness.

It is interesting to note that the initial chest radiograph failed to demonstrate a right as well as left hemidiaphragm elevation given that the phrenic nerve stimulation studies demonstrate profound bilateral weakness. However, a previous study published by our laboratory¹ showed that chest radiographs have imperfect sensitivity and specificity for diagnosing diaphragm paralysis. Hence, formal respiratory muscle testing should be considered when diagnostic suspicion is high, even if the plain radiograph appears reassuring.

The pulmonary function tests were typical for diaphragm dysfunction. The FVC is understandably reduced due to a lack of diaphragmatic excursion with a concomitant fall in FEV₁, such that the FEV₁/FVC ratio is >80% (ie, a restrictive deficit). Additionally the gas transfer is also mildly reduced, but is supernormal when corrected for alveolar volume. This again is typical for diaphragm dysfunction in the setting of an extrapulmonary restrictive deficit, but would also be consistent with other extrapulmonary restrictive conditions such as scoliosis which were absent in our patient. This is because the underlying lung parenchyma is healthy, unlike, for example, a pneumonitis-induced restrictive deficit where gas transfer will be limited by the diseased lung parenchyma. Finally, a fall in vital capacity of >20%² consistent with this patient's orthopnoea is useful in suggesting a diagnosis of diaphragm weakness. This is because patients with diaphragm weakness cannot defend their thorax against their abdominal contents when supine, but do not need to do so when erect since the effect of gravity is to draw the viscera caudally. Similarly, when erect in water, patients with diaphragm weakness are unable to counter the inward and upward pressure exerted by water on the abdominal viscera. Indeed in a previous study we determined that the magnitude of change due to water immersion was not dissimilar to adopting the supine posture and was associated with an increase in respiratory rate and drive to breathe.³ Although not subjected, to our knowledge, to experimental study, the same mechanism may be presumed to be the cause of dyspnoea when the patient bends forward (classically either to tie their shoelaces or when exiting their car).

Hyperemesis has long been associated with barotrauma, especially to the oesophagus; indeed Boerhaave first described his eponymous syndrome of oesophageal perforation post-hyperemesis in 1724. Literature review revealed only one other case of vomiting and hemidiaphragm elevation.⁴ Their case, however, had no trigger for the vomiting and also had a myocardial infarction, raising the possibility that vomiting was in fact the initial manifestation of the myocardial infarction; this conjecture is supported somewhat by the rapid resolution of their case, whereas our patient sustained symptoms lasting 3 months.

The phrenic nerves—the sole motor supply to the diaphragm—have a particularly long course, making them vulnerable to damage along their tortuous 30–40 cm path.⁵ The precise embryogenesis of the diaphragm is still the subject of scholarly debate, but it is conventionally considered to have two portions, the costal and crural.⁶ The right and left muscles fuse to form a single sheet of muscle but retain separate innervation and control by their respective phrenic nerves.⁷

Learning points

1. Vomiting in healthy humans can generate sufficiently high pressures to cause phrenic nerve injury and consequent diaphragm weakness.
2. Management is usually initially conservative and should focus on rehabilitation. Our patient benefited from this approach and has almost returned to his previous quality of life.

During emesis there are two main phases—retching and expulsion. The retching phase is vital to generating pressure. In this phase the diaphragm works synchronously with the gastric muscles such as the rectus abdominus to build up intragastric pressure. The crural diaphragm is also at high tension and does not allow the passage of food cranially. In the expulsive phase, there is dissociation between the crural and costal portions and this allows relaxation of the crura around the lower oesophageal sphincter and the release of gastric pressure with concomitant expulsion of gastric contents upwards. Gastric pressure recording has been undertaken during vomiting⁷; the maximum pressure obtained (394 cm H₂O) was greater than was observed during coughing and considerably greater than observed in normal subjects undertaking a maximal voluntary cough.⁸ It thus seems plausible that vomiting can, at least in fit individuals, be associated with sufficiently high pressures to cause damage to intrathoracic structures. We were therefore not surprised on careful literature review also to find cases where vomiting was followed by pneumomediastinum,⁹ pneumopericardium,¹⁰ displacement of medically implanted devices¹¹ and arterial trauma,¹² as well as the more well known clinical entities of Boerhaaves syndrome and the Mallory–Weiss tear.

Therapeutic options may be considered as either conservative or operative. It is reported that there is at least a 20% chance of spontaneous recovery over the following 2 years with axonal regeneration.¹³ Therefore, an initial period of observation is usually warranted. For those where recovery is not complete, hemidiaphragmatic paralysis is not usually too troublesome and patients may benefit from taking exercise, pulmonary rehabilitation—specifically inspiratory muscle training¹⁴ and weight loss. For those who remain symptomatic, surgical interventions have been described with diaphragm plication, phrenic nerve repair when there has been surgical dissection¹³ and occasionally diaphragm pacing when the phrenic nerve is intact, but it is recommended that such treatments be employed only after specialist review.

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