CASE REPORT

A case of progressive orthopnoea and shoulder pain secondary to bilateral diaphragmatic paralysis

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SUMMARY

A 45-year-old man with a raised body mass index presented with an acute history of right lower chest pain and increasing breathlessness. C reactive protein, D dimer and cardiac echocardiography were negative, with mild bibasal atelectasis the only positive reported finding from erect chest X-ray and CT pulmonary angiogram. He was discharged with antibiotics for a chest infection. He remained severely breathless and was referred to the sleep-related breathing disorders clinic. He described shoulder pain, daytime tiredness and sleeping upright due to a "suffocating" sensation. The video demonstrates the clinical findings. The CT topogram confirmed bilateral diaphragmatic paralysis. Spirometry demonstrated an 80% reduction in forced vital capacity in the supine position, when compared with erect. Consultation with a neurologist yielded the diagnosis of neuralgic amyotrophy, leading to bilateral diaphragmatic paralysis. The respiratory symptoms have been controlled with night-time non-invasive ventilation, allowing him to sleep supine.

BACKGROUND

The diaphragm is the most important muscle for breathing. Contraction of the domes of the diaphragm creates a negative intrathoracic pressure, triggering the initiation of respiration. Diaphragmatic paralysis, of any cause, can lead to respiratory failure, which may require ventilator support.

Neuralgic amyotrophy (NA), as a cause of phrenic nerve palsy and unilateral or bilateral diaphragmatic paralysis, is described in a number of case reports. However, the diagnosis can often be delayed due to the common presenting symptoms such as dyspnoea, orthopnoea and shoulder pain, focusing investigations into more common cardiorespiratory pathology. This potential delay is well demonstrated in this case report, with the patient undergoing an array of tests aimed at investigating and treating heart failure, pneumonia and a pulmonary embolus. Ultimately the diagnosis was made when paradoxical movement of the diaphragm in the supine position was noted on clinical examination (see online supplementary video).

This series of events demonstrates the need for ongoing education. The diagnosis can easily be overlooked in favour of more common cardiorespiratory pathologies, despite a typical history and presentation of the disease. It is very important to consider the possibility of diaphragmatic weakness in this context, particularly when these cardiorespiratory

diseases have been excluded or there is a poor response to treatment. Furthermore, where imaging is available, the position of the diaphragm should be reviewed in the supine position. In this case the CT pulmonary angiogram (CTPA) topogram (figure 1) demonstrated bilateral raised diaphragms, which were not demonstrated on the erect chest X-ray (CXR) (figure 2).

This case is unique within the context of shoulder pain and bilateral phrenic nerve palsy as a manifestation of NA as it is the only case report demonstrating clinical signs and symptoms in video footage. The aim of this case report is to highlight the clinical presentation and classical examination findings of a patient with bilateral diaphragmatic palsy (BDP), in the hope that it will facilitate the timely diagnosis of BDP in such patients. The case report lends itself to be used as an accessible and understandable education modality for other healthcare professionals.

CASE PRESENTATION

A 45-year-old man presented to the hospital with left shoulder pain and shortness of breath, worse on lying flat and progressing over several weeks. Serum inflammatory markers were within normal range and an erect CXR demonstrated mild bibasal atelectasis. He was treated for both a lower respiratory tract infection and heart failure, based on the presumed symptom of orthopnoea and the CXR report. However, there was no significant response to antibiotic or diuretic therapy and so further investigations were undertaken. A transthoracic echocardiogram demonstrated good left ventricular function, D dimer was within normal range and a CTPA was negative for a pulmonary embolus but again reported mild bibasal atelectasis. The patient was discharged from the hospital and continued the course of oral antibiotics at home. A referral for overnight sleep studies was made on the basis of a raised body mass index and increasing difficulty sleeping at night due to dyspnoea.

This man was seen again in the sleep-related breathing disorders (SRBD) clinic following the overnight sleep study. His Epworth sleep score (ESS) at this time was only 2/24. The data from the sleep studies were suggestive of rapid eye movement (REM) sleep-related apnoea.

The sleep eight-channel polygraph result demonstrated that the patient slept upright for most of the night and only 11 min lying down with significant paradoxical chest and abdominal wall movements

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Figure 1 Chest X-ray (erect).

throughout the study. His apnoea-hypopnoea index (AHI) index was 4.3/hour.

At the clinic the man offered additional information on the history of events, stating that the symptoms started with sudden-onset shoulder and scapula pain, lower ribcage pain, increasing breathlessness and the need to sleep upright due to a feeling of suffocation when lying flat. At this point he admitted to undertaking the sleep study while sleeping upright in a chair. Lung function tests demonstrated an 82% reduction in forced vital capacity (FVC) from sitting to supine.

Clinical examination demonstrated paradoxical movement of the chest and abdominal wall during inspiration, suggestive of bilateral diaphragmatic weakness. On review of the CTPA topogram images from the previous hospital admission, there was demonstrably raised hemidiaphragms. This finding was not commented on in the CT report at the time.

The first part of the video clip (see online supplementary video) records the patient's account of the progression of his symptoms. The second part of the clip demonstrates the clinical examination features described above.

Consultation and investigation by a neurologist yielded the diagnosis of bilateral diaphragmatic paralysis secondary to neuralgic amyotrophy (postinfective). A negative test for

antiganglioside antibodies along with no proximal myopathy made a diagnosis of Pompe disease unlikely.

The respiratory symptoms have been controlled with nocturnal non-invasive ventilation (NIV), with arterial blood gases (ABG) demonstrating a reduction in carbon dioxide and bicarbonate retention, and static lung function test showing improvements in forced expiratory volume in 1 s (FEV_1) and FVC following initiation of NIV. The percentage change in sitting to supine FVC has reduced from 80% to 10%. Most importantly, the patient has had significant symptom relief and is now able to sleep lying flat. NIV has been continued long term and the improvement of the FVC shown above has been a gradual process.

INVESTIGATIONS

Transthoracic echocardiogram: normal left ventricular function

CXR: Bibasal atelectasis

CTPA: Bibasal atelectasis; bilateral raised hemidiaphragms on the topogram view (not reported)

ESS: 2/24

Pre-NIV ABG: pH 7.44, pCO_2 5.9, pO_2 9.1, HCO_3^- 29

Post-NIV ABG: pH 7.44, pCO_2 5.2, pO_2 7.9, HCO_3^- 25

Spirometry, April 2012, FVC, FEV_1

Sitting (S): 2.49/5.15, 1.73/4.16

Lying (L): 0.48/5.15, 0.36/4.16

% Reduction (S/L): 80%, 80%

Spirometry, January 2017

Sitting: 4.12/5.05, 2.86/4.04

Lying: 3.76/5.05, 2.5/4.04

% Reduction (S/L): 10%

DIFFERENTIAL DIAGNOSIS

1. Pulmonary embolism (PE)
2. Heart failure
3. Pneumonia
4. Anxiety and panic disorder
5. SRBD
6. Bilateral phrenic nerve palsy secondary to NA (postinfective).

TREATMENT

Nocturnal NIV.

OUTCOME AND FOLLOW-UP

From a respiratory perspective, follow-up has been with the NIV service. He has shown a steady improvement in static lung function and symptom control.

DISCUSSION

Amyotrophic neuralgia, also known as brachial neuralgia, brachial neuritis or Parsonage-Turner syndrome, was first described 70 years ago as a neuropathic disease of the brachial plexus. In later years, it was found to affect other motor nerves, including the phrenic nerve, due to inflammation of the C3–C5 nerve roots and can lead to unilateral or bilateral diaphragmatic paralysis.^{1–5}

NA is an idiopathic condition, often preceded by a viral or infectious precipitant. Case reports of bilateral phrenic nerve paralysis due to NA after herpes zoster infection and tetanus antitoxin, as well other toxic precipitants such as chemotherapy, have been described.¹⁴

Bilateral phrenic nerve paralysis is relatively rare. The most common causes for impairment in diaphragmatic function include neuromuscular disorders, trauma, iatrogenic conditions, tumour compression, infectious and inflammatory conditions,

Figure 2 CT topogram (supine) showing raised hemidiaphragms bilaterally.

NA and idiopathic phrenic nerve paralysis. Presenting symptoms will often include progressive dyspnoea, particularly when supine, and cases associated with NA can commonly feature shoulder pain as a preceding symptom. Ikegami *et al* described a case with worsening orthopnoea over a 6-month period presenting with shoulder pain, similar to our case.³

In clinical practice, patients presenting with orthopnoea to the acute medical take is commonplace, and the differential diagnosis often includes heart failure. However, although rarer, phrenic nerve dysfunction should be in the differential diagnosis in these cases.

A thorough history and clinical examination is imperative in all cases. The video footage of our patient clearly demonstrates paradoxical movement of the thorax and abdomen with respiration when moving from sitting to supine. Suggested investigations include blood gas analysis, lung function testing and CT imaging to review for tumours, including thymoma associated with myasthenia gravis. Pulmonary function tests are a useful screening tool in such cases. An FVC reduction from sitting to supine of

20% or more is highly suggestive of diaphragmatic weakness. This can be further supported with nerve conduction studies. Fluoroscopies with sniff tests are also useful in establishing phrenic nerve palsy. Bedside ultrasound is a useful modality in initial assessment and obviates the need for traditional fluoroscopy in most cases. In our case this was not performed as a CT scan had been performed on admission, which showed bilateral raised hemidiaphragms; hence, this was not required in our case. A negative test for antiganglioside antibodies along with no proximal myopathy made a diagnosis of Pompey's disease unlikely in our case.

Current treatment options for bilateral phrenic nerve palsy include respiratory muscle training, long-term NIV and surgical diaphragm plication in selected patients. NIV is indicated in patients developing type 2 respiratory failure but also can lead to an improvement in static lung function and symptoms of orthopnoea. Having said that there are cases that showed a poor response to NIV.^{4 5}

There are published case reports describing bilateral phrenic palsy secondary to NA. To the best of our knowledge, our case is unique in being the first video case report demonstrating the clinical features of bilateral phrenic palsy at the peak of the patient's symptoms. We feel that this is an invaluable aspect of our case and an excellent educational resource for other clinicians.

Contributors Planning, design, video footage, literature search and report were conducted by all authors: AH, WAK, SM. The overall report was based on the equal input of all the authors.

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Patient's perspective

'When I was started on the NIV machine it was first time in 3 months that I had a good night sleep. The choking sensation and inability to take a breath has dramatically improved'.

Learning points

- ▶ Bilateral phrenic nerve palsy (BDP) should be included in the differential of unexplained breathlessness and orthopnoea.
- ▶ Neuralgic amyotrophy commonly presents with shoulder pain. It is a cause of both unilateral and bilateral diaphragmatic paralysis due to phrenic nerve palsy.
- ▶ The diagnosis and the respiratory disability should be confirmed with radiological and physiological studies, including lying and standing forced vital capacity, fluoroscopy or bedside ultrasound and antiganglioside antibodies, to exclude Pompey's disease.
- ▶ Recovery from BDP secondary to neuralgic amyotrophy may be prolonged, and non-invasive ventilation is an effective supportive treatment option.

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