CASE REPORT

Sniffing out a hidden cause of breathlessness

Alexandra Marie Nanzer,¹ John Janssen,² Matthew Hind¹

¹Respiratory Department, Royal Brompton and Harefield NHS Foundation Trust, London, UK
²Royal Brompton and Harefield NHS Foundation Trust, London, UK

Correspondence to
 Dr Alexandra Marie Nanzer,
 A.Nanzer-Kelly@rbht.nhs.uk

Accepted 6 June 2014

SUMMARY

A 78-year-old man presented with severe exertional dyspnoea. He suffered from mild chronic obstructive pulmonary disease, congestive cardiac failure and seropositive myasthenia gravis. Clinical examination of his chest and heart were unremarkable but he had speech dyspnoea and was unable to count to 20 in a single breath. Consecutive sniff nasal inspiratory measurements (SNIP) fell from 55 to 33 cm H₂O and forced vital capacity (FVC) fell from 3.4 to 2.4 L. A diagnosis of myasthenic crisis was carried out and treatment with non-invasive ventilation, intravenous immunoglobulin and high-dose oral prednisolone was initiated. The patient responded well and was discharged following a short period of rehabilitation. A high index of suspicion and a careful clinical examination with the help of two simple bedside tests, FVC and SNIP, allowed correct and timely treatment of his condition.

BACKGROUND

Up to a fifth of all patients with myasthenia gravis can experience a myasthenic crisis as its initial presentation, or during the course of the disease. Although the majority of patients who present with myasthenic crisis (MC) have an identifiable precipitant such as infection, medication or surgery, in up to 40% no trigger is found and patients may present in a subtle and precipitous manner as illustrated in this case. Patients with marked bulbar weakness or low baseline vital capacities of <20–25 mL/kg are most at risk for respiratory failure; paradoxical breathing or supine dyspnoea are other warning signs.

CASE PRESENTATION

A 78-year-old ex-pilot reported a 4-month history of progressive exertional dyspnoea. On presentation he was comfortable at rest with no orthopnoea but described breathlessness on minimal movement and speech dyspnoea. There was no cough, fever or chest pain. A keen gardener, he had been unable to leave his chair for several weeks prior to presentation. There was a complex history of congestive cardiac failure, previous multiple pulmonary emboli and seropositive myasthenia gravis. He was an ex-smoker. Medication included warfarin, furosemide, spironolactone, ramipril, verapamil, pyridostigmine, alendronic acid and prednisolone 10 mg daily.

On examination he was euvolaemic with no evidence of cardiac failure, his chest was clear though he was unable to count to 20 in a single breath; and there was no abdominal paradox. Neurological examination was unremarkable.

INVESTIGATIONS

Laboratory results revealed a therapeutic International Normalised Ratio of 2.4 and low D-dimer. Previous contrast CT scans were reviewed and no major pulmonary emboli were seen that could account for his symptoms. We noted centrilobular emphysematous changes within the lung parenchyma. Pulmonary function tests revealed scalloping of the flow volume loop, transfer factor for carbon monoxide 65.2% predicted and K₊ channel opener 93.9% predicted. An echocardiogram was normal as were daytime arterial blood gases; the mean overnight transcutaneous carbon dioxide was 5.91 kPa. We asked the patient to perform consecutive measurements of sniff nasal inspiratory pressures (SNIP) and spirometry: SNIP pressures fell from 53 to 33 cm H₂O (normal >70 cm H₂O for men) and vital capacity from 3.4 to 2.4 L.

DIFFERENTIAL DIAGNOSIS

Our patient had several cardiovascular risk factors known to cause exertional breathlessness: valvular disease, systemic hypertension and left ventricular failure. However, there was no orthopnoea and a normal echocardiogram was reassuring. Chronic obstructive pulmonary disease and previous pulmonary embolisms were the only pre-existing pulmonary disorders. Lung function tests confirmed the former to be mild and a review of the CT scans confirmed patent vascular structures. Our patient was neither febrile nor hypoxic and he did not show signs of disordered breathing pattern suggestive of either a cerebral pathology or panic and anxiety disorders. What became most apparent was a distinct fatigability on minimal movement and speech dyspnoea pointing towards a respiratory muscle problem.

TREATMENT

We diagnosed MC, initiated respiratory support with non-invasive ventilation and started intravenous immunoglobulin. Pyridostigmine was stopped, prednisolone was increased to 50 mg daily and azathioprine was introduced. By the fourth day, he reported a dramatic response: he was able to count to 30 in a single breath, talk normally and walk 30 m; SNIP increased to 76 cm H₂O with no detectable fatigability; vital capacity was 3.4 L with no supine change. He was discharged following a short period of rehabilitation.

OUTCOME AND FOLLOW-UP

One week later, he reported feeling like 'a new man'; walking had improved, better than at any time over the preceding few months, and he was

CrossMark

To cite: Nanzer AM, Janssen J, Hind M. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-204996

able to tend his garden. SNIP measurements had increased further to 102 cm H₂O with no fatiguability. Prednisolone was slowly reduced and azathioprine continued.

DISCUSSION

Myasthenia gravis (MG) is a rare autoimmune disorder affecting an estimated 75 people/million.¹ Most patients with MG have weakness and muscle fatiguability and up to 20% of patients will experience an MC.^{2,3} Indications for mechanical ventilation

include forced vital capacity ≤ 15 mL/kg (normal ≥ 60 mL/kg), hypoxaemia or hypercapnia; and although there are no randomised controlled data assessing the value of SNIP in MG or MC, a value ≤ 20 cm H₂O is regarded as high risk. In addition to measuring SNIP, the maximal inspiratory mouth pressure PI (max) is a valuable and reliable additional test for the assessment of inspiratory muscle strength in patients with neuromuscular disorders.⁴ Once ventilation is secure acute therapy is initiated and usually includes either plasma exchange therapy or intravenous immunoglobulins with comparable efficacy. Long-term therapy includes acetylcholinesterase inhibitors, corticosteroids, thymectomy and immunomodulating agents. With optimal therapy, prognosis is favourable.⁵

Contributors AMN was responsible for drafting and revising of the article and was involved in managing the case. JJ was involved in managing the case and reviewed the manuscript. MH was responsible for the final approval and for managing the case.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Vincent A, Palace J, Hilton-Jones D. Myasthenia gravis. *Lancet* 2001;357:2122–8.
- 2 Amato AA. Noninvasive mechanical ventilation in patients with myasthenic crisis. *Nat Clin Pract Neurol* 2008;4:356–7.
- 3 Spillane J, Hirsch NP, Kullmann DM, et al. Myasthenia gravis—treatment of acute severe exacerbations in the intensive care unit results in a favourable long-term prognosis. *Eur J Neurol* 2013;21:171–3.
- 4 Hart N, Polkey MI, Sharshar T. J Neurol limitations of sniff nasal pressure in patients with severe neuromuscular weakness. *Neurosurg Psychiatry* 2003;74:1685–7.
- 5 Jayawant S, Parr J, Vincent A. Autoimmune myasthenia gravis. *Handb Clin Neurol* 2013;113:1465–8.
- 6 Ali SS, O'Connell C, Kass L, et al. Single-breath counting: a pilot study of a novel technique for measuring pulmonary function in children. *Am J Emerg Med* 2010;29:33–6.
- 7 American Thoracic Society/European Respiratory Society. ATS/ERS Statement on respiratory muscle testing. *Am J Respir Crit Care Med* 2002;166:518–624.

Learning points

- The use of vital capacity is familiar to all physicians but we highlight two simple, reproducible bedside tests to help estimate ventilatory function: single breath count, which in a paediatric cohort of asthma patients has been shown to accurately estimate forced vital capacity (FVC) where 10 counts, in cadence to a metronome set at 2 bps, correlate with 1 L FVC and sniff nasal inspiratory measurements, a volitional measure of diaphragmatic function.⁶
- We recommend the American Thoracic Society/European Respiratory Society statement on respiratory muscle testing, which includes a paragraph on volitional tests of respiratory muscle strength.⁷
- A high index of suspicion is required for myasthenic crisis. Our patient described no orthopnoea when lying quietly but severe dyspnoea following minimal movement and difficulty in finishing sentences; neither respiratory muscle weakness nor fatiguability was apparent at rest.
- A careful clinical history supported by demonstration of respiratory muscle fatiguability allowed correct and timely treatment of his condition.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit

<http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow