CASE REPORT

Difficult to wean: think of the diaphragm

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SUMMARY

A 65-year-old woman, treated for non-ST segment elevation myocardial infarction and unexplained type 2 respiratory failure, was referred to our hospital in view of difficulty in weaning off the ventilator. She was evaluated in detail for persistent hypercapnia. Ultrasound of the diaphragm showed minimal excursion of diaphragm while she was off the ventilator and fluoroscopy confirmed bilateral diaphragmatic palsy. As extensive radiological, immunological and microbiological workup ruled out other possible aetiologies, a diagnosis of idiopathic Bell's palsy of the diaphragm was made. She was treated with valacyclovir and steroids after which she gradually recovered and was weaned off the ventilator.

BACKGROUND

As the majority of patients with diaphragmatic palsy are asymptomatic, the exact incidence is not known.¹ Non-traumatic bilateral diaphragmatic palsy is usually an underdiagnosed condition, the mean time for diagnosis being 2 years.² The diagnosis of Bell's palsy of the diaphragm should be thought of in cases of acute bilateral idiopathic diaphragm paralysis only when other possible aetiologies have been ruled out. Bell's palsy of the phrenic nerve is analogous to idiopathic paralysis of the seventh cranial nerve and the proposed aetiologies for this condition are postviral infection mainly by herpes simplex and varicella zoster.³ It is important to consider this entity when there is a diagnostic dilemma as there could be improvement in the diaphragmatic function with a therapeutic trial of antiviral drugs and steroids. This prevents unnecessary medical interventions and diagnostic delay.

CASE PRESENTATION

A 65-year-old woman, treated for urinary tract infection with injectable amikacin 1 week ago, presented with worsening of breathlessness since 1 day. Initial evaluation revealed non-ST elevation myocardial infarction (ECG: ST depression in antero-septal leads, elevated troponin T, echocardiogram: hypokinesia of interventricular septum) and type 2 respiratory failure (arterial blood gas (ABG): PaO₂ 74, PaCO₂ 93). She was treated with low molecular weight heparin, antiplatelets and statins for non-ST elevation myocardial infarction. She was intubated and ventilated in view of unexplained respiratory failure. Over the next few days her cardiac function improved and repeat echocardiography, before weaning, showed good ejection fraction without regional wall motion abnormalities. Despite being symptomatically better and

haemodynamically stable, she had failed three spontaneous breathing trials, each resulting in hypercapnia. She was referred to our hospital in view of difficulty in weaning off the ventilator. At presentation, her vital signs were stable. She was a known diabetic and hypertensive, compliant on treatment. There was no history of chronic cough, breathlessness, snoring, daytime sleepiness, neurological problems or of drug overdose. She had a normal cough reflex, good neck muscle power, no evidence of muscular wasting or fasciculations and bilateral flexorplantar reflex. Other systemic examinations were unremarkable. Detailed evaluation for cause of persistent hypercapnia was conducted.

INVESTIGATIONS

MRI of the brain and cervical spine were normal, ruling out brain stem stroke, multiple sclerosis and high cervical cord lesions. CT of the thorax and neck ruled out mass lesion causing compression of phrenic nerve. Tests for myasthenia were negative (repetitive nerve stimulation, neostigmine test and antiacetylcholine receptor antibodies). The neostigmine test was performed as follows. At the start of the test, the patient was comfortable on ventilator support and movements of the diaphragm were assessed using ultrasonography. After giving intravenous neostigmine, the ventilator support was gradually tapered and she was assessed clinically and ultrasonographically for movements of the diaphragm at regular intervals over 1 h. There was no improvement postneostigmine; hence myasthenia gravis was considered an unlikely possibility. Electromyography was performed to rule out motor neurone disease. Nerve conduction velocities were performed to rule out Guillain-Barre syndrome. Virological studies of cerebrospinal fluid were negative ruling out subclinical postencephalitic sequelae. Details of the investigations are summarised in table 1.

DIFFERENTIAL DIAGNOSIS

Summarised in table 2.

TREATMENT

A possibility of Bell's palsy of the diaphragm was considered, as all the known aetiologies for bilateral diaphragmatic palsy were ruled out. She was treated with valacyclovir 1000 mg three times a day for 14 days along with high dose methylprednisolone (1 gram) for 5 days followed by prednisolone (1 mg/kg) as maintenance therapy.

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Table 1 Investigations

CBP, RFT with serum electrolytes, LFT and TSH	Normal
ABG analysis	Type 2 respiratory failure (Po ₂ 74 and Pco ₂ 93)
Troponin T	0.46 (normal <0.02)
ECG	ST depression in anteroseptal leads
Echocardiography	Hypokinesia of interventricular septum and apex (ejection fraction 52%)
Repeat ECHO (before weaning trials):	Normal cardiac function (ejection fraction 62%)
Fluoroscopy/ultrasonography	Minimal excursion of diaphragm off ventilator (<2 mm)
MRI of the brain and spinal cord	Normal
CT of the thorax and neck	No mass lesion
Nerve conduction studies	Normal
Cerebrospinal fluid analysis	Normal
Cerebrospinal fluid PCR for herpes simplex, varicella zoster and Epstein-Barr virus	Negative
Neostigmine test	Negative
Repetitive nerve stimulation, antiacetylcholine receptor antibodies	Negative
Electromyography	Normal

ABG, arterial blood gas; CBP, complete blood picture; ECHO, echocardiogram; LFT, liver function test; RFT, renal function test; TSH, thyroid stimulating hormone.

OUTCOME AND FOLLOW-UP

She made a gradual recovery and was weaned off the ventilator over the next 3 weeks. Repeat ultrasonography showed normal movements of the diaphragm and ABG showed no evidence of CO₂ retention. She was later on discharged from the hospital. At 1-year follow-up, she was asymptomatic with a normal ABG.

Table 2 Differential diagnosis

Possible aetiologies	Points against diagnosis
Guillain-Barre syndrome	Reflexes preserved Cerebrospinal fluid—normal (after 10 days) Nerve conduction studies—no conduction blocks
Brain stem stroke	MRI of the brain—normal
Cervical cord compression	MRI of the cervical spine—normal
Bilateral phrenic nerve compression	CECT of the thorax—normal
Motor neurone disease	Short history No fasciculations Electromyography—normal
Myasthenia gravis	Neostigmine test—negative Antiacetylcholine receptor antibodies—negative Repetitive nerve stimulation test—negative
Neuromuscular blockade postaminoglycoside treatment	Unlikely after 1 week of exposure
Myopathy	Electromyography—normal
CECT, contrast-enhanced CT.	

DISCUSSION

Bilateral diaphragmatic paralysis is more often seen in the background of severe generalised muscle weakness. When the diaphragm is the only muscle involved, determining aetiology poses a diagnostic challenge and requires a high clinical suspicion. Although a battery of tests is available, most of them are non-specific.^{4 5}

Transdiaphragmatic pressure measurement is considered as the gold standard for diagnosing bilateral diaphragmatic paralysis, but it is invasive and not readily available.¹ A suitable alternative is non-invasive ultrasonographic measurement.⁶

Once a diagnosis of bilateral diaphragmatic paralysis is made, a workup for all possible aetiologies should be performed. The diagnosis of myasthenia gravis may be difficult to make and is often misleading as antiacetylcholine receptor antibody titres may be negative and repetitive nerve stimulation may not show decremental responses in patients with isolated respiratory muscle involvement.⁷ In such a scenario, the neostigmine test is necessary to prove or disprove the diagnosis. This is one of the rare cases in which the neostigmine test was used to rule out myasthenia gravis in a patient with isolated bilateral diaphragmatic palsy on ventilator where negative antiacetylcholine receptor antibodies and repetitive nerve stimulation could not be relied on.

The treatment of idiopathic bilateral diaphragmatic palsy with valacyclovir and steroids was based on a preliminary study which considered the condition as Bell's palsy of the diaphragm and treatment with antiviral drugs and steroids showed improvement in diaphragmatic function.³ Patients are expected to make a spontaneous recovery which may be prolonged up to 2–3 years,⁸ but can be brought down to 4–6 weeks with treatment.³ Hence a therapeutic trial is warranted in patients presenting with idiopathic diaphragmatic palsy.

Learning points

- ▶ Bilateral diaphragmatic palsy should be considered in the differential diagnosis in patients presenting with type 2 respiratory failure.
- ▶ Bell's palsy of the diaphragm is to be considered in an acute presentation of bilateral diaphragmatic palsy when other aetiologies are ruled out.
- ▶ Therapeutic trial of antiviral drugs and steroids should be given in Bell's palsy of the diaphragm as it hastens recovery.

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Competing interests None.

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