An unusual cause of obstructive sleep apnoea

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Obstructive sleep apnoea is usually associated with obesity and is caused by recurrent collapse of the pharyngeal airway during sleep at multiple levels. Vocal cord paralysis is an uncommon cause of obstructive sleep apnoea. We describe a patient with obstructive sleep apnoea caused by idiopathic vocal cord paralysis. He also had features of autonomic failure including orthostatic hypotension and urinary retention.

Review of the literature showed that patients with the Shy–Drager syndrome can have these manifestations. However, our patient did not have the neurological features of the Shy–Drager syndrome, apart from the autonomic failure. We believe that vocal cord paralysis and sleep apnoea can occur in association with features of autonomic failure, in the absence of,

CASE REPORT

A 60-year-old retired safety guard with non-atopic asthma has been followed up in our asthma clinic since 1993. He has a 75 pack/year smoking history and his spirometric values were as follows: forced expiratory volume in 1 second (FEV_1) was 2.09 litres (88% predicted), forced vital capacity (FVC) 3.26 litres (112% predicted), FEV_1/FVC 63%. After inhalation of salbutamol 400 μg with a metered dose inhaler, FEV_1 improved to 2.49 litres (20% improvement), FVC was 3.48 litres and FEV_1/FVC 72%. His asthma was well controlled with terbutaline and budesonide inhalations and theophylline. Other significant history included frequency of urine and sense of incomplete voiding since 1994. This had been treated as benign prostatic hypertrophy by transurethral resection of the prostate in another hospital without benefit.

He complained of frequent syncopal attacks while walking in 1996. A marked postural drop in blood pressure was noted in the clinic. Supine blood pressure was 140/90 mmHg and erect blood pressure was 70/50 mmHg. He was not taking any antihypertensive drugs. He was admitted to our unit for further investigations.

Physical examination did not reveal any abnormality apart from the postural hypotension. In particular, he was not hypovolaemic. Chest examination was normal. Phonation was normal. There were no neurological features of parkinsonism, cerebellar or corticospinal disturbance, or peripheral neuropathy. The prostate was not clinically enlarged. Complete blood count and serum biochemistry were normal. Urinalysis was normal.

He was not diabetic. Serology for antinuclear factor, rheumatoid factor and anti-double stranded DNA was negative. Thyroid function test was normal and a short tetracosactrin test showed normal response. His postural drop in blood pressure was investigated with an arterial line placed in his left radial artery. Supine blood pressure was 180/90 mmHg and heart rate was 92/min. He was then placed at a 60° tilt on a tilt table. His blood pressure fell progressively to a low of 61/40 mmHg in 9 minutes accompanied by dizziness, and the heart rate was 110 beats/min. Valsalva manoeuvre showed a lack of overshoot in phase IV. Submaximal handgrip showed no hypertensive response.

His urinary symptom was examined with cystometry, which showed incomplete emptying of bladder with impaired sensation, compatible with autonomic failure. Other selected tests of the autonomic system, namely sympathetic skin response to intracutaneous injection of histamine and pupillary response to instillation of 1:1000 adrenaline, were normal. Nerve conduction studies on peripheral nerves were normal.

While he was in hospital, he was noted to have loud inspiratory stridor and paradoxical thoraco-abdominal breathing movement during sleep. On direct enquiry, he admitted to have hypersomnolence (Epworth sleepiness scale 11/24) and his family members confirmed he has loud 'snoring' at home. Polysomnography confirmed obstructive sleep apnoea syndrome with an apnoea–hypopnoea index of 24/hour. Baseline arterial oxygen saturation (SaO_2) by pulse oximetry was 98%. Recurrent desaturations below 95% occupied about 40% of total sleep time and the lowest SaO_2 recorded was 87%. These episodes were associated with stridor, cessation or diminution of airflow and thoraco-abdominal paradox. These events occurred as soon as he fell asleep and were present in stage 1 and stage 2 sleep. His sleep was severely disturbed with frequent awakening related to these episodes. No stage 3, stage 4 or rapid eye movement sleep was recorded on the night of study. A nasendoscopy was performed and examination of the nose, pharynx and larynx while awake were normal.

Repeat examination with intravenous sedation by midazolam (3 mg) was performed. Stridor, obstructive apnoea and desaturation were reproduced and the vocal cords were seen to occupy the paramedian position and remained adducted with each inspiratory effort. A diagnosis of bilateral abductor vocal cord paralysis was made. Examination of the tracheobronchial tree was normal. Reversal of these abnormalities occurred with intravenous flumazenil, and phonation was normal when the patient was awake.

A computed tomography of the neck and thorax showed no abnormality along the laryngeal nerves. A magnetic resonance scan of the brain revealed no abnormality. A trial of continuous positive airway pressure (CPAP) was offered. A maximum CPAP of 13 cmH_20 was delivered which was unable to correct the obstruction and he could not tolerate higher pressure. Failing CPAP, the patient consented to a tracheostomy. Tracheostomy with a fenestrated tube (Shiley™, St Louis, USA) for use during sleep was fashioned. Follow-up polysomnography with tracheostomy showed total relief of his obstructive sleep apnoea.

The patient received regular follow-up. His postural hypotension was partially controlled with fludrocortisone and pressure stockings (supine blood pressure 140/90 mmHg, erect blood pressure 90/60 mmHg) with good symptom relief. He reported relief of somnolence (Epworth sleepiness scale 6/24). When he was last seen in May 1998, there were no motor neurological manifestations of the Shy–Drager syndrome.

or preceding other manifestations of the Shy–Drager syndrome.

DISCUSSION

Vocal cord paralysis and autonomic failure have been reported in the Shy–Drager syndrome (Israel and Marino, 1977). Shy and Drager (1960) originally described a syndrome of progressive autonomic failure and parkinsonism. The full syndrome comprises the following features:

- Orthostatic hypotension
- Urinary and rectal incontinence
- Loss of sweating
- Iris atrophy
- Extraocular paresis
- Parkinsonism, including tremor, rigidity and slowness of movement
- Pyramidal and cerebellar signs.

Thomas and Schirger (1970) described a further 57 cases. In these earlier reports, there was no description of vocal cord paralysis.

Israel and Marino (1977) reported a patient with the Shy–Drager syndrome who developed respiratory failure resulting from bilateral vocal cord paresis requiring emergency tracheostomy. A patient with the Shy–Drager syndrome, sleep apnoea and unilateral vocal cord paresis was reported to die during sleep (Guilleminault et al, 1977).

The largest series of vocal cord paralysis in the Shy–Drager syndrome was reported by Williams et al (1979). Nine out of twelve patients with the classical Shy–Drager syndrome had varying degrees of bilateral abductor paresis and two had unilateral paresis. They presented with snoring, stridor and sleep apnoea. Tracheostomy was performed in five patients. One patient who declined a tracheostomy died from a respiratory arrest during sleep in this series. All patients in this series had parkinsonism, orthostatic hypotension, sexual and sphincter dysfunction. Those with more severe vocal cord paresis tended to have more severe autonomic and motor manifestations as well. Some patients also developed cerebellar and bulbar dysfunction.

Our patient had vocal cord paralysis, obstructive sleep apnoea and evidence of autonomic failure with orthostatic hypotension and sphincter disturbance. He was different from the cases reported elsewhere in that he had no extrapyramidal features of the Shy–Drager syndrome.

The Shy–Drager syndrome is now thought to overlap with olivo-pontocerebellar atrophy and striatonigral degeneration, all being umbrellaed under the term multiple system atrophy (MSA). In the largest survey to date of clinicopathological features of 203 pathologically proven MSA cases, parkinsonism was absent in 13% of cases (Wenning et al, 1997). Indeed, symptoms of autonomic failure may predate the onset of motor disturbance by up to several years (Polinsky, 1984). Respiratory stridor was reported in 13% of the patients in the same series.

Vocal cord paralysis causing obstructive sleep apnoea seems to be associated with a significant mortality in the series described (Guilleminault et al, 1977; Williams et al, 1979) and it is prudent to relieve the obstruction in these patients. Williams et al treated their patients with tracheostomy and our patient was also treated with tracheostomy with good response. Continuous positive airways pressure (CPAP) was not successful in our patient, although it is generally useful in the usual form of sleep apnoea caused by pharyngeal airway collapse. It is possible that while CPAP is able to 'splint' the pharyngeal airway, it may not be able to splint the paralyzed vocal cords.

CONCLUSION

Clinicians need to be aware of the association of obstructive sleep apnoea, vocal cord paralysis and autonomic failure in the uncommon condition of MSA. This constellation may occur with or without other neurological features of MSA. **HM**

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