Clinical Reports

Obstructive sleep apnoea associated with syringomyelia

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Summary: We present a case of obstructive sleep apnoea in association with syringomyelia. We describe the successful treatment of the respiratory obstruction by continuous positive airway pressure and then by surgical means. This rare combination of conditions and the management is reviewed.

Introduction

Syringomyelia-syringobulbia has been reported to cause either isolated central sleep apnoea¹ or isolated obstructive sleep apnoea.^{2–5} The anatomy and behaviour of these lesions allows them to interfere with either the central drive to breathe or with the innervation to oropharyngeal muscles involved in maintenance of local tone. Here we report a case of syringomyelia associated with profound obstructive sleep apnoea which responded dramatically to treatment.

Case report

A 32 year old farm labourer presented with an 8 month history of progressive daytime tiredness and of falling asleep at work. He had previously been well, though he smoked 40 cigarettes a day and drank at least four bottles of red wine and 15 pints of beer per week. He snored loudly at night and had noticed persistent numbness in his right upper limb and painless ulcers of his right elbow and hand. In addition, he frequently burnt himself when cigarettes fell onto his chest when he went to sleep. Sometimes the pain would wake him, but in one area of his chest he could not feel himself being burnt.

On examination he was plethoric and obese, weighing 107 kg at a height of 1.74 m. He had ulceration of his right hand and elbow as well as cigarette burns in a band across his chest. Cranial nerve examination was normal as was his speech. His oropharynx, however, revealed huge tonsils and soft tissue hypertrophy. Tone and power in the limbs was normal apart from grade 4/5 weakness of finger abduction and adduction of the right hand.

Reflexes were absent in the right arm but were normal elsewhere. Gait and coordination were also normal. There was absent pain and temperature sensation in the distribution of C5–C8, and T5–T7 dermatomes on the right as well as T6 and T7 on the left. Light touch and posterior column sensation was normal.

Investigations

Renal, liver and thyroid function tests were normal, the erythrocyte sedimentation rate was 10 mm/hour, and syphilis serology was negative. However, the haemoglobin was 17.3 g/dl with a packed cell volume of 0.52. Arterial blood gases in air were as follows: pH 7.40, PaCO₂ 6.5 kPa, PaO₂ 8.2 kPa, bicarbonate 31 mmol/l, oxygen saturation 91%. The chest X-ray showed patchy basal atelectasis and the electrocardiogram P-pulmonale and an axis of +90. There was a thoracic kyphoscoliosis and a healed fracture of the sixth cervical vertebra on the plain spinal films. Plain X-rays of his right elbow and wrist showed evidence of neuropathic arthropathy.

Magnetic resonance imaging demonstrated a syrinx at the level of C2 to T7, with the cerebellar tonsils protruding through the foramen magnum by 5 mm, corresponding to a type 1 Arnold-Chiari malformation.⁶ It also demonstrated the massive enlargement of the soft palate and uvula.

At spirometry the forced expiratory volume in 1 second (FEV₁) was 2.97 l (predicted 4.11 l) and forced vital capacity (FVC) was 4.34 l (predicted 4.90 l). There were low maximum inspiratory and expiratory mouth pressures of 2.1 kPa (predicted 9.98 kPa) and 4.7 kPa (15.96 kPa), respectively, suggesting weakness of respiratory musculature. Overnight sleep studies were performed with finger oximetry, chest wall excursion monitoring and careful observation. These revealed recurrent

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episodes of obstructed respiration with hypoxaemia: the arterial oxygen saturation dropping to below 50% on five occasions during the night (Figure 1). Fluoroscopic screening of the oropharynx was also undertaken during midazolam-induced sleep. This demonstrated the obstruction of the pharyngeal lumen by the floppy redundant soft palatal tissues. At no time during the sleep studies did complete cessation of chest wall movement occur, and the pattern of these movements were characteristic of obstructive sleep apnoea.

Management

The patient was treated initially with continuous positive airway pressure (CPAP) during sleep via a tightly fitting nasal mask at a mean pressure of 10 cm water. There was a dramatic improvement in oxygen saturation during use (Figure 2), with a

good symptomatic response. The patient remained on CPAP at night for the next 18 days. In view of this evidence he underwent uvulopalatopharyngoplasty. One year after presentation he has a settled sleep pattern (Figure 3), and has stopped drinking. He has lost 12 kg in weight and has recovered his former mental alertness. His neurological condition is stable.

Discussion

Sleep apnoea is classified into three broad categories with some overlap between the groups.^{7,8} It can be a purely central problem, with disruption of the respiratory centres of the medulla, or more commonly it can be an obstructive problem. In the latter group a previously narrow airway becomes obstructed at night, due to the physiological relaxation of oropharyngeal muscle tone and this

Figure 1 Overnight oximetry at presentation.

Figure 2 Overnight oximetry showing effects of CPAP for 1 hour.

Figure 3 Overnight oximetry following surgery.

stimulates increasingly vigorous respiratory efforts to overcome it. Finally, sleep apnoea can be a combination of these two mechanisms – when recurrent obstruction-induced hypoxaemia and hypercapnia also progresses to a down-grading of the central drive to breathe.

Central sleep apnoea has been reported with several brainstem lesions including infarction of the medulla,⁹ olivopontocerebellar degeneration,¹⁰ encephalitis,¹¹ and Arnold-Chiari malformation.¹² It may also be idiopathic as the classical 'Ondine's Curse'.¹³

Obstructive sleep apnoea most often occurs in obese men, particularly those who drink alcohol heavily at night. It is common in chronic mouth breathers and in those with anatomical narrowing of the upper airway.⁸ It has been reported, however, in neurological conditions which can cause airway obstruction such as poliomyelitis,¹⁴ Shy-Drager syndrome,¹⁵ and motor neurone disease.¹⁶

Syringomyelia-bulbia with or without Arnold-Chiari malformation have been found to be associated with central sleep apnoea,^{1,17} and syringomyelia has caused obstructive sleep apnoea via vocal cord paralysis.⁵ Two cases have also described cranial nerve dysfunction and oropharyngeal muscle weakness with airway occlusion due to a syrinx extending into the brainstem. Both cases responded to tracheostomy and demonstrated no interruption of the central drive to breathing.^{2,3}

Here we report a case of obstructive sleep apnoea and pure syringomyelia: this is the second time this combination has been reported, but in this case there was no evidence of vocal cord paralysis. We cannot, however, exclude an element of central sleep apnoea since our study did not include polysomnography. Our patient was obese with soft tissue hypertrophy of the palate and drank excess

alcohol. Syringomyelia-induced denervation of the respiratory muscles may have caused weakness contributing to his difficulty in overcoming the upper airway obstruction.

The first line of treatment for obstructive sleep apnoea consists of simple measures, such as weight loss and abstinence from alcohol in the evenings. Tricyclic antidepressant drugs that decrease the time in rapid eye movement sleep and increase the tone in the muscles of the upper airway have also been used.¹⁸ Unfortunately they have a large number of side effects related to their anticholinergic properties.

Nasal CPAP increases the patency of the pharyngeal lumen by the application of positive pressure via a tightly-fitting nasal mask.¹⁹ The degree of pressure applied is determined by the response at sleep study and the range is usually between 0.5–1.5 kPa. There are various problems with this treatment that are not uncommon. The mask itself can be very uncomfortable, especially at first, and if fitted incorrectly can cause sores on the skin. Secondly, the maintenance of positive pressure within the pharyngeal lumen is restricted if the mouth falls open during sleep, and sometimes this has to be prevented. Finally, nasal congestion renders the system considerably less effective.

Treatment can sometimes be surgical, as in this case. The operation of uvulopalatopharyngoplasty consists of the removal of 'excess' mucosal tissue in the pharynx including the tonsils, uvula and a variable amount of the soft palate.²⁰ One of the most serious complications of the procedure occurs if too much tissue in the soft palate is removed, which can result in escape of air and fluid through the nose. The procedure usually provides good relief of snoring, but is less successful at preventing apnoeic episodes. The definition that is usually

applied in studies assessing the success of such surgery is at least a 50% reduction in sleep-disordered breathing events. If this definition is taken, the 'response' rate is usually found to be about 50%.²¹ It is not yet clear how to predict who will respond best to treatment by surgery. However, some studies do suggest that those patients with obstruction at the level of the oro- rather than the hypo-pharynx do better.²² There is no agreement as to whether body weight correlates with response.²³ The operation therefore has a relatively

low success rate, and is probably best reserved for those in whom visualization and imaging demonstrates severe soft tissue obstruction of the pharyngeal lumen, particularly at the oropharyngeal level.

We suspect that other cases of syringomyelia-syringobulbia, as well as other neurological conditions such as those mentioned above, may benefit from identification and treatment of the obstructive apnoea syndrome to which they may be more prone.

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