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Abductor vocal fold palsy in the Shy-Drager syndrome presenting with snoring and sleep apnoea

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Abstract
The case of an elderly male with Shy-Drager syndrome is presented. His presentation to the Sleep Clinic for assessment of snoring illustrates bilateral abductor vocal fold palsy as a rare presentation of the syndrome. This case emphasizes the need for thorough investigation of all patients with sleep-related breathing disorders with video and sound recordings prior to anaesthesia and surgery.

Key words: Shy-Drager syndrome; Snoring; Sleep apnoea syndromes; Vocal fold paralysis

Introduction

Bilateral vocal fold palsy is a recognised presenting sign of the Shy-Drager syndrome (Williams *et al.*, 1979). This condition involves progressive autonomic failure with orthostatic hypotension, urinary and faecal incontinence, cerebellar, pyramidal and extrapyramidal signs, above or in combination (Shy and Drager, 1960).

The Shy-Drager syndrome was diagnosed in an elderly male patient following his referral to the Sleep Clinic. Inadequate initial assessment of the patient had resulted in inappropriate treatment, viz. intranasal steroids for his rhinitic nose, in an attempt to treat 'snoring'.

The diagnosis of Shy-Drager syndrome was made only after demonstrating nocturnal stridor by review of video and sound recordings which are part of our full polysomnographic assessment of patients with sleep-related breathing disorders.

Case history

The patient, a 66-year-old man, presented with late onset snoring. He had a history of maturity onset diabetes and allergic rhinitis. He had been treated with topical nasal steroids with alleviation of his nasal symptoms but no improvement in the snoring. His wife reported some unsteadiness of gait and nocturnal urinary incontinence in the preceding months but these were not marked.

He was referred to the Sleep Clinic for further assessment. Examination including flexible nasendoscopy showed limitation of abduction of the vocal folds. He underwent a sleep study which involved overnight recording of pulse oximetry, heart rate, chest and abdominal movements along with nasal airflow. The apnoea/hypopnoea index was 22 (normal <15) and oxygen saturation fluctuated between the upper 80's and 90's per cent for most of the study. This demonstrated results consistent with mild obstructive apnoea although the nurse observer noted that his snoring sounded 'odd' and was stridulous at times.

He was next referred for fibreoptic endoscopy of the upper airway under light anaesthesia ('sleep nasendoscopy'). This procedure had to be terminated (unusually) because the patient developed laryngospasm. The surgeon commented on the lack of secretions which might have caused this. He was deemed unsuitable for palatal surgery on basis of the observations made.

He underwent an overnight study using nasal continuous positive airways pressure which abolished the 'snoring' and maintained his oxygen saturation in the mid 90's. His wife by this stage reported worsening of his ataxia and incontinence along with memory loss.

He was referred back for full polysomnography including video recording and digital audio tape (DAT) sound. His noisy breathing was in fact biphasic stridor which was his 'snoring'. His apnoea/hypopnoea index was similar to the original sleep study with oxygen saturation repeatedly in the upper 80's. ECG monitoring revealed frequent bradycardia. Sleep staging was disrupted by very frequent cortical arousal and reduced stage IV and REM sleep. He subsequently was referred to a neurologist who made a definitive diagnosis of Shy-Drager syndrome.

Discussion

Stridor during sleep, reported as 'snoring', is an uncommon but recognised presenting feature of the Shy-Drager syndrome. It is common in the later stages of the disease when bilateral vocal fold palsy often requires tracheostomy to maintain the airway and to protect the lungs from aspiration, frequently the cause of death.

Loud and persistent snoring is a common disorder which may affect up to 60 per cent of elderly males (Lugaresi *et al.*, 1980). As well as marital disharmony, the recognition of the harmful effects of the associated sleep apnoea syndrome has lead to the emergence of dedicated Sleep Laboratories and Clinics for the assessment of patients with sleep disorders.

Assessment at the RNTNE sleep laboratory includes overnight video recording and polysomnography followed

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by fibreoptic endoscopy of the upper airway under light anaesthesia (Croft and Pringle, 1991). The patients may subsequently be referred for uvulopharyngopalatoplasty or laser palatopexy which have become common and effective treatments for palatal flutter in snoring (Ellis *et al.*, 1993). The role of other surgical procedures for the treatment of snoring due to tongue base and hypopharyngeal collapse and for sleep apnoea syndrome remains to be proven.

Patients with Shy-Drager syndrome are at increased risk during anaesthesia as they may develop severe hypotension or exhibit respiratory depression. (They may be sensitive to inducton agents). Postural hypotension may be marked. Defective lacrimination, poor pupillary response and reduced sweating can make assessment of anaesthetic depth difficult. Additional hazards include those associated with an absent gag reflex, gastric dilatation and a high pain-threshold (Sweeney *et al.*, 1985).

This case re-iterates the need for thorough assessment of patients with sleep disorders and a multi-disciplinary approach to treatment. It is only by doing so that the rarer causes of disorders of respiration during sleep will be identified and inappropriate or potentially harmful surgery avoided.

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