Obstructive Sleep Apnoea-Hypopnoea Syndrome And Idiopathic Intracranial Hypertension: Coincident, Comorbid, Or Causal Relationship?

In obese patients, papilloedema may be due to idiopathic intracranial hypertension or obstructive sleep apnoea-hypopnoea syndrome, diagnoses with widely differing therapeutic implications. However, the overlap between these conditions may be aetiological, as well as clinical.

INTRODUCTION
Patients with obstructive sleep apnoea-hypopnoea syndrome (OSAHS) may present to the neurology clinic with various symptoms, including loss of consciousness, stroke, excessive daytime somnolence, and cognitive decline. Symptoms and signs suggestive of raised intracranial pressure (ICP) are also recognised features of OSAHS, namely headache with early morning waking and exacerbation with recumbency and papilloedema. These clinical features may be mistaken for those of idiopathic ('benign') intracranial hypertension (IIH; pseudotumor cerebri). This is an important differential diagnosis to be aware of, with distinctive therapeutic implications. However, the relationship between these two conditions may not be simply exclusive.

CASE REPORT
A 43 year-old woman was referred to hospital with shooting pains around the right eye, unrelated to posture or activity. Examination revealed visual acuities of 6/9 right and 6/5 left (uncorrected) and bilateral papilloedema (see Figure). Automated visual field plotting showed enlargement of the blind spots, more evident on the right. MR imaging of the brain and orbits was normal, with no evidence of venous sinus thrombosis. Lumbar puncture showed an opening pressure of 27 cm H2O; CSF cell count, glucose and protein were normal. A diagnosis of idiopathic intracranial hypertension was made and treatment with acetazolamide commenced.

At subsequent neurological consultation, it emerged that the patient was a long-standing snorer with frequent nocturnal awakenings (corroborated by her bed partner), excessive daytime sleepiness and a tendency to ‘catnap’ during the day. In addition to the papilloedema, she was noted to be obese (weight 115 kg; height 1.69 m; body mass index [BMI] 39 kg/m2), with crowding of the oropharynx. On the Epworth Sleepiness Scale, a validated measure of subjective daytime sleepiness which assesses the likelihood of falling asleep in certain situations, the patient’s score was moderately high (14/24). A sleep study (overnight home oximetry) showed cyclic oxygen desaturations throughout the night (mean oxygen saturation = 93%). The desaturation index (DI), the number of oxygen desaturations ≥ 4% per hour of sleep, was 13; DI = 5 may be used to define sleep-disordered breathing. A diagnosis of obstructive sleep apnoea-hypopnoea syndrome (OSAHS) was made. Acetazolamide was stopped without complication, and she was referred for a trial of nocturnal continuous positive airway pressure (CPAP).

DISCUSSION
The differential diagnosis of OSAHS is broad: loss of consciousness may be mistaken for epilepsy, excessive daytime somnolence for narcolepsy, and cognitive decline for dementia. OSAHS may also present with features suggestive of raised ICP, namely headache, papilloedema and visual failure, although some authorities report early morning headache to be a rare feature. These symptoms and signs are thought to be secondary to episodic nocturnal hypoxaemia and hypercapnia, consequent upon nocturnal hypoventilation, with cerebral vasodilation.

As illustrated in the current case, the clinical syndrome may be mistaken for idiopathic intracranial hypertension (IIH). However, this patient did not fulfill the modified Dandy criteria for the diagnosis of IIH, since headache with features of raised ICP was absent. The modest elevation of CSF opening pressure is also unusual in IIH, where pressures of >300 mm H2O are more typically seen. However, the relationship between OSAHS and IIH may not be simply one of exclusion; the two conditions may be related. In a tertiary neuro-ophthalmology practice, 9 of 53 patients diagnosed with IIH (F:M = 5:1:2), 37 admitted to snoring and daytime somnolence. Of these, 14 underwent polysomnography (F:M = 12:2; mean age 39.4 +/- 11.9 years; mean BMI = 46.0 +/- 9.5 kg/m2) and six were found to have OSAHS and seven the related condition of upper airway resistance syndrome. A multicentre retrospective study at three tertiary care academic ophthalmology centres identified 32 men with IIH, six of whom had sleep apnoea, treatment of which with CPAP improved the symptoms and signs of IIH. Although these series are highly selected, nonetheless they raise the possibility that sleep-related breathing problems may be more common in IIH than expected by chance alone.

The exact relationship between OSAHS and IIH remains uncertain. Sleep-related breathing disorders may be a risk factor for IIH, or they may be simply comorbid conditions with no causal association. Clearly both are related to obesity, but the reversed sex incidence of IIH and OASAS indicates that there must be other factors in operation. A large prospective study might identify any such factors. In the meantime, the inter-relationship of OSAHS and IIH should be recognised. Hence, it would seem prudent to recommend that when a diagnosis of IIH is established, patients (and their bed partners) be questioned for symptoms of sleep disturbance (excessive daytime somnolence, snoring, nocturnal awakenings, apnoeic episodes, road traffic accidents), with a view to selecting those appropriate for investigation of possible OSAHS. Men with IIH should perhaps be subject to particular scrutiny, since IIH is rare and OSAHS common in men. The conditions have differing therapeutic implications: diuretics such as acetazolamide for IIH; CPAP for OSAHS, and possibly surgery for those intolerant of CPAP.

See next page for References
References