CASE REPORT

Declining vision: saved by a tracheostomy

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SUMMARY

A 37-year-old woman presented to our hospital with a history of headaches, blurred vision and declining visual acuity. She had a history of obstructive sleep apnoea associated with Treacher Collins syndrome, a rare congenital disorder characterised by craniofacial deformities and obstruction of the upper airways. On funduscopy we noted bilateral papilloedema and retinal haemorrhages. A CT scan of the brain excluded a space occupying lesion and contrast-enhanced CT did not show evidence of venous sinus thrombosis. The cerebrospinal fluid opening pressure on lumbar puncture was normal. We made a diagnosis of visual loss and papilloedema secondary to chronic CO₂ retention from obstructive sleep apnoea. Her vision and headaches improved remarkably following a permanent tracheostomy. This case highlights the association between obstructive sleep apnoea and papilloedema and shows that the associated visual loss can ultimately be restored with a tracheostomy.

BACKGROUND

Papilloedema is a recognised but rare presentation of obstructive sleep apnoea syndrome. We described a woman who presented with papilloedema and visual loss for which no plausible aetiology was found other than chronic hypercapnia from obstructive sleep apnoea.

CASE PRESENTATION

A 37-year-old female librarian presented to our hospital with a 5-month history of blurred central vision and daily, generalised headaches. She had been diagnosed in infancy with Treacher Collins syndrome, a rare congenital disorder characterised by craniofacial abnormalities and obstruction of the upper airways. She subsequently had corrective facial surgery in her teenage years to improve the expansion of her jaw. At the time of surgery, a diagnosis of obstructive sleep apnoea was made and this had since been managed with regular use of nocturnal continuous positive airway pressure (CPAP). Other medical details included childhood allergic asthma for which she took loratadine, a second generation H₁ histamine antagonist. She had no history of hypertension or diabetes, had never smoked, and was not pregnant, or on the contraceptive pill. On physical examination, she was not obese and her blood pressure was 125/70 mm Hg. There were no cranial nerve signs or limb weakness. Both pupils reacted sluggishly to light but external eye movements and visual fields were normal. Funduscopy revealed bilateral retinal haemorrhages and papilloedema (figure 1A). Visual acuity was 6/6 in the right eye and 6/9 in the left

eye. Intraocular pressure was not elevated measuring 14 mm Hg in both eyes.

INVESTIGATIONS

Routine blood tests including full blood count, inflammatory markers, renal function, liver enzymes, lipid profile, thyroid hormones and plasma glucose were all within normal limits. Arterial blood gas analysis showed hypoxia and hypercapnia with a compensated respiratory acidosis (table 1). CT scan of the brain did not show evidence of a space occupying lesion or raised intracranial pressure and CT with venous contrast excluded venous sinus thrombosis. We did not perform MRI of the brain because she had metallic implants from the previous maxillofacial surgery. A lumbar puncture performed with the patient in the left lateral position revealed an opening cerebrospinal fluid pressure of 20 cm H₂O with normal cellular and biochemical constituents.

Figure 1 (A) Funduscopy images of the patient before tracheostomy. Papilloedema before tracheostomy. (B) Funduscopy images of the patient after tracheostomy. Papilloedema improved post-tracheostomy.

To cite: Hett KL, Chutter LJ, Hassan M, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-201117

Table 1 Arterial blood gas analysis before and after tracheostomy

	Laboratory reference range	5 November 2008 pre-tracheostomy	3 August 2009 post-tracheostomy
pH	7.35–7.45	7.40	7.41
Pco ₂	4.7–6.0 kPa	7.5	4.1
Po ₂	>10 kPa	8.1	12.9
Bicarbonate	22–26 mmol/L	33.7	19.1
O ₂ Hb	95–100%	89%	97%

Before tracheostomy, the arterial blood gas analysis showed hypoxia and hypercapnia with a compensated respiratory acidosis. The arterial gases are normal after tracheostomy.

O₂Hb, oxygenated haemoglobin.

DIFFERENTIAL DIAGNOSIS

The occurrence of headaches and papilloedema in a young woman should raise a suspicion of idiopathic intracranial hypertension. However, it is important to exclude other causes of headaches and papilloedema, notably space occupying lesions, venous sinus thrombosis, hydrocephalus and malignant hypertension. These conditions were excluded in our patient by the normal systemic blood pressure together with a normal head CT. Furthermore, although our patient's presenting features suggested raised intracranial pressure, the normal cerebrospinal fluid opening pressure on lumbar puncture did not support the diagnosis of idiopathic intracranial hypertension. Moreover, she had none of the traditional risk factors for idiopathic hypertension, such as obesity or hormonal contraceptive use. Thus, having excluded other plausible aetiology we made a tentative diagnosis of papilloedema secondary to chronic CO₂ retention from obstructive sleep apnoea.

TREATMENT

We adjusted the pressure settings on her CPAP device in order to improve her oxygenation levels. This proved ineffective as she remained hypoxic with hypercapnia and continued to suffer with headaches on a daily basis. Her vision remained poor with further reduction in her visual acuity in both eyes. As there was no relief in her symptoms and arterial gas profile, a decision was taken to perform a permanent tracheostomy 2 months after presentation.

OUTCOME AND FOLLOW-UP

One month after tracheostomy, she still had disc oedema on funduscopy but 8 months later there was almost complete resolution of papilloedema associated with considerable improvement in the arterial gas levels (figure 1B). By this time her headaches had fully resolved and visual acuity had improved to 6/6 in both eyes. She remained in good health with normal vision and normal oxygen and CO₂ concentrations.

DISCUSSION

This case highlights the importance of recognising the association between obstructive sleep apnoea and papilloedema and illustrates that visual loss due to papilloedema may ultimately be reversed with effective management of hypoxia and hypercapnia.¹ Obstructive sleep apnoea is common in the general population.² Most patients are obese, but as in the case presented the syndrome can develop in non-obese individuals with upper airway obstruction. Treacher Collins syndrome is a rare autosomal dominant inherited disorder of craniofacial development seen in approximately 1/50 000 live-births. The features which

are usually evident at birth consist of downward slanting eyes, underdeveloped pinna and mandibular hypoplasia. Airway obstruction is a frequent accompaniment, and in one series obstructive sleep apnoea syndrome was reported in 46% of individuals with the syndrome. The level of obstruction can vary from the nasal septum to the trachea but the most significant obstruction usually occurs in the pharynx. Surgery at a single level is unlikely to resolve sleep apnoea since the obstruction often involves multiple levels.³

Although the cardiovascular complications of obstructive sleep apnoea syndrome are well documented,² the ophthalmic manifestations have so far received little attention. It is unclear how sleep apnoea causes papilloedema, but several mechanisms, all of which ultimately increase intracranial pressure, have been implicated.⁴ These include increased cerebral vasodilation arising from hypercapnia, elevated central venous pressure due to increased airway obstruction⁵ and the presence of systemic arterial hypertension.⁴ Although obstructive sleep apnoea syndrome is a recognised cause of intracranial hypertension, in addition, one case series showed that patients with sleep apnoea had normal intracranial pressure during the day and only developed intracranial hypertension during sleep-related apnoeic episodes.¹ The finding of a normal daytime cerebrospinal fluid pressure in our patient is in keeping with these reports and emphasises that normal intracranial pressures should not necessarily preclude the diagnosis of obstructive sleep apnoea as the cause of papilloedema.

Clinicians should be alert to the possibility of obstructive sleep apnoea syndrome in patients with unexplained papilloedema. With the current obesity epidemic, clinicians will increasingly encounter patients with sleep apnoea, many of whom will remain undiagnosed. Recognition of ophthalmic complications in such patients will be crucial in preventing permanent visual loss. Reports have documented visual improvement through effective use of CPAP, and in some cases after tracheostomy.¹ Our case illustrates that where continuous airway pressure is ineffective, a tracheostomy should be considered, as this will correct respiratory failure and can ultimately restore vision.

Learning points

- ▶ Patients with obstructive sleep apnoea syndrome may develop papilloedema and visual loss.
- ▶ A tracheostomy may be required to reverse visual loss, especially in patients with craniofacial abnormalities.
- ▶ Patients with obstructive sleep apnoea syndrome may have a normal daytime cerebrospinal fluid pressure.

Acknowledgements The authors are grateful to Dr Robin Corkill and Mr Raghu Ram for their kind advice in the preparation of the manuscript. Dr Mohamed Hassan looked after the patient along with Dr Anthony Gibson, Dr Corkill and Mr Ram.

Contributors All of the above authors were involved in drafting, writing and critically reviewing the case report. All have given approval for the final draft.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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