Headaches and papilloedema in a pregnant woman with polycystic ovarian syndrome

Adam Morton FRACP

Queensland Diabetes Centre, Mater Hospital, Raymond Terrace, South Brisbane, Queensland 4101, Australia

Summary: Idiopathic intracranial hypertension (IIH) is a rare condition, its highest incidence being in obese women of reproductive age. A woman with polycystic ovarian syndrome and IIH is presented, a new diagnosis during pregnancy resulting in a change in her management.

Keywords: neurology, obesity, sleep medicine, endocrinology

The mother has given signed informed consent for publication of this case report.

CASE REPORT

A 29-year-old primigravid woman at 25 weeks gestation with a dichorionic diamniotic twin pregnancy achieved following clomiphene induction was referred to medical antenatal clinic. Four years earlier she had been diagnosed with idiopathic intracranial hypertension (IIH) after presenting with headaches, bilateral papilloedema and early peripheral visual field loss on computer perimetry. Lumbar puncture had shown normal cerebrospinal fluid (CSF) constituents but an elevated opening pressure of 33 cm, magnetic resonance venography excluded venous sinus thrombosis. She had been treated with acetazolamide 250 mg twice a day, and had subsequently undergone laparoscopic gastric banding with a weight loss of 15 kg, with some improvement in her symptoms. Her past history was also significant for polycystic ovarian syndrome (PCOS), and anxiety treated with mirtazapine. Physical examination revealed an obese young woman with body mass index 48.5 kg/m², crowded oropharynx, neck circumference of 58 cm, bilateral papilloedema and normal visual fields to confrontation. Specific questioning revealed longstanding symptoms of snoring, headaches, daytime somnolence and nocturnal apnoeic events suggestive of obstructive sleep apnoea (OSA). Urgent polysomnography confirmed severe OSA in supine and lateral positions, apnoea hypopnoea index of 90 per hour and nadir oxygen desaturation of 74% in rapid eye movement sleep. She spent 50 minutes of the night with oxygen saturations less than 90%. Continuous positive airway pressure (CPAP) at a pressure of 16 cm of water provided good control of her OSA, and she continued on this for the remainder of her pregnancy. At 36 weeks gestation, she developed generalized itch

with elevated bile salts consistent with intrahepatic cholestasis of pregnancy. In view of her medical issues, her twins were delivered at 37 weeks gestation by caesarean section after induction of labour was unsuccessful. Both babies were well with birthweights of 2860 and 2706 g, respectively. Postpartum the mother's acetazolamide was discontinued, and she is asymptomatic with resolution of her papilloedema and normal visual fields while using CPAP despite her weight being unchanged.

DISCUSSION

IIH is a rare condition, with the highest incidence being in obese women of reproductive age (4–21 per 100,000). Pregnancy and exogenous oestrogen are thought to promote or worsen IIH. The treatment is aimed at preventing visual loss and alleviating symptoms. Carbonic anhydrase inhibitors (acetazolamide), glucocorticoids, diuretics and serial lumbar punctures have all been used to lower intracranial pressure. While acetazolamide in high doses may produce birth defects in animals, there is little clinical evidence to support any adverse effect on pregnancy outcome in humans, and it may be offered after appropriate informed consent.¹ Visual acuity and visual field testing should be performed every two months during pregnancy. The mode of delivery should be based on obstetric indications – there is no evidence that vaginal delivery or caesarean section is advantageous in IIH. Increased CSF pressure may occur with inadequate analgesia, uterine contractions, narcotic analgesia and general anaesthesia. Early regional anaesthesia is preferred (except in patients with a lumbo-peritoneal shunt), and the second stage of labour may be shortened by instrumental delivery.

IIH and OSA share common risk factors (obesity), symptoms (headache) and ocular signs (papilloedema). One study found that 37 of 53 patients with IIH had symptoms of sleep disturbance.² Fourteen of these 37 individuals underwent polysomnography, OSA being diagnosed in six and upper airway resistance syndrome in seven patients. Another study identified

Correspondence to: Adam Morton
Email: Adam.morton@mater.org.au

six (33%) patients with sleep apnoea from a group of 18 men with IIH.³ Resolution of papilloedema with CPAP therapy enabled three patients to discontinue treatment with acetazolamide, and two patients continued acetazolamide with the addition of CPAP resulting in improved papilloedema. Apnoeic events have been shown to be associated with marked elevations of intracranial pressure, and patients with severe OSA may demonstrate continuously elevated intracranial pressure.^{4,5} Thus OSA has been proposed as a recognizable cause of IIH.

OSA is highly prevalent in PCOS, having been reported in up to 70% of women with this condition.⁶ OSA has been associated with adverse outcomes in pregnancy including gestational hypertension, preeclampsia, fetal growth restriction and lower mean Apgar scores.⁷

In one study of 65 women with IIH, 37 (57%) were found to have PCOS.⁸ It has been postulated that the high prevalence of IIH in women with PCOS may be due to thrombophilia-hypofibrinolysis with subsequent thrombosis resulting in reduced CSF resorption in the arachnoid villi.

In conclusion, a pregnant woman with a past history of PCOS and diagnosis of IIH was found to have severe OSA. Her symptoms and signs of IIH resolved with CPAP treatment. As obesity is a central clinical feature of IIH, PCOS and OSA, this triad of disorders will occasionally occur by chance. As adverse pregnancy outcomes may occur in women with OSA, and CPAP treatment may result in improvement in the symptoms and signs that are common to OSA and IIH, it is important that women with PCOS or IIH who present for

preconception counselling or pregnancy care are asked about symptoms of sleep-disordered breathing.

REFERENCES

- 1 Lee AG, Pless M, Falardeau J, Capozzoli T, Wall M, Kardon RH. The use of acetazolamide in idiopathic intracranial hypertension during pregnancy. *Am J Ophthalmol* 2005;**139**:855–9
- 2 Marcus DM, Lynn J, Miller JJ, Chaudhary O, Thomas D, Chaudhary B. Sleep disorders: a risk factor for pseudotumor cerebri? *J Neuroophthalmol* 2001;**21**:121–3
- 3 Lee AG, Golnik K, Kardon R, Wall M, Eggenberger E, Yedavally S. Sleep apnea and intracranial hypertension in men. *Ophthalmology* 2002;**109**:482–5
- 4 Sugita Y, Iijima S, Teshima Y, *et al.* Marked episodic elevation of cerebrospinal fluid pressure during nocturnal sleep in patients with sleep apnea hypersomnia syndrome. *Electroencephalogr Clin Neurophysiol* 1985;**60**:214–9
- 5 Kirkpatrick PJ, Meyer T, Sarkies N, Pickard JD, Whitehouse H, Smielewski P. Papilloedema and visual failure in a patient with nocturnal hypoventilation. *J Neurol Neurosurg Psychiatry* 1994;**57**:1546–7
- 6 Sanders MH. Increased risk of obstructive sleep apnea in obese women with polycystic ovary syndrome (a review of two related articles). Articles reviewed: 'Increased prevalence of obstructive sleep apnea syndrome in obese women with polycystic ovary syndrome' and 'Polycystic ovary syndrome is associated with obstructive sleep apnea and daytime sleepiness: role of insulin resistance'. *Sleep Med* 2002;**3**:287–9
- 7 Lamberg L. Sleeping poorly while pregnant may not be 'normal'. *JAMA* 2006;**295**:1357–61
- 8 Glueck CJ, Aregawi D, Goldenberg N, Golnik KC, Sieve L, Wang P. Idiopathic intracranial hypertension, polycystic-ovary syndrome, and thrombophilia. *J Lab Clin Med* 2005;**145**:72–82

(Accepted 10 September 2010)