A patient with low pressure idiopathic intracranial hypertension and multiple cranial neuropathies

Introduction

Idiopathic intracranial hypertension is a disorder of increased intracranial pressure of unknown cause. Idiopathic intracranial hypertension can present with a wide spectrum of neurological signs, especially cranial nerve palsies, which are mostly thought to be caused by a pressure-dependent stretching mechanism. Treatment is guided by aetiology whenever possible; otherwise drainage of CSF by ventriculostomy or shunt is needed.

This case illustrates the finding of multiple cranial nerve palsies in 'low pressure' idiopathic intracranial hypertension and indicates the need to revise the currently used 'modified Dandy criteria' for idiopathic intracranial hypertension. These criteria do not allow for any focal neurological deficit other than the sixth cranial nerve palsy and do not include the occasional finding of a normal CSF pressure.

A finding of unilateral simultaneous third, fifth and seventh nerve palsies in one patient at the same time is uncommon and total resolution within minutes of a therapeutic lumbar puncture has rarely been reported.

Discussion

Idiopathic intracranial hypertension is a disorder of increased intracranial pressure of unknown cause. It is a disorder predominantly of overweight women in the childbearing years. Idiopathic intracranial hypertension can present with a wide spectrum of neurological signs, especially cranial nerve palsies which are thought to be caused by a pressure-dependent stretching mechanism. Treatment is guided by aetiology whenever detected, and at times drainage of CSF by ventriculostomy or shunt is necessary if repeated lumbar punctures are needed.

As Capobianco et al (1997) reported, cranial nerve palsies in idiopathic intra-

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Figure 1. Patient on presentation showing left pupil-sparing third cranial nerve palsy and left upper motor neuron seventh cranial nerve palsy. She also had reduced facial sensation in the cutaneous distribution of the left trigeminal nerve.

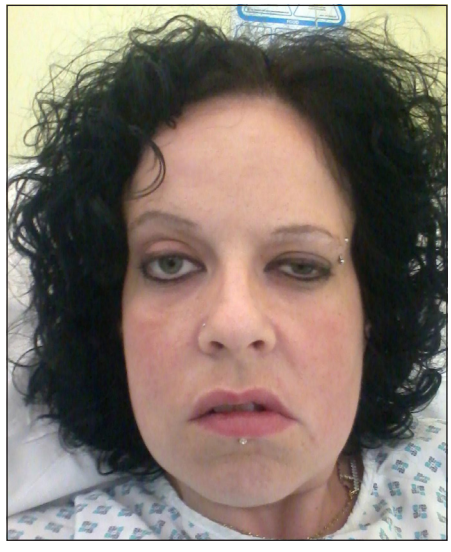
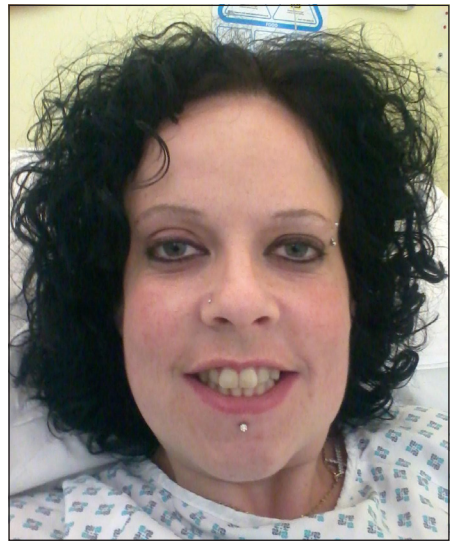


Figure 2. Patient showing resolution of her third and seventh cranial nerve immediately following removal of 12 ml of CSF by lumbar puncture.



Case Report

A 35-year-old Caucasian woman with diagnosed idiopathic intracranial hypertension presented to the emergency department with severe occipitofrontal headache and blurred vision and was found to have a left pupil-sparing third cranial nerve and left upper motor neuron seventh cranial nerve palsies (*Figure 1*). She also had reduced sensation on the left side of her face. Visual fields were normal on confrontation, papilloedema was absent and Snellen's visual acuity was 6/6. Her body mass index was 25 kg/m^2.

A provisional diagnosis of stroke was made in the emergency department, and she was given stat aspirin awaiting a computed tomography scan of the head.

Computed tomography imaging of the head was normal, and the patient was admitted under medicine. After reviewing the available records, a therapeutic lumbar puncture was performed to relieve the headache. The opening pressure was surprisingly low at 16 cm CSF. Five minutes into the procedure, the headache, blurred vision and cranial neuropathies resolved completely (*Figure 2*). A total of 12 ml of CSF was removed, with a closing pressure of 9 cm CSF. Biochemically, the CSF was acellular with protein and glucose in the normal range.

She had had two previous lumbar punctures performed since the diagnosis 4 years previously; a shunt was contemplated but never inserted. The last lumbar puncture had been performed 1 year previously, and the opening pressure was noted to be 'low' as well. She recalled having a 'droopy eyelid' with previous presentations, but the facial weakness and numbness were new for her.

The magnetic resonance imaging and cerebral venous sinus venography done at the time of initial diagnosis excluded any mechanical (e.g. venous sinus thrombosis) and anatomical (Dandy–Walker or Arnold–Chiari malformations) causes of her raised intracranial pressure. She had never taken oral contraceptive pills, and no other known confounding factors or medications were known. The patient was discharged the following day on acetazolamide and with an outpatient neurosurgical follow up.

cranial hypertension are more common in prepubertal children than in adults. Sixth nerve palsy is documented in 10–40% of patients in most cases, but third nerve palsies are uncommon (Tan, 2010).

This is an unusual case of idiopathic intracranial hypertension in which multiple (third, fifth and seventh) cranial nerve palsies resolved while a therapeutic lumbar puncture was performed to relieve the headache (Chari and Rao, 1991). CSF pressures can vary, and a normal reading in a patient with idiopathic intracranial hypertension may reflect an atypically low reading for that patient.

This case illustrates the finding of multiple cranial nerve palsies in a low pressure idiopathic intracranial hypertension and indicates the need to revise the modified Dandy criteria for idiopathic intracranial hypertension, which do not allow for any focal neurological deficit other than the sixth cranial nerve palsy and does not include the occasional finding of a normal CSF pressure. Complete recovery of multiple cranial nerve palsies after treatment indicates that these are false localizing signs and indicates a pressure-related phenomenon (Digre and Corbett, 2001).

The Friedman–Jacobson criteria for the diagnosis of idiopathic intracranial hypertension specify lumbar puncture opening pressure values that are largely based on experience with little supporting normative data. Papilloedema may not always be present in idiopathic intracranial hypertension, but the frequency of true idiopathic intracranial hypertension without papilloedema is controversial, and the threshold for diagnosing it varies among clinicians.

Concepts regarding the pathogenesis of idiopathic intracranial hypertension continue to evolve; venous hypertension is certainly implicated even though it is uncertain whether venous sinus stenosis is the cause or effect of increased intracranial pressure (Friedman, 2010). BJHM

Capobianco DJ, Brazis PW, Cheshire WP (1997) Idiopathic intracranial hypertension and seventh nerve palsy. *Headache* **37**(5): 286–8
Chari C, Rao NS (1991) Benign intracranial hypertension--its unusual manifestations. *Headache* **31**(9): 599–600
Digre KB, Corbett JJ (2001) Idiopathic intracranial hypertension (pseudotumor cerebri): A reappraisal. *Neurologist* **7**: 2–67
Friedman DI (2010) Idiopathic intracranial hypertension with Dan and beyond: the 2010 Jacobson Lecture. *J Neuroophthalmol* **30**(4): 380–5
Tan H (2010) Bilateral oculomotor palsy secondary to pseudotumor cerebri. *Pediatr Neurol* **42**(2): 141–2

LEARNING POINTS

- Idiopathic intracranial hypertension can atypically have low opening pressure on lumbar puncture.
- Idiopathic intracranial hypertension can present with multiple cranial neuropathies, hence a diagnosis of stroke should be made with caution.
- Multiple cranial nerve palsies are reported in cases of idiopathic intracranial hypertension, with sixth cranial nerve palsy the most common. Total resolution of these can occur following a therapeutic lumbar puncture as described in this case.
- The 'modified Dandy criteria' for idiopathic intracranial hypertension only allow for a sixth cranial nerve palsy and do not include the possibility of a normal CSF pressure.
- Papilloedema may not always be present in idiopathic intracranial hypertension.

Caseous calcification of the mitral valve annulus

A 73-year-old woman was referred with hypertension. Blood tests were normal. Echocardiography showed a mass arising from the mitral valve annulus (*Figure 1*).

Mitral annular calcification is fairly common in the elderly and inexplicably is predominant in women. More extreme (caseous) calcification is unusual. Lesions look smooth on echocardiogram with a rim of calcium and a central lucent region, suggesting liquefaction. The caseous 'putty-like' centre is acellular and culture negative.

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This is only the third case identified in 14 years in the authors' cardiology department (prevalence on echocardiography of <0.0007%). All three cases were remarkably similar, being slim elderly Asian women (aged 73, 78 and 81 years).

Such lesions usually follow a benign course and occasional reports describe spontaneous resolution, presumably as a result of rupture and extrusion of contents. Cardiac surgery can be considered when there is a significant obstructive element. BJHM

Figure 1. Transthoracic echocardiographic (a) parasternal and (b) apical views showing a mass (arrows) within the left atrium (2.0 x 2.3 cm). Ao = aorta; LA = left atrium; LV = left ventricle.