CASE REPORT

Multiple cranial nerve involvement with idiopathic intracranial hypertension

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Learning points for clinicians

• Idiopathic intracranial hypertension is a relatively frequent pathology (1/100 000). Papilloedema is a clinical feature in these patients. Optical coherence tomography imaging provides an objective, noninvasive and reproducible technique to assess papilloedema, which can be performed even in children, aiding the diagnosis and the monitoring.

A previously overweight (body mass index = 37.2) 14-year-old girl presented with a 2-day history of holocranial headache and 3 h of horizontal diplopia and ptosis of the right eye. She also complained of recent episodes of loss of vision in the right eye. A neurological examination revealed a right-sided ptosis with a limitation of the right inferior oblique (partial third nerve palsy). There was also an abduction deficit of the right eye (sixth nerve palsy) (Figure 1A) and left-sided facial paralysis (upper motor neurone seventh palsy). Visual acuity was 20/20 in both eyes, with normal colour vision and pupillary reactions. Visual field testing indicated generalized constriction with marked inferonasal defects. Fundus examination revealed bilateral disc swelling with circumpapillary haemorrhages (Figure 1B). These observations were confirmed with optical coherence tomography (OCT) imaging of the optic discs. There was severe diffuse thickening of the nerve fibre layer (NFL) with a loss of the typical double hunch pattern. The mean NFL thickness was 230 µm and 210 µm in the right and left eye, respectively.

Head computed tomography and magnetic resonance imaging (gadolinium enhancement) were normal. Lumbar puncture was performed, which revealed a high opening pressure (32 cm H₂O), and cerebrospinal fluid (CSF) analysis was normal. All blood tests performed presented normal ranges. A diagnosis of idiopathic intracranial hypertension (IIH) was made with involvement of the right second, third and sixth cranial nerves and left seventh cranial nerve. Acetazolamide was given at a dose of 250mg every 8 h along with a potassium supplement.
After 1 month of treatment, the headache and abduction deficit resolved, and there was mild residual papilloedema. Repeat OCT imaging revealed a mean NFL thickness of 170 μm and 150 μm in the right and left eye, respectively, with a recovery to the typical double hunch pattern. Visual fields returned to normal.

IIH is a condition defined by elevated intracranial pressure without apparent aetiology, associated with normal CSF analysis together with normal central nervous system imaging.1 IIH has been described in adults and adolescents with a female preponderance and a link with obesity, particularly recent weight gain.2 This condition has been described in children, although less frequently.3 Children with IIH may present with diplopia due to an associated nerve palsy, which usually resolves promptly with acetazolamide treatment, as seen in the presented case.4 However, the involvement of four cranial nerves (II, III, VI and VII) in the setting of IIH presented here is an exception. Surgical treatment options for IIH include optic nerve sheath fenestration and CSF diversions (lumboperitoneal or ventriculoperitoneal shunting). Bilateral papilloedema is a feature and OCT imaging provides an objective, noninvasive and reproducible technique that can be performed in children, aiding the diagnosis and the monitoring of papilloedema in the context of IIH.5 It is crucial for any child or adult presenting with sudden onset headache to examine the fundi to determine the status of the optic nerve head as visual acuity is initially normal in this condition.

Conflict of interest: None declared.

References