Intracranial hypotension causing pituitary enlargement

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DESCRIPTION

A woman aged 67 years was referred with pituitary enlargement identified on CT scan. Imaging was performed to investigate a progressive, incapacitating, 12-month history of nausea, vertigo, gait disturbance and recurrent falls. Her presentation included chronic occipitofrontal headaches, which had increased in severity and were exacerbated by upright posture. She had a history of lumboperitoneal (LP) shunting for idiopathic intracranial hypertension, systemic hypertension, ischaemic heart disease and pulmonary disease secondary to cigarette smoking. Physical examination demonstrated normal cranial nerves, bilateral upper motor neuron signs in both upper and lower limbs and severe gait instability.

Her anterior pituitary function testing showed mildly elevated prolactin levels (69.4 μg/L, range 5.2–26.7 μg/L). Adrenocorticotropic hormone (ACTH) deficiency could not be completely excluded on basal testing (ACTH 1.6 pmol/L, range 0.0–10.0 pmol/L; cortisol 245 nmol/L). Other pituitary axes were unremarkable. Formal perimetry confirmed normal visual fields. On MRI examination, the pituitary gland was homogenously enlarged (13x9x12 mm) with a convex superior margin abutting the optic chiasm. There was diffuse pachymeningeal enhancement, tonsillar herniation and a syrinx at C2 level.

A diagnosis of intracranial hypotension (IH) was made based on clinical presentation and radiological appearance (figure 1). The LP shunt was revised and her signs and symptoms gradually resolved. MRI examination performed 6 months postoperatively confirmed normalisation of her pituitary anatomy (6x9x10 mm) and resolution of the abnormal features. Basal hormonal levels were retested (ACTH 16 pmol/L; range 5.2–26.7 μg/L; cortisol 420 nmol/L) and her hyperprolactinaemia had resolved (21.9 pmol/L). This patient had acquired IH due to overdrainage of her LP shunt. IH can be misdiagnosed as pituitary hyperplasia or adenoma.

Figure 1  MRI finding reveal signs consistent with intracranial hypotension. Sagittal and coronal images T1 postgadolinium images of the pituitary gland (A–D) and sagittal T2 images through the cervical cord (E–F). Images in the left column (A, C and E) are taken during lumboperitoneal (LP) shunt overdrainage. They demonstrate pituitary enlargement of 13x9x12 mm (1), which abuts the optic chiasm (2), mild diffuse pachymeningeal enhancement (3), sagging of the midbrain, tonsillar herniation (4) and a small cervical syrinx at C2 (5). The images in the right column (B, D and F) are taken 6 months after revision of the LP shunt and demonstrate resolution of these abnormal features with pituitary gland dimensions of 6x9x10 mm.

Learning points

- Spontaneous or acquired intracranial hypotension (IH) can lead to pituitary enlargement, presumably due to engorgement of pituitary vasculatures to compensate for low intracranial pressure from cerebrospinal fluid loss.
- Hormonal dysfunction, notably hyperprolactinaemia, has been reported and may be caused by an effect of IH on the pituitary stalk.
- The cardinal signs on MRI are diffuse pachymeningeal enhancement, subdural fluid collections, engorgement of venous sinuses, pituitary enlargement and herniation of the cerebellar tonsils.

Contributors The case was a patient of KW at a public hospital clinic. AYW interpreted the clinical images. DWCC prepared the draft of this clinical picture. KW reviewed and revised the manuscript. All authors approved the final submitted version. The authors acknowledge that they participated sufficiently in the work to take public responsibility for its content.
Images in...

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