PICTORIAL MEDICINE

Spontaneous intracranial hypotension: uncommon but important

Case report

A 44-year-old woman presented with sudden onset of occipital headache and neck stiffness for 3 weeks. The headache was pulling in character and worse in the morning (visual analogue scale pain score of 8). Coughing, straining when passing a stool, and postural change (either going from a lying to an erect position or vice versa) provoked the headache. It was associated with vertigo, nausea, and vomiting. She gave no history of trauma to her head and neck and had not experienced any fever, rashes, photophobia, tinnitus, or deafness. She did, however, notice night sweats, anorexia, and subjective weight loss. She had undergone LASIK for myopia and her medical history was otherwise unremarkable. She did not use oral contraceptive pills.

She consulted a private practitioner and a traditional Chinese practitioner without significant relief of her symptoms. She then attended the accident and emergency department of a public hospital 1 week after the onset of her headache. A neurological examination performed at that time elicited no abnormalities. She was afebrile and her vital signs were stable. Postural hypotension was not elicited. Computed tomography (CT) of her brain was normal. She was discharged with prochlorperazine, betahistine, diclofenac sodium slow release, tramadol, and famotidine.

These medications did not relieve her symptoms and the headache spread to her forehead. She revisited the same accident and emergency department 2 weeks later and was admitted. A detailed neurological examination again found no abnormalities. She had no papilloedema, neck rigidity, cranial nerve deficits, long tract signs or cerebellar signs. Multiple drugs, including propoxyphene/paracetamol, naproxen, amitriptyline, nortriptyline, bromazepam and diazepam were tried, without success. Neck physiotherapy relieved her neck stiffness but not her headache.

Blood tests, including a full blood count, erythrocyte sedimentation rate, C-reactive protein, clotting profile, vasculitic markers, tumour markers, liver and renal functions were all normal. A repeated CT of the brain showed a left subdural fluid collection and effacement of the basal cistern (Fig 1). A lumbar puncture performed in the lateral recumbent position revealed an opening pressure of 50 mm H_2O, a red blood cell count of 12 /mm^3, a white blood cell count of 6 /mm^3, a protein level of 0.57 g/L and a glucose level of 4.1 mmol/L (the paired serum glucose level was 5.9 mmol/L). A Gram smear and bacterial culture, acid-fast bacillus smear and culture, and cytology for malignant cells were all negative. Digital subtraction angiography of the cerebral vessels was completely normal.

Magnetic resonance imaging (MRI) of the brain, performed after the CT scans, found the following interesting features: (1) bilateral subdural fluid collections; (2) cerebellar tonsillar ectopia resembling a type I Chiari malformation (Fig 2); and (3) diffuse, smooth contrast enhancement of the intracranial dura (Fig 3).

These findings were typical of spontaneous intracranial hypotension (SIH). Therefore, contrast magnetic resonance myelography (intrathecal 0.7 mL Dotarem, a gadolinium-containing contrast agent [0.5 mmol/mL], off-label use) was performed, which demonstrated a cerebrospinal fluid (CSF) leak on the left side of the thecal sac at the S1/2 level (Fig 4). Initially we planned to perform an epidural blood patch but her headache and subdural collections

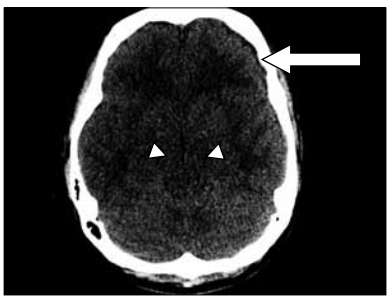

FIG 1. Left subdural fluid collection (arrow) and effacement of the basal cistern (arrowheads)

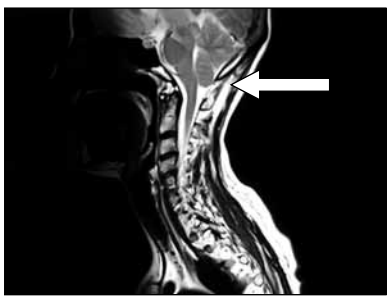

FIG 2. Cerebellar tonsillar ectopia resembling a type I Chiari malformation (arrow)

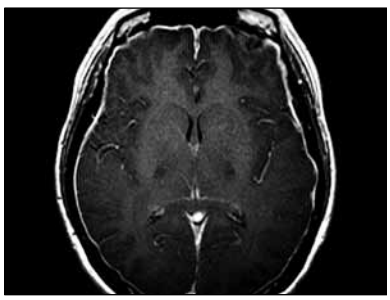

FIG 3. Diffuse, smooth contrast enhancement of the intracranial dura

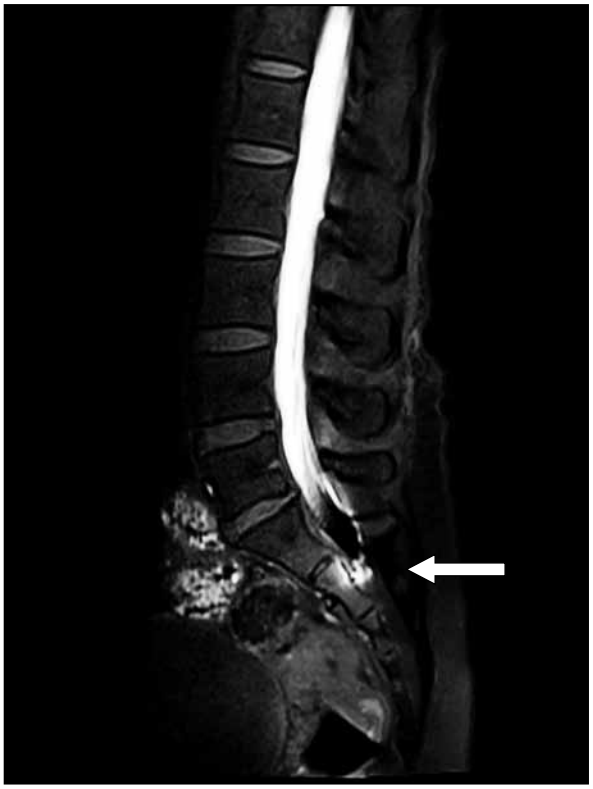

FIG 4. Cerebrospinal fluid leak demonstrated by MR myelography at S1/2 level (arrow)

resolved spontaneously after bed rest. She had no connective tissue disorder features.

Discussion

Spontaneous intracranial hypotension is an uncommon cause of headache, yet clinicians should familiarise themselves with its clinical and radiological features. Misdiagnosis not only causes delayed treatment, but also exposes patients to unnecessary, high-risk diagnostic and therapeutic procedures. In 18 patients studied by Schievink,[1] 17 (94%) received an initial incorrect diagnosis and the median diagnostic delay was 5 weeks. Some patients had undergone invasive procedures including meningeal biopsy, a decompressive craniotomy for Chiari malformation and the evacuation of subdural collections.

Positional headache is the clinical hallmark of SIH. It is likely to be due to the reduction in CSF volume and downward displacement of the brain, causing traction on pain-sensitive structures such as the dura. Alternatively, it may involve compensatory dilatation of the pain-sensitive intracranial venous structures. Occipital or suboccipital regions are commonly affected although any other sites are possible. The onset of headache can be gradual or acute which, together with neck stiffness, mimics subarachnoid haemorrhage or meningitis. The International Headache Society has published diagnostic criteria[2] for "headache attributed to spontaneous (or idiopathic) low CSF pressure" (Section 7.2.3).

Associated neurological symptoms are common and may mimic other more serious neurological conditions. Examples include neck pain or stiffness, nausea and vomiting, photophobia, vertigo, hearing loss, and diplopia. These symptoms are either due to meningeal irritation or stretching of the cranial nerves. Patients may even develop coma.

Five characteristic imaging features can be visible on MRI. The mnemonic is SEEPS[3]—(1) subdural fluid collections; (2) enhancement of the pachymeninges; (3) engorgement of venous structures (dural venous sinuses or large cerebral veins); (4) pituitary hyperaemia; (5) sagging of the brain (bowing of the optic chiasm over the pituitary fossa, flattening of the pons against the clivus, descent of the cerebellar tonsils mimicking a Chiari type I malformation). To define the location and extent of the CSF leak, CT or magnetic resonance myelography is the investigation of choice. Alternatively, radionuclide cisternography may be used to study the CSF leak.

YF Cheung, FHKAM (Medicine)
Email: cyfz02@ha.org.hk
Division of Neurology, Department of Medicine
KW Tang, FRCR
Department of Radiology and Imaging
Phillip YC Chan, MRCP
Division of Neurology, Department of Medicine
YW Wong, FHKAM (Surgery)
FC Cheung, FRCS
Department of Neurosurgery
John HM Chan, FRCP
Division of Neurology, Department of Medicine
Samuel CL Leung, FRCS
Department of Neurosurgery
Patrick CK Li, FRCP
Division of Neurology, Department of Medicine
Queen Elizabeth Hospital
Kowloon, Hong Kong

References

1. Schievink WI. Misdiagnosis of spontaneous intracranial hypotension. Arch Neurol 2003;60:1713-8.
2. Headache Classification Subcommittee of the International Headache Society. The International Classification of Headache Disorders. 2nd ed. Cephalalgia 2004;24 Suppl 1:9-16.
3. Schievink WI. Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. JAMA 2006;295:2286-96.