Spontaneous intracranial hypotension: a cause of severe acute headache

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Severe acute headache is a common presenting symptom to an accident and emergency department. Spontaneous intracranial hypotension (SIH) is an increasingly recognised cause of these symptoms and has characteristic clinical and imaging findings. SIH is characterised by headache worse on standing, low opening cerebrospinal fluid pressures at lumbar puncture and uniform pachymeningeal enhancement with gadolinium enhanced magnetic resonance imaging of the brain, all in the absence of dural trauma. Atypical presentations occur and severe neurological decline can rarely be associated with this condition. A review of five patients presenting recently to our institution with classical imaging findings together with a review of the literature is presented.

Patients presenting to the accident and emergency department with severe acute headache are commonly treated as subarachnoid haemorrhage until proven otherwise. The differential diagnosis in patients with negative computed tomographic (CT) scan and lumbar puncture includes spontaneous intracranial hypotension (SIH).

SIH is characterised by headache worse on standing, low opening cerebrospinal fluid (CSF) pressures at lumbar puncture, and uniform pachymeningeal enhancement with gadolinium enhanced magnetic resonance imaging (MRI) of the brain, all in the absence of dural trauma. Atypical presentations occur and severe neurological decline can rarely be associated with this condition. We report on five cases of spontaneous intracranial hypotension, the clinical presentations, imaging findings, opening pressures at lumbar puncture, and CSF analysis and provide

a review of the recent literature on the condition.

CASES

The clinical and radiological presentations of five patients (three female, two male) attending our institution between November 2004 and May 2006 with a diagnosis of SIH were reviewed retrospectively (table 1). Four of the patients had first presented via the accident and emergency department. All patients had normal CT scans of the brain and then underwent MRI. T1 (TE 14 ms, TR 500 ms) pre- and post-contrast and T2 (TE 86 ms, TR 3000 ms) sequences were obtained using a Siemens 1.5 Tesla Magnetom Symphony MRI scanner (Maestro Class, Erlangen, Germany). The contrast agent used was Prohance (gadoteridol) 0.2 ml/kg up to a maximum of 15 ml.

All patients showed uniform thickening (fig 1) and enhancement of the

pachymeninges following intravenous contrast administration (figs 2 and 3). One patient was noted to have tonsillar herniation (fig 4) and dilatation and enhancement of the cervical epidural venous plexus. All patients had further imaging of the spine to look for possible site of CSF leak, but none were found.

DISCUSSION

SIH was originally described by Schaltenbrand in 1938 and the MRI findings were later described in 1991.^{1,2} The clinical features are those that occur after dural puncture but arising spontaneously. Orthostatic headache relieved on lying down is the most common presentation. This contrasts with the headache of meningitis which is exacerbated by lying down. However, cranial nerve palsies (VI, VII and VIII), nausea, vomiting, coma, tinnitus, vertigo, cervical myelopathy and parkinsonism can be presenting symptoms.³ Most, but not all, series report a female predominance of up to 3:1. Most case reports are of female patients and our series conforms with most of the published data with a female to male ratio of 3:2.¹⁻⁴ The incidence of SIH has been reported as being between 1 in 50 000 and 5 in 100 000, although comprehensive epidemiological data are not yet available.^{5,6}

SIH diagnosis has been thought to require a CSF opening pressure <60 mm H₂O and the absence of a dural tear. However, more recently, several papers have postulated that the physiology of the condition is based on hypovolaemia of CSF rather than its reduced pressure within the central nervous system.^{4,7} Three of our patients had low opening CSF pressures at lumbar puncture. One patient required

Table 1 Patient details

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Presenting complaint	Orthostatic headache	Nose pain. Worse on Valsalva manoeuvre	Orthostatic headache, nausea, neck stiffness, photophobia	Orthostatic headache, nausea, vomiting	Orthostatic headache, vomiting
Onset of symptoms	Gradual 2 months	Gradual 3 months	Gradual 1 week	Sudden	Sudden
CSF opening pressure	18 mm H ₂ O	No lumbar puncture	60 mm H ₂ O	18 mm H ₂ O	10 mm H ₂ O
CSF findings	Nil	Nil	Raised protein	Nil	Raised protein
Imaging findings	Uniform pachymeningeal enhancement	Uniform pachymeningeal enhancement	Uniform pachymeningeal enhancement	Uniform pachymeningeal enhancement	Uniform pachymeningeal enhancement
Management	Steroids—no benefit. Considering dural patch	Considering dural patch	Rehydration, rest, analgesia, laxatives. Awaiting blood patch	Oral prednisolone—symptoms return when dose decreased	Considering dural patch

CSF, cerebrospinal fluid.

Figure 1 (A) T2 weighted axial image showing thickened pachymeninges of the posterior falx (arrow). (B) T2 weighted axial image at the same level as in (A) showing normal appearances of the falx.

six attempts due to repeated “dry tap”, and another patient had an opening pressure of 60 mm H₂O but otherwise classical clinical and imaging findings of SIH.

The Monro–Kellie hypothesis that the sum of the volumes of the brain, CSF and intracranial blood are constant at any time, but are in a state of constant flux, is the mechanism frequently used to explain the MRI findings in this condition. A reduction in the volume of the CSF will require an increase in volume of one or both of the other components. The most reliably demonstrated area of increased volume on imaging is the pachymeninges which show diffuse thickening and enhancement with gadolinium enhanced MRI due to lack of blood–brain barrier and an increase in the

volume of venous blood in this compartment.⁸

The patients have variously been treated with analgesia, bed rest, caffeine, steroids and senna and lactulose (to avoid constipation). The success of conservative treatment has been limited and blind epidural blood patch is being considered by four patients in the absence of a demonstrable target on MRI.

The differential diagnosis of diffuse meningeal enhancement includes infective or inflammatory meningitis, metastasis and post-surgical changes, and adequate clinical history may be the only way to refine the diagnosis. Other demonstrable mechanisms of compensatory volume increase include engorgement of the dural venous sinuses, enlargement of epidural venous plexuses, subdural fluid collections and

enlargement of the pituitary gland. The imaging findings are reversible and resolution of abnormal imaging usually accompanies a cessation in the patients’ symptoms.⁷

As well as the effect on the volume of the other intracranial structures, the CSF has a buoyant effect and with its volume diminished can lead to sagging of the brain within the cranial cavity causing traction on the cranial nerves, epidural bridging veins, and descent of the cerebellar tonsils. The vessels in the cranium are sensitive to stretching and it is traction on these structures that is thought to result in the orthostatic headache.⁴

Once the diagnosis has been made or confirmed with brain MRI, further imaging is aimed at demonstrating a site of CSF leakage. It has been proposed that patients with Marfan syndrome and other connective tissue diseases are more likely to have CSF leaks due to dural ectasia seen in these conditions.⁹ The most common sites are found in the cervical and lumbar spine, and MRI, CT myelography and radio-nuclide myelocisternography have all been used to locate the CSF leak. On MRI the site of leak is seen as a non-enhancing or mildly enhancing epidural fluid collection on gadolinium enhanced images.^{10 11}

Management of SIH is dependent on the severity of the patients’ symptoms and how well they respond to conservative treatments. Bed rest, analgesia, adequate hydration, caffeine and oral corticosteroids can be used in the acute stage. Epidural blood patch, CT-guided fibrin glue injection at the site of the leak, and open surgical intervention are used in the chronic phase.¹² In the majority of patients the disease has a benign course; however, obtunded patients are seen in SIH and there has been at least one death ascribed to this condition. The cause of obtundation is transtentorial herniation and in these cases of severe neurological decline lumbar intrathecal saline infusion has been used to arrest or reverse the patients’ demise. Subdural collections (blood or serous fluid) are documented and may require neurosurgical drainage.¹³

All our cases except one presented via the accident and emergency department with symptoms of sudden onset headache and a strong postural relationship, being worse on standing. The differential diagnosis for these patients on presentation to an accident and emergency department includes subarachnoid haemorrhage, meningitis and post-coital headache. The differential diagnosis for a more chronic presentation would include colloid cyst, cerebral

Figure 2 (A) T1 weighted post-contrast coronal and axial images respectively showing thickening and enhancement of the pachymeninges of the falx and tentorium cerebelli (arrows). (B) T1 weighted post-contrast coronal and axial images respectively showing normal dural enhancement.

Figure 3 (A) T1 weighted post-contrast coronal and axial images respectively showing thickening and enhancement of the pachymeninges of the falx and tentorium cerebelli (arrows). (B) T1 weighted post-contrast coronal and axial images respectively showing normal dural enhancement.

Figure 4 T2 weighted sagittal image showing tonsillar herniation (arrow).

venous sinus thrombosis and headache associated with positional vertigo. Careful history taking, in particular the postural relationship of the headache, should alert the clinician to the possible diagnosis of SIH.

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