

An Acute Severe Headache

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Keywords

Spontaneous Intracranial Hypotension, Headache, Subarachnoid Haemorrhage

Key Points

- Headaches worse on erect posture should alert clinicians to intracranial hypotension.
- Cranial imaging, including MRI, may be entirely normal at the time of initial presentation.
- Treatment of spontaneous intracranial hypotension should include bed rest and consideration of an epidural blood patch if the symptoms fail to resolve.

Answer

1. The MRI scan shows bright meninges and bilateral subdural collections (see Figure 3)
2. The likely diagnosis is spontaneous intracranial hypotension
3. Initially bed rest is recommended; if the headache persists, an epidural blood patch may be required.

Discussion

Severe headaches are a frequent presentation to the Emergency Department, accounting for around 1% of presentations.¹ Of these, 13–16% will have a sinister underlying pathology¹ but accurate diagnosis of headaches on the medical take remain difficult for non-specialists. A retrospective study from the UK found that, of those presenting to Emergency Department with sudden onset severe headaches, only 60% were given a specific headache diagnosis, with one third of these given their diagnoses despite an inadequate history.²

Low pressure headaches are commonly recognised following iatrogenic dural puncture; it is far less common to see these symptoms in the absence of such a procedure.³ The literature is made up almost entirely of case reports. This is partly due to its often non-specific clinical presentation as well as the lack of any diagnostic tests.

Primary low pressure headaches are thought to be due to spontaneous CSF leaks: mechanical stresses cause dural tears or provoke meningeal diverticula.⁴ The lower cervical and upper thoracic spine are particularly vulnerable and in some cases they can be associated with underlying connective tissue disorders.⁵ Case reports often describe some sort of precipitant such as heavy lifting (weightlifting as in our case) but also straining, stretching, coughing or exercise.⁶

The hallmark of intracranial hypotension is postural or orthostatic headache: headache that is exacerbated on

upright posture and relieved on supine posture. In our case, the headache reached maximal intensity within seconds of upright posture and so such an association is easy for patient and clinician to identify. Unfortunately, this is not always the case⁷ and so this cardinal clinical feature may be difficult to illicit. The headache is commonly described to localise in the occipital area⁸ but can be more generalised. Its severity does not correlate well with the degree of intracranial hypotension on investigation parameters⁹ and it is very likely that many milder cases go undiagnosed. To make clinical diagnosis more difficult, Intracranial Hypotension can mimic more familiar and dangerous headache syndromes. Focal neurology in the form of cranial nerve palsies¹⁰ have been described, as has confusion and coma,¹¹ tremor,¹² neck stiffness¹³ and “acute thunderclap headaches”.¹⁴ The non-specific nature of the condition in combination

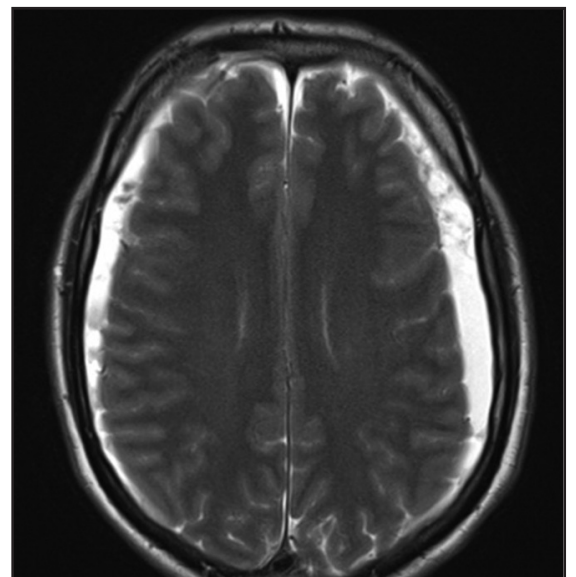


Figure 3. T1 weighted cranial MRI showing bilateral subdural collections and bright meninges

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with unfamiliarity amongst non-specialist clinicians means that many cases are likely misdiagnosed.

Cranial CT proves insensitive to features of intracranial hypotension though MRI can show a number of features including diffuse meningeal enhancement, venous sinus engorgement, subdural fluid collection, pituitary gland enhancement and downward displacement of the brain.⁸ T1 weighted midline sagittal views best demonstrate downward displacement of the brain and gadolinium enhanced T1 weighted coronal views through the sella and pituitary best demonstrate pachymeningeal enhancement.⁷ If attempting to localise the CSF leak, CT Myelography is often considered to be the gold standard,¹⁶ however heavily T2 weighted MR sequences of the spine have been shown to be at least as sensitive¹⁷ and have the benefit of being non-invasive and more readily available. In our case, MRI features of intracranial hypotension were not seen until 3 weeks after clinical presentation. One study suggests that spinal MR changes might precede brain MR changes particularly if investigating early¹⁸ however we are not aware of any studies exploring the value of repeat imaging.

Lumbar Puncture opening pressures of less than 60mmH₂O in the lateral decubitus position is typical of intracranial hypotension however normal pressures do not exclude the diagnosis.¹⁹ A wide variety of factors have been proposed to account for this, such as time and compensatory mechanisms as well as the theory that it is decreased CSF volume rather than CSF pressure that is the underlying the pathology.²⁰ Biochemical CSF analysis might demonstrate elevated protein, leukocytosis and even xanthochromia, thought to reflect changes in dilated meningeal blood vessel permeability and reduced

CSF flow.^{21,22} Ultimately, MRI and Lumbar Puncture is at best 80% sensitive in severe cases;¹⁹ negative findings do not exclude the diagnosis and investigations may mimic other headache syndromes so must be interpreted in the context of clinical suspicion.

Once a diagnosis of Spontaneous Intracranial Hypotension is made, management strategies are largely based on anecdotal evidence. In most cases conservative measures with strict bed rest and hydration are reported to help.²² If conservative measures fail then autologous epidural blood patching could be considered. This describes the 'blind' (i.e. without knowing the exact site of the CSF leak) injection of 10–20 mls of the patient's blood into the epidural space. The blood is thought to provide volume replacement and cause dural tamponade sometimes giving immediate relief to patients.²² Such is its efficacy; response to epidural blood patching is considered a diagnostic criterion in the International Classification of Headache Disorders, 3rd edition (beta version).¹⁵ Targeted blood patching can be performed if the site of the leak is discovered though it is not clear if this more effective than blind blood patching. Unfortunately, blood patching in Spontaneous Intracranial Hypotension is not as effective as when applied to iatrogenic dural puncture, with success rates of only around 30% rising to 50% with repeated procedures.²³ Other strategies before surgical intervention have been reported in case reports, such as multi-level blood patching,²⁴ glucocorticoid steroid therapy²⁵ and intrathecal saline infusions.²⁶

Conflict of interest

Nothing to declare.

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