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Review

Spontaneous low pressure headache – A review and illustrative patient

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ABSTRACT

Low pressure headache typically occurs as a complication of dural puncture. "Spontaneous" low pressure headache is a relatively rare but under-recognised cause of intractable headache. Clinical suspicion of this condition warrants imaging of the brain to confirm the diagnosis; spinal imaging may be needed to identify the site of the leak. Epidural blood patching may be necessary to seal the leak – CT fluoroscopy may be helpful in delivering the patch directly to the site of the leak. Surgical intervention may be required in intractable cases. We describe a patient with spontaneous intracranial hypotension and review the clinical and radiological features of this syndrome.

1. Introduction

Cerebrospinal fluid (CSF) hypovolaemia after a dural puncture is a well-recognised cause of postural headache, and is most often the result of lumbar puncture. Apparently spontaneous CSF leak due to a dural tear can give rise to severe postural headache and other symptoms. Minor trauma and underlying connective tissue disorders may predispose to spontaneous intracranial hypotension (SIH) by causing a dural tear. Although SIH is a rare cause of headache, it is under-recognised, which results in unnecessary investigations and delayed diagnosis and treatment. Although the clinical presentation of this condition varies widely, characteristic neuroradiological features may be helpful in confirming the diagnosis, localising the leak and sealing it. We report an illustrative patient, a young woman who developed SIH after minor trauma. Neuroimaging was helpful in confirming the diagnosis, and the site of CSF leak was identified by CT myelography. Introduction of blood patches at the conventional lumbar site was ineffective, but CT fluoroscopy facilitated blood patching to the thoracic site of the leak, which was successful, and obviated the need for surgical intervention. Our patient has remained symptom free for more than two years.

2. Methods

The material covered in this review is based on systematic review of journal articles in PUBMED (1950 to 2010) using the terms "low pressure headache", "spontaneous intracranial hypotension", "CSF leak", "CSF hypovolemia" and "postural headache". Selected articles were predominantly those published in the last 15 years but older articles, which were frequently referenced, were also included.

3. Illustrative patient

A 38-year-old woman presented with extremely severe headache, neck discomfort, tinnitus and vomiting within hours after bending to pick up a toy from the floor. She looked distressed, but there were no abnormal signs. The pain was much worse when she sat or stood, and was bearable in the recumbent position only if regular morphine was administered. She recalled minor lower back pain during a netball match six months earlier and had suffered from Meniere's disease since the age of 20 years. Her general health was good.

Brain MRI with contrast showed pachymeningeal thickening and enhancement (Fig. 1). Her spinal MRI revealed a large epidural CSF collection, which extended over numerous segments (Fig. 2). A shallow disc protrusion was seen at T12/L1.

Despite three weeks of strict bed rest, hydration, analgesia, antiemetics and caffeine administration, our patient remained prostrated. A conventional lumbar epidural blood patch (EBP) was performed without benefit on three occasions. A radio-isotope study did not demonstrate a CSF leak, but contrast CT myelography revealed a leak at T12–L1, presumably due to a dural tear at the site of the disk protrusion (Fig. 3). A targeted blood patch was performed at this level under fluoro-CT guidance. The headache

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Fig. 1. Coronal T1-weighted MRI brain with gadolinium of a 38-year-old woman with spontaneous low pressure headache showing diffuse enhancement of the pachymeninges.

resolved completely within 24 hours, but transient radicular pain occurred in the legs.

About five weeks later, intolerable headache recurred after the patient stumbled. A directed blood patch under image intensification was again successful; radicular irritation was avoided by gabapentin and prednisone cover. The patient has since remained headache free for more than two years.

Fig. 2. Sagittal T2-weighted MRI of the spine showing extensive cerebrospinal fluid leak.

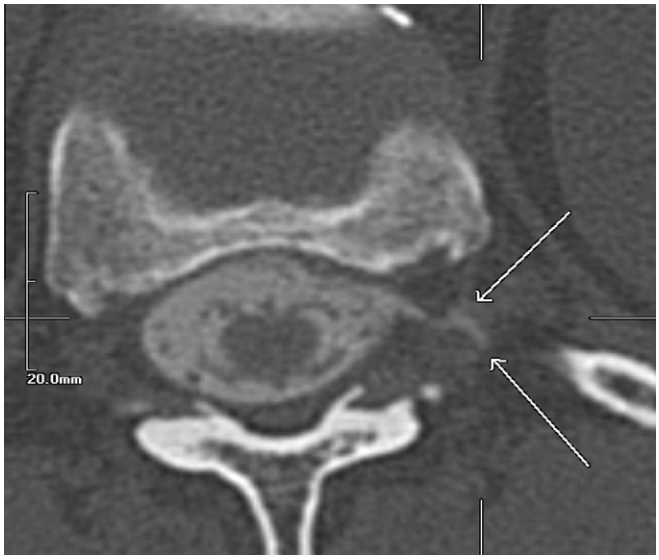

Fig. 3. Axial CT myelogram with contrast demonstrating an active cerebrospinal fluid leak (arrow) at T12–L1, presumably due to a dural tear at the site of the disk protrusion.

4. Epidemiology

The estimated annual incidence of SIH is 5 per 100,000 according to an emergency department-based study.[1] Data from comprehensive population-based studies are not available. SIH occurs more frequently in women, with a female-to-male ratio of approximately 2:1. It is more common in young and middle-aged individuals but reports in children and the elderly have been described. In a recent case series, chronic subdural haematoma in elderly individuals on oral anticoagulation has been a manifestation of spontaneous CSF leak; the haematomas resolved after treatment of the CSF leak.[2] Such cases might be under-recognised. Peak incidence is in the fourth decade. Individuals with underlying connective tissue disorders are predisposed to spontaneous dural tears resulting in SIH. Minor trauma in the days or weeks prior to the presentation of SIH is not uncommon.

5. Pathophysiology

Spontaneous low pressure headache was described by Schaltenbrand in 1938, and results from CSF leak at a spinal level.[1] CSF hypovolemia results in reduced buoyancy of the brain, which leads to sagging of the brain in the cranial vault and stretching of pain sensitive structures. This gives rise to the postural symptoms characteristic of this condition. The intracranial volume is compensated for by dilation of intracranial vessels. According to the Monroe-Kellie hypothesis, intracranial volume, which is a sum of brain parenchyma, cerebrospinal fluid and vascular components, remains constant.[3] CSF hypovolemia due to CSF leak is compensated for physiologically by an increase of vascular component (dilatation of compliant intracranial blood vessels) as brain parenchymal volume remains constant. Vascular congestion may contribute to headache.[4]

In the presence of intravenous contrast, the dilated pachymeningeal vessels appear to be congested as they are outside of the blood–brain barrier (BBB). However, leptomeningeal vessels have an intact BBB, and should not enhance in SIH. If they do so, an alternative diagnosis should be considered. Subdural fluid collections (hygromas) may form to restore the reduced intracranial volume.[5]