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Spontaneous Intracranial Hypotension: Underdiagnosed and Undertreated

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Abstract

Presentation
A 27 year old female presents with a six week history of back pain, postural headaches, blurred vision, and tinnitus.

Diagnosis
Spontaneous intracranial hypotension (SIH) secondary to a spinal cerebrospinal fluid (CSF) leak.

Treatment
Surgical repair of the dural defect.

Discussion
SIH is underdiagnosed and poorly managed, leading to considerable patient morbidity. Recognition of the clinical signs and symptoms, and the performance of appropriate investigations in the form of non-invasive and invasive imaging, is key to the diagnosis and treatment of SIH.

Introduction

First described in 1938, spontaneous intracranial hypotension (SIH) is caused by loss of cerebrospinal fluid (CSF) from the spinal canal, and results in postural headaches clinically, and in rare cases can lead to coma and death[1]. Incidence is estimated at 5 per 100,000, but this may be an underestimation as SIH is frequently misdiagnosed as migraine, tension headache, viral meningitis, or malingering[2].

Case Report

A 27 year old female presents with a day history of back pain, postural headaches, blurred vision, and tinnitus. Her headaches were worse on sitting up and relieved by lying flat, and of such severity that she could not function normally and spent all of her time lying down. She

has no history of connective tissue disorders. Initial non contrast CT brain revealed no abnormalities, but an MRI brain with contrast performed six weeks later due to unremitting symptoms revealed bilateral subdural collections, pachymeningeal enhancement, engorgement of the venous sinuses, a reduced mamillopontine distance, and effacement of the suprasellar and prepontine cisterns, strongly suggestive of SIH (Figure 1). An MRI of whole spine revealed a spinal longitudinal extradural CSF collection (SLEC) in the lumbar spine. An initial digital subtraction myelogram (DSM) re-demonstrated contrast in the ventral epidural space but could not locate a definite point of CSF leak. Repeat DSM four weeks later identified a CSF leak ventrally in the midline at the level of T8/9 (Figure 2).

She was brought to the operating theatre for a CSF leak repair. A T8 laminectomy was performed via a posterior midline approach, followed by a midline durotomy. The spinal cord was gently mobilised and rotated to reveal a pinhole defect in the dura ventrally, but this defect could not be safely closed primarily with a stitch due to the need for unacceptable levels of retraction on the spinal cord with risk of neurological injury. A dural patch was employed instead and the durotomy closed in a water-tight fashion. The patient recovered both clinically and radiologically and was discharged well.

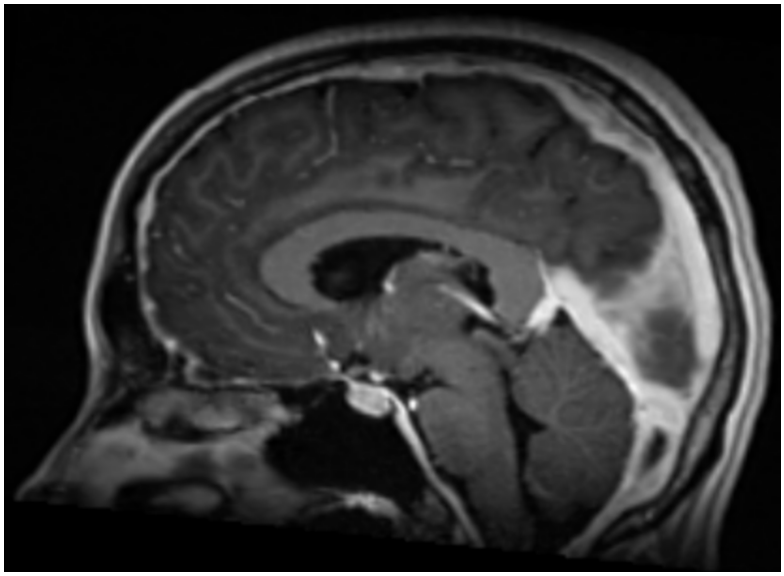

Figure 1: Sagittal T1 MRI Brain with contrast showing the features of intracranial hypotension.

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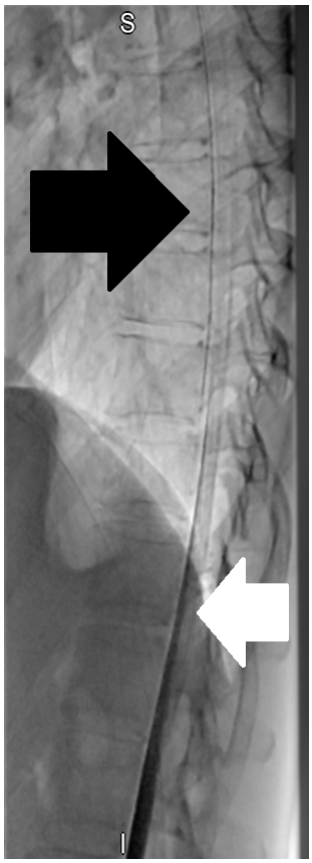

Figure 2: Lateral DSM demonstrating intrathecal contrast (white arrow), and contrast in the extradural space (black arrow).

Discussion

SIH is diagnosed according to the International Classification of Headache Disorders (ICHD-3) clinically in patients with an orthostatic headache, in the absence of trauma known to cause CSF leakage, and with either a low opening pressure on lumbar puncture or typical radiological signs of intracranial hypotension[3]. The Bern score, based on features found on contrast enhanced MRI brain, is used to predict the probability of identifying a CSF leak or CSF-venous fistula at myelography, and our patient meets all the criteria[4].

Three types of CSF leaks have been described. Type 1 is caused by a ventral dural defect thought to be caused by an osteodiscogenic spur which was the case in our patient. Type 2 is caused by a defect as the nerve root sleeve exits the thecal sac. Type 3 is caused by a direct fistula between the thecal sac and surrounding epidural veins. Type 1 leaks may result in a SLEC being identified on spinal imaging as was in our case, and the defect, often the size of a pinhole, is identified by specialised DSM in a head-down and prone position as the contrast leaks while flowing cranially. Type 2 and 3 leaks may not result in a SLEC and occur laterally in which the DSM needs to be performed in a lateral position to identify the pathology[1].

Conservative management of SIH consists of bed rest, oral hydration, caffeine, and use of an abdominal binder. Traditionally, intervention was by injection of autologous blood into the spinal extradural space, also known as a blood patch[2]. This technique is effective for CSF leaks caused by spinal taps as the defect is located dorsally and could be "patched", but is unlikely to benefit ventral defects, particularly where the defect is several levels above the lumbar spine, as was in our case. Type 1 and 2 leaks refractory to blood patches and conservative therapy are treated with microsurgical repair while type 3 leaks are treated with radiological embolization.

In conclusion, SIH is a condition caused by spinal CSF leak, is not uncommon, and can be challenging to diagnose and treat. SIH should be suspected in the setting of an orthostatic headache. Neuroimaging followed by specialist referral and workup can lead to the appropriate diagnosis, treatment, and a good outcome.

Declarations of Conflicts of Interest:

None declared.

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