Diagnosis and management of SIH

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Background-Spontaneous intracranial hypotension (SIH) defines a clinical condition characterized by debilitating postural/orthostatic headaches secondary to spontaneous spinal cerebrospinal fluid (CSF) leak and/or CSF hypotension.

Introduction -International Classification of Headache Disorders (ICHD-3), SIH is diagnosed when headache has developed spontaneously and in temporal relation to a CSF leak (evident on imaging) and/or CSF hypotension (lumbar puncture with measuring opening pressure.

Headache is the main and most common symptom of SIH. Pain usually progresses rapidly over a few hours. The typical headache is bilateral (but headache can be unilateral) generally occipital–nuchal, orthostatic, and identical to the headache of post lumbar puncture syndrome. Thunderclap headache can also occur and confuse the picture. Orthostatic headache might worsen after a few seconds/minutes or several hours of the patient being upright, and might improve or disappear after a few seconds/minutes/hours of rest in supine position. The orthostatic nature might become less obvious over time. Headache could be associated with nausea/vomiting, neck stiffness, and cochlear–vestibular signs including tinnitus, ear fullness, echoing, or distortion of sounds, hypoacusia, dizziness, or even rotational vertigo.

SIH is an underdiagnosed disorder, mainly because absence of orthostatic headache, normal imaging findings, or normal lumbar puncture opening pressures can occur in SIH; therefore, this diagnosis cannot be excluded in patients who do not present with all the typical features of this disorder.

Diagnostic imaging - MRI with gadolinium contrast of the brain and spine considered initial imaging modality. Diffuse pachymeningeal enhancement is most common MRI feature but there is other finding with different specifity and sensitivity. MRI of the spine without gadolinium is typically performed along with brain MRI to identify features suggestive of CSF leak.

Other diagnostic imaging such as MR myelography, CT myelogram or radioisotope cisternography might be indicated if:

1. Initial brain and spine MRI are abnormal but non-diagnostic

2. Clinical suspicion for the diagnosis is high despite normal initial imaging.

CT myelography is the most reliable test to show the exact site of the leak.

Lumbar Puncture – LP can document low CSF pressure in suspected cases of SIH. However, CSF pressure may be normal even in the presence of an active leak.

Treatment - Management of SIH relies on observational data and expert opinion and all patients need initial symptomatic management. Overhydration, corticosteroids, Caffeine and theophylline used with various efficacy. Epidural Blood Patch (EBP) is the gold standard initial treatment. It is usually performed under fluoroscopic guidance with an autologous blood volume ranging from about 30–50 mL mixed with 5 mL of iodinated contrast agent. When the first EBP does not result in clinical benefit, it is possible to perform one or two additional patches at least 7 days apart from each other.

Complication- Early complications of SIH should be suspected in any patients reporting changes in headache, such as the pain becoming non-postural, which may suggest intracranial hypertension. Subdural hematomas are not an infrequent complication of SIH, and they are commonly chronic, with or without an acute hemorrhagic component. A frequent but under-reported complication of CSF leak closure is rebound intracranial hypertension.

Discussion- The original conception of SIH was as a condition caused by low CSF pressure. Early reports of the condition considered low pressure to be the defining pathophysiological disturbance; Subsequent work, however, has clearly shown this conception to be incomplete. Currently, there is no single diagnostic test that excludes SIH with a high level of sensitivity.

Conclusion - In clinical practice SIH is generally underdiagnosed; patients may have headaches for decades before the diagnosis is considered. Although the most common presenting symptom is orthostatic headache, SIH should be suspected in patients with headaches that are daily from onset (NDPH) and refractory to every medical treatment.