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Review Article

Intracranial hypotension with spinal pathology

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Abstract

BACKGROUND CONTEXT: Intracranial hypotension (IH) is a syndrome in which volume depletion of the cerebrospinal fluid (CSF) results in various neurological symptoms. Most commonly, a small tear or defect in the spinal dural sac is the underlying lesion that results in a CSF leakage and IH. Causes of IH can be classified as 1) spontaneous (primary), and 2) secondary. Knowledge of IH is essential to spine surgeons, because a small proportion of patients with spontaneous IH require spine surgery, and secondary causes of IH include spine trauma, various spine-related diagnostic/therapeutic procedures (iatrogenic), and degenerative spine disorders.

PURPOSE: The purpose of this review is to update knowledge of IH pertinent to spine surgery.

STUDY DESIGN/SETTING: Review of the literature.

METHODS: A systematic review of the literature was conducted using PUBMED. The literature regarding IH with spinal pathology published from 1966 to 2005 was searched and reviewed comprehensively.

RESULTS/CONCLUSIONS: The lower cervical and upper thoracic spine are the predilection sites for a CSF leak in patients with spontaneous IH. Mechanical stress may have a role in the pathogenesis of spontaneous IH, and meningeal diverticula and connective tissue disorders may be important risk factors. Iatrogenic causes of IH include lumbar puncture, spine surgery, and chiropractic manipulation. Rarely, degenerative spine disorders are the secondary cause of IH. Although orthostatic headache is the characteristic symptom of IH, a small proportion of patients present with neck pain or radicular symptoms of the upper extremity. Imaging studies of the spine have contributed significantly to elucidation of the pathophysiology of IH as well as identification of the site of a CSF leak. Typical spinal radiographic findings of IH include extra-arachnoid or extradural fluid collections, meningeal enhancement, engorgement of the epidural venous plexus, and tonsilar descent into the foramen magnum. Most patients with spontaneous IH respond favorably to conservative management, including autologous epidural blood patch. Surgical intervention is reserved for those who fail the conservative management. Although immediate clinical improvement is usually achieved by surgery, the long-term outcomes of surgical patients need to be investigated further. In patients who have a secondary cause of IH, treatment of the underlying lesion or condition may terminate a CSF leak and result in reversal of symptoms.

Keywords: Intracranial hypotension; Spine; Spontaneous; Pathology; Diagnosis; Iatrogenic; Treatment

Introduction

Intracranial hypotension (IH) is a clinical syndrome in which absolute or relative hypovolemia of the cerebrospinal fluid (CSF) results in various neurological symptoms [1]. An increasing number of publications on the subject in recent years suggests that IH is no longer a rare syndrome. Although causes of IH are diverse, they can be classified as 1) spontaneous (primary), and 2) secondary (Table 1). The latter can be divided further into: 2-i) traumatic, mostly iatrogenic; 2-ii) degenerative spine disorders; 2-iii) truly hypovolemic, mostly from systemic dehydration; and 2-iv) overdrainage of the CSF by neurosurgical procedures. The last two entities, true hypovolemia and CSF overdrainage, are not included in this review, because of their irrelevancy to spine surgery.

It is rare for patients with IH to be seen by spine surgeons primarily: those patients almost invariably present with complaints of orthostatic headache, and neurologists

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Table 1
Causes of intracranial hypotension (IH)

1. Primary (Spontaneous) IH
i. Unknown cause
ii. Weakness of the dural sac, often associated with prior trivial trauma
Meningeal diverticula
Abnormalities of connective tissue
2. Secondary IH
i. Traumatic/Iatrogenic
Definite trauma which creates a spinal dural tear
Lumbar puncture
Spine surgery
Chiropractic manipulation
Other cranial surgery
ii. Degenerative spinal disorders
iii. Truly hypovolemic state (reduced body water)
iv. Overdrainage of the cerebrospinal fluid by neurological surgery (shunt/endoscopic)

Modified from: Mokri B. Low cerebrospinal fluid pressure syndromes. Neurol Clin 2004;22:55–74. Copyright permission from Elsevier Science.

and anesthesiologists are most frequently in charge of treatment. Knowledge of IH is essential to spine surgeons, however, because a small proportion of patients with spontaneous IH require spine surgery, and complications of spine surgery as well as degenerative spine disorders can be secondary causes of IH. We attempted to update knowledge of IH pertinent to spine surgery by a review of the literature.

Methods

A review of the literature was conducted using PUBMED, an Internet medical literature search engine. Key words for the literature search included intracranial hypotension, spine, spontaneous, pathology, diagnosis, iatrogenic, and treatment. The literature published from 1966 to 2005 was searched and reviewed comprehensively.

Etiology

Spontaneous

Many patients with IH present without known predisposing factors or history of a major trauma, and the condition has been called ‘‘spontaneous’’ IH [1–3]. Epidemiologically, there have been few published data on the prevalence or annual incidence of spontaneous IH, because of lack of population-based studies. In Olmsted County, Minnesota, the prevalence of spontaneous IH in 1995 was approximately one in 50,000 persons [4]. There was a clear female preponderance for the syndrome: a male to female ratio was between 1:1.75 and 1:4 [4–7]. The mean age at the time of presentation was 38 to 42 years in both genders [4–7]. It is unknown whether the prevalence or incidence of spontaneous IH is affected by ethnicity.

Most patients with spontaneous IH have a CSF leak in the spinal dural sac. Although the spinal level of a CSF leak can usually be determined with diagnostic imaging studies, the actual site of a leak cannot always be identified. In a study by Sencakova et al., the spinal level of a CSF leak was determined in 16 (64%), and the actual site could be identified in 9 (36%) of 25 patients with spontaneous IH [5]. The leak was present in the cervical spine in 6 (24%), thoracic spine in 9 (36%), and lumbar spine in 1 (4%) patient [5]. The results were similar to those reported by Schievink et al. [8]. In their study, a CSF leak in the spinal dural sac was identified in all of 25 patients, with some patients having multiple leaks [8]. The majority of those patients had a leak either in the cervical (14 patients, 56%) or thoracic spine (13 patients, 52%). A leak in the lumbar spine was rare, with only two patients (8%) [8]. A dural sleeve surrounding a spinal nerve root is the predilection site of a CSF leak, and meningeal diverticula, Tarlov’s cysts, or ‘‘nude’’ nerve roots are occasionally found [1,3,9–11]. Because most patients with spontaneous IH are treated nonsurgically, macroscopic or microscopic inspection of a lesion in the spinal dura has rarely been conducted [1,3,9–11].

Despite the term ‘‘spontaneous’’ IH, many authors think that mechanical stress has an important role in its pathogenesis, by acting on the point of focal weakness in the dural sac [1–3]. It is not uncommon for patients with spontaneous IH to recall having sustained a trivial trauma. In a series by Chung et al., 7 (23%) of 30 patients with spontaneous IH recalled a prior history of a trivial neck or head trauma after a detailed interview [12].

Connective tissue disorders may be important risk factors for spontaneous IH. Nine (16%) of 58 patients in a study by Mokri et al. [13] and 7 (38%) of 18 patients in a study by Schievink et al. [14] were found to have connective tissue disorders. In the latter study, three patients had a minor skeletal feature of Marfan syndrome, two had Ehlers-Danlos syndrome type II, and the other two had joint hypermobility [14]. No radiographic evidence of cardiovascular anomalies was found in those seven patients. The spinal level of a CSF leak, a male:female ratio, and the mean age at presentation in IH patients with connective tissue disorders seem to be no different from those without the disorders. Imaging studies often reveal multiple meningeal diverticula, indicating increased susceptibility of patients with connective tissue disorders to spontaneous IH [13,14]. The actual incidence of spontaneous IH in patients with full-blown Marfan or Ehlers-Danlos syndrome, however, has rarely been reported in the literature.

Traumatic/Iatrogenic

IH has been reported to occur after a major spine trauma, including a penetrating spine injury [15] and a thoracic spine fracture [16]. Most cases of traumatic IH are