Case Report

Frontotemporal brain sagging syndrome: Craniospinal hypovolemia secondary to a T6-T7 cerebrospinal fluid-venous fistula

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INTRODUCTION

Spontaneous intracranial hypotension (SIH) is characterized by orthostatic headaches that result from low cerebrospinal fluid (CSF) pressure, accompanied by neck stiffness and hearing deficits. In 2011, Wicklund et al.¹ coined the term frontotemporal brain sagging syndrome (FTBSS) for...
those patients with SIH that was indistinguishable from a behavioral frontotemporal dementia (FTD). Brain magnetic resonance imaging (MRI) may confirm the diagnosis of FTBSS by showing symmetrical pachymeningeal enhancement, brain sagging, cerebellar tonsillar ectopia, intracranial venous sinuses dilation, and subdural collections, attributed to a spinal CSF leak or hyperabsorption.\textsuperscript{8,16} Here, we present a case of FTBSS secondary to a T6-T7 CSF-venous thoracic fistula.

CASE REPORT

A 56-year-old male presented in 2002 with severe orthostatic bifrontal headaches and fatigue after performing scuba diving. From 2002 to 2005, his behavior was inappropriate and he was diagnosed with major depression. For and by 2009, he continued with orthostatic headaches plus lumbar pain, vertigo, tinnitus, nausea, and once vomiting. By 2015, he complained of memory dysfunction, apathy, neck pain, severe fatigue, and orthostatic headaches; he was admitted to the hospital with a suspected viral meningencephalitis and was treated with antivirals, antibiotics, and levetiracetam with minimal improvement. Later in that year, he was given a short cycle of prednisolone and showed partial improvement. From 2016 to 2018, his symptoms exacerbated, and he developed hypersomnolence, sleep apnea, memory impairment, inappropriate behavior, bradypsychia, bradydalia, hand and oral automatisms, urinary urgency, neck pain, ataxia, tremor, dysarthric speech, and dysphagia. We admitted him in 2018 after worsening of the state of consciousness, memory impairment, and inappropriate behavior.

His physical examination was normal, but the neurological examination revealed fluctuating consciousness (normal to clouded), inattention, disorientation, disinhibited behavior, impaired judgment, bradydalia, and memory impairment. On fundoscopy, there was blurring of the medial optic margins, elevation of the optic disc, and loss of venous pulsations (II). Bilateral absent gag reflex, flattening of the palatal arch (IX, X), dysphonia (X), hypotrophy, and fasciculations of the tongue (XII). Motor strength was 4/5 bilaterally with generalized hyperreflexia and both rest and intention tremor.

Neuropsychological test battery (e.g., MoCA, WAIS, CPT, Tower of London test, TAVEC, WCST, and Rey complex figure test) was all abnormal, which showed mild deficits in attention and planning, plus severe deficits in working, explicit, and episodic memory. A lumbar puncture revealed an opening pressure of 0 cm H₂O, but normal CSF laboratory findings. The electroencephalography showed dominant background alpha rhythm, left occipital amplitude asymmetry without epileptic activity. The brain MRI showed severe brain sagging, minimal cerebellar tonsillar descent, bilateral temporal lobe herniation, and pachymeningeal enhancement [Figure 1].\textsuperscript{14} The thoracic MRI showed multiple diverticula. A computed tomography (CT) myelogram performed in prone, supine, and lateral positions revealed a right T6 thoracic meningeal diverticulum associated to a CSF-venous leak and a T7 small meningeal diverticulum [Figure 2].

Surgical technique

A two-level T6-T7 laminotomy was performed to address the CSF-venous leak (e.g., using fluoroscopic and neuronavigation). The T6 and T7 nerve roots were exposed, the diverticulum was isolated/dissected and ligated from the surrounding veins (e.g., ligated with vascular Sugita straight clips placed at the origin of the root sleeves) [Figure 2].

One year later, cognitive, behavioral, and cranial nerve symptoms progressively improved; now 2 years postoperatively, he remains asymptomatic. Neuropsychological tests, 6 months and 1 year after surgery, revealed significant improvement. Sleep apnea improved, but he still requires continuous positive airway pressure. The postoperative MRI’s (48 h, 1 month, and 12 months after surgery) showed progressive improvement of the sagging index/uncal herniation [Figure 3].\textsuperscript{11}

DISCUSSION

In 2002, Hong et al.\textsuperscript{4} published the first case of SIH associated to memory loss, personality, and behavioral changes. In 2011, Wicklund et al.\textsuperscript{22} coined the term FTBSS, characterized by behavioral disinhibition, apathy/inertia, loss of empathy, perseverative behavior, hyperorality, memory impairment, auditory and/or visual hallucinations, hypersomnolence, orthostatic headache, neck pain, tinnitus, dysarthria, dysphagia, tremors, orofacial dyskinesias, nausea, and unsteady lurching gait.\textsuperscript{15}

Although SIH, also called craniospinal hypovolemia, is caused by leakage of CSF volume from the spine, CSF pressure is normal in the majority of patients and it only decreases when the rate of CSF volume loss cannot be furthered compensated.\textsuperscript{6,10,14} The most important intracranial complications are in the short-term subdural hematomas or hygromas and in the long-term FTBSS\textsuperscript{15,17} which leads to a mechanical disruption of the frontotemporal, deep midline, and brainstem tracts.\textsuperscript{22} Orthostatic headaches are secondary to a CSF depletion, leading to undue traction to vascular structures\textsuperscript{11} and brain displacement toward the skull base meninges. Headache is induced when 10% of the volume is drained in the upright position, and by over 40% decrease of the negative vertex CSF pressure.\textsuperscript{12} This results in a compensatory increase of the intracranial blood volume, explaining the characteristic pachymeningeal gadolinium enhancement, spinal venous engorgement, and subdural collections.\textsuperscript{11}

Etiology of FTBSS

The etiology of FTBSS is usually attributed to a spontaneous spinal CSF leak, however in the largest clinical series, 12/29
In our case, a CSF-venous leak was meticulously identified. They seem to occur in the thoracic spine and the thoracolumbar junction, are more frequently paravertebral, and most of them are associated with a nerve root sleeve diverticulum. Kranz et al.\(^5\) suggested that a rupture of an enlarged arachnoid granulation into its adjacent vein is a possible explanation of these rare leaks.

**Treatment options**

Treatment options for FTBSS depend on the CSF leak type \([\text{Table 1}].\)\(^1,1,3,8,10,13,15,16,18,21,23\) CSF-venous fistulas (Type 3 leaks) do not respond in the long-term to targeted blood patches and the main options include sacrificing a nonappendicular nerve root and/or direct venous fistula obliteration.\(^10\) In our case, we

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Figure 1: Preoperative brain magnetic resonance imaging showed severe brain sagging signs, in the axial plane (a) a distorted midbrain anatomy was observed at the level of the tentorial incisura in which an anteroposterior midbrain elongation (35 mm) and temporal lobe herniation were evident. Notice the anterior and posterior parahippocampal herniation (b). In the sagittal plane, there was sagging of the brainstem, cerebellar tonsillar descent, shortened pontomammillary distance (2.8 mm/abnormal <5.5 mm) (c), flattening of the pontomesencephalic angle (10.5°/abnormal <50°) (d), narrow vein of Galen/straight sinus angle (50.7°), aqueduct displacement (e), and cerebellar tonsillar descent using the McRae line (2.4 mm/abnormal >5 mm) (f). Notice the generalized pachymeningeal enhancement (g). In the coronal plane, the third ventricle was thin and elongated, the mammillothalamic tract was displaced downwardly, and bilateral uncal herniation was observed (h and i).\(^2\)
Figure 2: Hyperdense paraspinal vein sign[6,18] was observed on computed tomography (CT) myelogram. A curvilinear high attenuation vascular structure was observed in connection with a T7 nerve root diverticulum (a and b). The contrast filling paraspinal vein and CSF leak was better observed on CT myelography in the prone and right lateral decubitus position. Notice the paraspinal vein (c) and the 3D reconstruction of the thoracic diverticula (d). CSF-venous leak was localized, and the thoracic roots were exposed (e). Multiples veins were coagulated (f) during the root dissection; vascular Sugita straight clips were placed at the origin of the root sleeves (g).

Figure 3: MRI showed axial, coronal, and sagittal planes (a-l). Notice the progressive hypotension resolution over the time. Anterior-posterior midbrain diameter was 35 mm preoperative versus 25 mm postoperative (1 year). Pontomammillary distance was 2.8 mm preoperative versus 8.1 mm postoperative (1 year). Pontomesencephalic angle was 10.5° preoperative versus 51° postoperative (1 year). Vein of Galen/straight sinus angle was 50.7° preoperative versus 68.1° postoperative. Sagging index (anterior-posterior midbrain diameter/pontomammillary distance) improved significantly 1 year after surgery (12.5 vs. 3.08). A sagging index higher than 10 suggests an atypical clinical presentation for craniospinal hypovolemia.[1]
decided to clipped and ligated a T6 CSF-venous fistula and a T7 diverticulum, following which symptoms and brain sagging improved [Figure 3]. FTBSS is a rare potentially reversible syndrome that has to be approached using spine MRI and CT myelogram, to determine the etiology of the CSF leak.

**CONCLUSION**

We should consider FTBSS in the differential diagnosis of an early onset FTD. Unfortunately, the CSF leak is rarely detected early in the clinical course. In this case, MR/myelo-CT was essential to identify the T6-T7 CSF-venous fistula that was appropriately surgically dissected/ligated and should be similarly used in other comparable cases.

**Declaration of patient consent**

Patient's consent not required as patients identity is not disclosed or compromised.

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**Conflicts of interest**

There are no conflicts of interest.

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