Cessation of spontaneous intracranial hypotensive symptoms following fibrin epidural patching in a patient with connective tissue disorder and chronic, recalcitrant spontaneous CSF leak

Stacy de la Motte, MD, Genevieve D’souza, MD, and Elliot J. Krane, MD

**Background**

Intracranial hypotension (IH) commonly results from a spontaneous CSF leak. IH is characterized by orthostatic headache, cervical pain, dizziness, tinnitus, autonomic changes, which improve in the supine position, and is often encountered in patients with connective tissue disorders.

Certain connective tissue disorders are known to be associated with meningeal abnormalities that potentially may lead to dural defects. Meningeal diverticula occur in Marfan syndrome, Ehler’s-Danlos syndrome type 2, neurofibromatosis, autosomal dominant polycystic kidney disease, and familial osteosclerosis.

**Case Report**

We present the case of a 13-year-old Caucasian male with an unspecified connective tissue disorder with Marfanoid features, who met the diagnostic pattern of CSF hypotension with chronic headache and neck pain.

Symptoms began in 2011, localized at the junction of the bottom of the skull and neck, in the midline and bilaterally. Pain was described as 5/10 on VAS scale, and positional. Episodes of pain were described as non-radiating, constant, non-pulsatile, starting in the morning with rising, and lasting for 2 days.

MRI demonstrated IH with flattening of the pons and accelerated CSF flow velocity in the cervicothoracic region, but was unable to identify the exact location of a dural tear.

**Figures 1 & 2:** The patient’s MRI demonstrating flattening of the pons and accelerated CSF flow velocity in the cervicothoracic region.

**Discussion**

Successful treatment of intracranial hypotensive symptoms with epidural fibrin sealant has been reported in post-dural puncture headache in adults secondary to long-term intrathecal catheterization.

Additionally, fibrin sealant is widely used to achieve watertight dural closure in neurosurgical and orthopedic operations.

This novel case illustrates that epidural patching with fibrin sealant is an alternative to autologous epidural blood patching, when epidural blood patching does not result in resolution of the symptoms in a pediatric patient in the setting of an underlying connective tissue disorder.

**References**
