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Clinical Note

Trigeminal Neuralgia in a Patient With Spontaneous Intracranial Hypotension

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Spontaneous intracranial hypotension and trigeminal neuralgia are examples of pain syndromes arising from shifting anatomical relationships in the posterior fossa. We report both conditions occurring in the same patient and resolving following surgical closure of a cervical nerve root sleeve dural defect. This case further elucidates the pathophysiologic basis of both forms of head pain.

Key words: trigeminal neuralgia, cerebrospinal fluid hypovolemia, subdural cerebrospinal fluid effusion, intracranial hypotension

Abbreviations: CSF cerebrospinal fluid, CT computed tomography, MRI magnetic resonance imaging, SIH spontaneous intracranial hypotension, TN trigeminal neuralgia

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Spontaneous intracranial hypotension (SIH) is an under-recognized cause of headache caused by cerebrospinal fluid (CSF) leakage resulting in CSF hypovolemia.[1-3] Patients typically describe a bilateral postural headache that is minimal or absent in the recumbent posture, but which greatly worsens in the upright posture. Associated symptoms may include nausea, vomiting, vertigo, or tinnitus.[1-3]

Cranial nerve palsies are known to occur in SIH. The abducens nerve is most commonly involved,[4-7] and the oculomotor,[5] vestibular, or trochlear[8] nerves[9] less frequently. We report what we believe to be the first case of trigeminal neuralgia (TN) in a patient with SIH.

Trigeminal neuralgia consists of abrupt, recurrent, paroxysmal pain in the distribution of one or more branches of the trigeminal nerve. Unlike SIH, the pain of TN is nonpostural. The typical pain of TN is sharp or electrical in quality and almost always unilateral. Many patients will describe a facial trigger zone separate from the site of pain.[10] SIH and TN each have an incidence of approximately 5 per 100,000 per year.[3,10] Onset of SIH is most commonly during the fourth or fifth decade,[3] and TN beyond the sixth decade,[10] although either can occur at any age.

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CASE HISTORY

A 56-year-old man developed persistent postural headaches following vigorous coughing during an upper respiratory tract infection. He described the headache as a daily, intense, dull nuchal ache radiating as a pressure sensation to the bifrontal head regions. The headache consistently worsened during standing, bending forward and physical exertion, and greatly improved or resolved within several minutes

Conflict of Interest: None

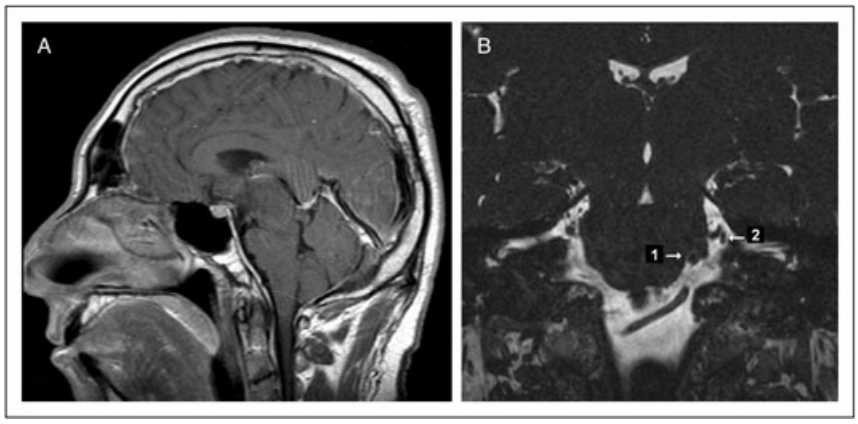

Figure.—(A) Sagittal T1-weighted MRI showing diffuse pachymeningeal gadolinium enhancement and downward displacement of the cerebellar tonsils preoperatively. Note also the anterior pontine vascular indentation. (B) Coronal high-resolution constructive interference in steady state (CISS) MRI through the posterior fossa showing a dolichoectatic vertebrobasilar artery system with a tortuous intracranial distal left vertebral arterial loop (1) effacing the inferiolateral pons caudal to the trigeminal nerve root entry zone. The transverse pontine vein approximates the left trigeminal nerve root (2).

of recumbency. Neurological examination was unremarkable.

Normal investigations included complete blood count, sedimentation rate, paranasal sinus computed tomography, cranial magnetic resonance angiography, and venography. CSF was acellular with a protein concentration of 79 mg/dL. Opening pressure measured reclining was 7-cm H_2O. Bacterial, tuberculine, and fungal cultures were negative. Magnetic resonance imaging (MRI) with gadolinium demonstrated diffuse pachymeningeal enhancement and cerebellar tonsilar ptosis (Figure A). Panmyelography disclosed elongation and irregularity of the left C8 nerve root sleeve with subtle adjacent extraforaminal contrast collection. Radionuclide cisternography showed a corresponding triangular focus of increased radiotracer activity extending lateral to the left C8 nerve root sleeve. These findings were consistent with a CSF leak. A series of 3 cervical epidural blood patches relieved the headache immediately, but 2 weeks following each procedure, the symptoms returned.

The postural headache had been present for 9 months when a new type of pain emerged. Abrupt paroxysms of brief (1-20 seconds), sharp, electric shock-like pain involved the left upper lip and gum. This TN pain was excruciating, overshadowing the intensity of the preexisting postural headache, and striking up to 15 times a day. The orofacial pain was nonpostural, being triggered not by the upright posture but instead by any slight touch of the left nasolabial fold, eating, speaking, or brushing the teeth. Dental examinations were normal. Gabapentin and baclofen were ineffective. Carbamazepine at 200 mg daily completely abolished the facial pain by the second day of treatment. Unfortunately, the facial pain returned several weeks later and was inadequately controlled by carbamazepine up to 800 mg daily. Side effects of gait ataxia and dysarthria prevented further dosage increases.

The patient underwent surgical repair of the C8 nerve root sleeve leak. Intraoperatively, a small dural outpouching was visualized laterally and inferiorly at the level of C8. The leak was sealed with an epidural silicon impression mold (Dur-A-Sil®, Insta-Mold Products, Oaks, PA) and Gelfoam® gelatin sponge. Postoperatively, both the SIH and TN pains resolved entirely. MRI 1 month later demonstrated resolution of meningeal enhancement and tonsillar ptosis. High resolution images disclosed a tortuous intracranial distal left vertebral arterial loop effacing the inferiolateral pons caudal to the trigeminal nerve root entry zone (Figure A,B). Additionally, the transverse pontine vein approximated the trigeminal nerve root (Figure B). Complete pain relief was sustained during 2 years of follow-up.

COMMENTS

Our patient had the misfortune of developing 2 distinct forms of cephalalgia, each of which was debilitating. Epidemiological, temporal, and structural observations support the conclusion that SIH contributed to the origin of his TN. A coincidental association is unlikely since both SIH and TN are uncommon disorders. The fact that immediate resolution of TN coincided with surgical closure of the spinal dural defect indicates that CSF hypotension caused or greatly contributed to this patient's TN. Accordingly, the neuroimaging abnormalities suggest several potential structural interrelationships.

The prevailing theory of SIH attributes the CSF leaks to fragile spinal meningeal diverticulae at radicular nerve root sleeves.[1-3,11] Depletion of CSF volume decreases brain buoyancy, resulting in downward displacement of the brain and traction of pain-sensitive vascular structures. Some patients will recall a recent history of minor trauma, but in most cases the CSF leaks are presumed to be spontaneous. Diagnosis is confirmed by gadolinium-enhanced MRI, which typically shows diffuse pachymeningeal enhancement due to engorgement of venous sinuses or the epidural venous plexus. Additional findings may include subdural fluid collections, pituitary hyperemia, and sagging of the brain with downward displacement of the cerebellar tonsils.[11] Radionuclide cisternography or myelography may disclose single or multiple CSF leaks, most frequently at the cervicothoracic region as in our patient.[11] CSF lymphocytic pleiocytosis and elevated protein content are common findings which may be due to increased permeability of dilated meningeal blood vessels or decreased lumbar CSF flow.[1-3] SIH frequently resolves spontaneously or with conservative treatment including bed rest and hydration.[1] Effective treatments for persistent cases include lumbar puncture with epidural autologous blood patch,[1,2,11] CT-guided fibrin glue injection at the site of leakage or, in severe intractable cases such as this one, open surgical closure of the dural defects.[12]

As SIH is known to impair the third,[4] fourth,[8] sixth,[4-7] and eighth[9] cranial nerves, it seems plausible that the fifth cranial nerve could be similarly vulnerable. The basis of cranial neuropathies in SIH is incompletely understood. One possible cause may be epidural venous engorgement with entrapment of individual cranial nerve roots, in this case, the trigeminal ganglion where it penetrates the dural terminus in Meckel's cave. Another may be altered vasa nervorum dynamics from traction on cranial nerves by the downward shift of posterior fossa contents. A further possibility may be a derangement of collagen fibers affecting both spinal dural nerve root sleeve and cranial nerve sheath structural integrity.

We believe that the probable mechanism of TN in this case was decompensated vascular compression. MRI in our patient disclosed 2 relevant findings. First was a tortuous left vertebral artery indenting the inferiolateral pons near the trigeminal nerve root entry zone (Figure A,B). Second was vascular contact with the left trigeminal nerve root (Figure B). We conclude that downward displacement of the brain stem from intracranial hypotension shifted the anatomical proximity of these blood vessels to the trigeminal nerve root or its entry zone, accentuating previously asymptomatic neurovascular contact sufficiently to provoke symptomatic TN.

The pathophysiology of TN is frequently related to neurovascular compression. Ultrastructural studies, neuroimaging and intraoperative findings, combined with relief of pain following surgical decompression support the widely held theory that idiopathic TN arises from pulsatile vascular indentation leading to focal demyelination and sensitization of the trigeminal nerve root.[10] Venous compression, most frequently by the transverse pontine vein, as in this case, causes approximately 6% of TN.[13] Vertebrobasilar dolichoectasia is also a recognized, albeit uncommon, cause of TN.[14,15]

In conclusion, the downward anatomical shift of the posterior fossa in this case of SIH caused not only a postural headache but also TN. The mechanism appeared to be decompensation of previously asymptomatic neurovascular compression. Surgical closure of the CSF leak completely relieved both types of pain.

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