Intracranial Hypotension With Parkinsonism, Ataxia, and Bulbar Weakness

Anthony S.-I. Pakiam, MD; Christine Lee, MD; Anthony E. Lang, MD

Objective: To describe a case of spontaneous intracranial hypotension with a previously unreported constellation of presenting features.

Design: Case report.

Setting: Tertiary care center.

Main Outcome and Results: We describe a patient with intracranial hypotension who presented with a parkinsonian syndrome and later development of ataxia and prominent bulbar symptomatology. Headache was not a feature of her initial presentation and was only reported after repeated questioning during later evaluations. Magnetic resonance imaging of the patient’s head revealed findings characteristic of intracranial hypotension. An \([^{18}\text{F}]\text{fluoro-m-tyrosine positron emission tomographic scan showed normal striatal activity, suggesting intact presynaptic nigrostriatal function. Opening pressure on lumbar puncture was reduced at 40 mm H}_2\text{O. A source of cerebrospinal fluid leakage was not identified on nuclear cisternography and the patient underwent lumbar epidural blood patching, which resulted in complete resolution of her signs and symptoms as well as in a marked improvement in her imaging findings.}

Conclusions: The clinical spectrum of intracranial hypotension can be broadened to include parkinsonism, cerebellar ataxia, and prominent bulbar dysfunction. As with more common manifestations of the disorder, these features may resolve after appropriate treatment.

Arch Neurol. 1999;56:869-872

Intracranial hypotension\(^1-5\) characteristically presents with a postural headache and low cerebrospinal fluid (CSF) pressure. Common accompanying features include nausea, vomiting, stiff neck, photophobia, and vertiginous symptoms. The syndrome may occur spontaneously, as first described by Schaltenbrand,\(^6\) follow rupture of an arachnoid (Tarlov) cyst, or have a precipitant, such as lumbar puncture, trauma, or cranial or spinal surgery.\(^1,3\) We describe a patient with an unusual constellation of presenting features in whom headache was not initially a prominent complaint.

REPORT OF A CASE

A 54-year-old right-handed woman was seen in our clinic regarding the possibility of Parkinson disease. One year earlier, she had experienced the onset of a right hand tremor that had mainly been present at rest but had spontaneously improved before her clinic visit. Also, her speech had become softer during the past year, and certain activities, such as eating or walking, were slower than before. She had noted no change in speed or ability to perform other activities, including the use of utensils, personal hygiene, and getting dressed or undressed. Writing had become extremely “sloppy” but not micrographic. She had experienced 3 falls, 1 as a result of tripping over an object and 2 from losing her balance while walking downhill. She had been treated for depressive symptoms with paroxetine (10 mg/d) for 8 months, with some improvement in her mood. The paroxetine therapy had been discontinued 3 months earlier. She was hospitalized 2 months after her initial clinic visit and had in the interim developed more of a nasal quality to her voice; dysphagia, which was more prominent with liquids than solids; and vomiting 2 or 3 times per week. She also admitted to a mild headache, which was holoccephalic, steady, and pressurelike in quality and occurred only when she coughed. Standing up did not precipitate her headaches, but lying down could make them less intense. She also reported epi-
sodic fatigue and having been “uptight” and making mistakes at work 2 to 3 years earlier. Also, in retrospect, she checked some notes that she had taken 3 1/2 years earlier and found that she had experienced a sensation of a “plugged” left ear 1 day after doing a handstand in a swimming pool and had headaches 4 days later. The following year, in addition to having difficulty at work, she had again noted a sensation of plugged ears. She had no visual, vertiginous, or autonomic symptoms; she had no history of head or neck trauma; and she had never had a lumbar puncture. Her only medication was alprazolam (0.25 mg nightly). There was a history of a tremor in her father, a paternal aunt, and 2 cousins. Her medical history was remarkable for breast cancer 5 years earlier, for which she had undergone a left mastectomy and had subsequently been treated with tamoxifen citrate, with no evidence of recurrence.

General examination revealed a tall, thin woman. Neurological examination revealed normal alertness and cognition. Her speech had a nasal quality and was hypophonic. Her palatal elevation was reduced and her gag reflex was weak. The findings of the remainder of her cranial nerve examination were normal. Myerson sign was absent. Muscle strength and tone were normal, with no cogwheeling. Only a very mild, intermittent high-frequency tremor was present in the extremities in all positions. Fast, repetitive movements demonstrated mild but definite bradykinesia, more so on the left than on the right. No dysmetria was present, and sensation was intact to all modalities. Her deep tendon reflexes were normal except for reduced ankle reflexes, and her plantar responses were flexor. Her gait was unsteady, and she was unable to tandem walk, although she could ambulate without support. Her postural reflexes were impaired, with several steps of retropulsion but no falling.

Magnetic resonance imaging (MRI) of the brain (Figure 1) revealed a downward displacement of posterior fossa structures and an elongation of the brainstem in the anteroposterior plane. Dural enhancement was present mainly in the posterior fossa. The findings of [18F]fluoro-m-tyrosine positron emission tomographic scanning were normal. Opening pressure on lumbar puncture was low at 40 mm H2O, and the results of CSF studies, including cytology, were otherwise normal. Cisternography using technetium pentetic acid failed to show a CSF leak after lumbar instillation. The patient underwent epidural blood patching with 20 mL of blood in the midlumbar area, after which she remained supine for 30 minutes but was not placed in the head-down position. After the blood patching, her voice and swallowing returned to normal within 2 days. Her headaches resolved rapidly. The neck stiffness, which had developed after the lumbar puncture, also resolved after the blood patching. Five weeks later, the findings of her neurological examination were normal. Her palatal elevation and gag reflex were normal, as were her walking ability and performance of rapid repetitive movements. Her writing skills also returned to normal. Follow-up MRI (Figure 2) after 7 weeks showed a normal position of the brainstem and cerebellar tonsils. There was mild persistence of the elongation of the midbrain in the anteroposterior direction.

Intracranial hypotension has been reported to occur not only after significant trauma, spinal or intracranial surgery, and lumbar puncture, but also after even minor trauma or strain. The syndrome may follow or occur at the time of sexual intercourse, paroxysmal coughing, bending down, or falling and landing on the buttocks, in association with back pain radiating into the lower extremities. In the latter case, CSF leakage is presumably precipitated by trauma causing a disruption in the nerve root sleeve at the lumbosacral level. Similarly, cases have been associated with repetitive strenuous upper extremity motions, such as racket sports or throwing movements when fishing, suggesting injury to nerve root sheaths at the level of nerve roots supplying the brachial plexus, through which stretch is transmitted. Our patient experienced minor strain the day before her first symptoms occurred, but there was no clear precipitating event, so the diagnosis in her case is best classified as spontaneous intracranial hypotension.

Our patient had an unusual presentation of spontaneous intracranial hypotension and was initially re-
ferred with a query of Parkinson disease because of a unilateral resting hand tremor, bradykinesia, and gait imbalance. Another atypical feature was the subsequent development of bulbar weakness manifesting as nasal-ity of speech and dysphagia. Cranial nerve abnormalities reported in other cases of intracranial hypotension have included symptoms referable to the optic nerve or chiasm manifesting as blurred vision,3,10 transient visual obscurations, and visual field deficits.8,11 Abducens palsy11 with resultant horizontal diplopia may be present, most likely as the result of distortion of the nerve from traction or pressure. Symptoms and findings referable to cranial nerve VIII,3,8,12 including sensorineural hearing loss, hyperacusis, tinnitus, vertigo, and a sensation of “fullness”,3 “popping,”9 or “echoing”4 in the ear, have also been reported. Some of these latter symptoms may be related to secondary alterations in intralabyrinthine pressure dynamics, which could also account for the sensation of “ear plugging” experienced by our patient. Finally, facial weakness5,13 or numbness5 and dysgeusia3 have been reported in association with intracranial hypotension. Cranial nerve palsies may be persistent for a prolonged period, despite immediate relief of postural headache after epidural blood patching.13

Potential pathophysiological mechanisms contributing to cranial nerve dysfunction in intracranial hypotension include intracranial venous distention, given the fixed volume of the intracranial space, and traction or pressure on affected cranial nerves, especially when downward displacement of the brainstem is evident, as was the case in our patient. The cardinal symptom of postural headache is also postulated to result from traction on pain-sensitive structures, including cranial nerves,3 rather than from the reduced intracranial pressure alone, since maneuvers to increase intracranial pressure, such as jugular venous compression or valsalva, increase rather than relieve the headache.5 The degree of brain descent and resultant contributory traction mechanism to the characteristic postural dependence of headache and other symptoms may be somewhat underestimated by MRI, since these images are by necessity obtained with the patient in the recumbent position. In addition to a traction mechanism affecting cranial nerves and pain-sensitive structures in the posterior fossa, a compressive effect on the brainstem can be postulated in light of the midbrain deformity present in our patient, in whom one manifestation was a partial parkinsonian syndrome. However, an [18F]fluoro-m-tyrosine positron emission tomographic scan14 did not provide evidence of the speculated presynaptic dopaminergic dysfunction of nigral origin. Depressed level of consciousness, described in a recent report of intracranial hypotension due to a torn dural root sleeve and ruptured Tarlov cyst, was also blamed on central herniation and compression of diencephalic structures.15

Abnormal findings on brain MRI in our patient were characteristic of intracranial hypotension and consisted of low-lying cerebellar tonsils; displacement of the brainstem, with effacement of the interpeduncular and prepon-tine cisterns; and thickening of the meninges with linear cisternal space surrounding the midbrain and mild persistent deformity. Subdural hematomas5 or subdural effusions and dilatation of cortical and spinal epidural veins resulting from the altered pressure dynamics may also be seen,3,12 and dural enhancement is characteristically diffuse.16-19 Cerebrospinal fluid pressure is typically,2 but not always,20 reduced to less than 60 mm H2O, and CSF analysis may disclose a variable presence of erythrocytosis, lymphocytic pleocytosis, and an elevated protein content.3,21 The site of CSF leakage was sought in our patient by nuclear cisternography, the findings of which were unrevealing. Alternative studies include spinal MRI and computed tomographic myelography, which is potentially more sensitive5 but was not performed in our patient, since the diagnostic lumbar puncture was not well tolerated. Recently, Pleasure et al15 emphasized the utility of thin-section MRI of the entire spine in refractory cases. Treatment may involve surgical ligation or epidural packing of leaking men-

Figure 2. Magnetic resonance imaging scan of the patient’s brain 5 weeks after treatment. A, Sagittal T1-weighted scan showing a return of the cerebellar tonsils to a normal position. B, Axial T2-weighted scan showing an increase in cisternal space surrounding the midbrain and mild persistent deformity.
ingeval diverticula if found. However, many investigators would first try a large-volume (20-30 mL) epidural blood patch in the lumbar region, followed by a 5-minute period with the patient’s head lowered 30°. In our patient, blood patching provided complete and rapid relief of symptoms along with radiographic improvement. When the initial blood patching fails to result in sustained benefit, subsequent investigations and/or another blood patching at the same or a higher level can be performed. Alternatively, a 24-hour epidural infusion of normal saline may be effective.

In conclusion, the clinical spectrum of intracranial hypotension can now be broadened to rarely include parkinsonism, cerebellar ataxia, and prominent bulbar symptomatology. As with other symptoms more commonly present in this condition, these features and accompanying radiological changes may resolve following appropriate treatment of the disorder.

Accepted for publication August 17, 1998.

The [18F]fluoro-m-tyrosine positron emission tomographic scanning was performed by Claude Nahmias, PhD.

Reprints: Anthony E. Lang, MD, Division of Neurology, Toronto Western Hospital, 399 Bathurst St, MP 11-306, Toronto, Ontario, Canada M5T 2S8.

REFERENCES