Hemifacial Spasm
An Unusual Manifestation of Idiopathic Intracranial Hypertension

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Increased intracranial pressure may produce a variety of clinical manifestations, some common and others rare. We present a patient with idiopathic intracranial hypertension whose initial symptom was hemifacial spasm. All signs and symptoms of intracranial hypertension resolved with acetazolamide.

Key Words: Hemifacial spasm—Idiopathic intracranial hypertension—Pseudotumor cerebri.

Idiopathic intracranial hypertension also known as pseudotumor cerebri, is a clinical syndrome of increased intracranial pressure in the absence of a mass or hydrocephalus and with normal spinal fluid constituents. Symptoms typically associated with elevated intracranial pressure include headache (94%), transient visual obscurations (68%), intracranial noises (58%), photopsias (54%), diplopia (38%), and visual blur or field loss (30%) (1). Round and Keane (2) profiled some of the less common clinical manifestations of elevated intracranial pressure among 101 patients with idiopathic intracranial hypertension. These included paresthesias (22%), arthralgias (13%), back or leg pain (5%), and ataxia (4%).

Involvement of cranial nerves other than optic and abducens nerves is another uncommon manifestation of intracranial hypertension. There are sporadic reports of oculomotor, trochlear, trigeminal, facial, and hypoglossal nerve palsies that resolved with lowering of the elevated intracranial pressure (3-9). In this paper, we describe a patient whose presenting symptom of idiopathic intracranial hypertension was transient facial nerve dysfunction. However, unlike previously reported cases, our patient had hemifacial spasm, a syndrome of facial nerve hyperexcitability, rather than facial nerve palsy.

CASE REPORT

A 46-year-old woman was referred for papilledema. Two months previously, she had developed intermittent twitching and "drawing-up" of the right side of her face with sustained closure of the right eye lasting 1-2 minutes. This occurred several times daily and was unassociated with other involuntary movements. Hemifacial spasm was observed and diagnosed by her local physician,
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who did not find any evidence of facial numbness, paresthesias, or palsies between episodes of hemifacial spasm. An electroencephalogram (EEG) and magnetic resonance imaging (MRI) of the head were normal. Specifically, no vertebrobasilar artery tortuosity or ectasia was noted. She was then started on carbamazepine with significant reduction in the frequency of her right hemifacial spasm. One month later, she noticed decreased visual field inferiorly in the left eye and complained of severe headaches and intermittent pulsatile tinnitus. She was referred for neuro-ophthalmologic evaluation. Her last episode of hemifacial spasm was 3 days prior to our evaluation.

Past medical history was unremarkable except for chronic obesity with a recent 10-lb weight gain. She had not ingested vitamin A or antibiotics. Current medications were carbamazepine and Premarin.

On examination, visual acuity was 20/20 OD and 20/25 OS. Pupils were 5 mm OU with a 1+ left afferent defect. Kinetic perimetry on a Goldmann perimeter demonstrated peripheral inferior constriction in the right eye and an inferior altitudinal defect in the left eye. Intraocular pressures were 18 mm Hg OU. Funduscopy revealed moderate bilateral disc edema with hyperemia. The remainder of the ophthalmologic and neurologic examination was normal. No facial muscle spasms were seen. There was no underlying facial weakness or synkinesis.

Serum chemistries and serologies were normal. Lumbar puncture showed an opening pressure of 350 mm H$_2$O. Spinal fluid analysis was normal. Acetazolamide 500 mg three times daily was begun, and the patient reported marked improvement of her headaches and resolution of tinnitus after 1 week. Carbamazepine was discontinued without recurrence of hemifacial spasm.

At 9 months follow-up, the patient remained asymptomatic. She had lost a total of 30 lb and was maintained on acetazolamide 500 mg twice daily. Her neuro-ophthalmologic examination was normal, and serial kinetic perimetry on a Goldmann perimeter disclosed no progression of the visual defects. Several months later, acetazolamide was decreased to 500 mg once daily, but recurrence of headaches and a “funny sensation” around her eyes prompted a return to twice daily dosage.

DISCUSSION

Idiopathic intracranial hypertension typically affects the optic nerves by transmission of increased cerebrospinal fluid pressure to the retrolaminar subarachnoid space. Axoplasmic stasis then leads to papilledema with or without associated visual loss. Involvement of other cranial nerves has been described in idiopathic intracranial hypertension, most commonly abducens palsy in 24% of cases (10). Various mechanisms for sixth nerve palsy as a remote effect of intracranial hypertension have been proposed. They include traction from brainstem shifts of the longitudinal segment of the sixth nerve, stretching of its transverse segment across the bony skull base, growing of the pontine surface, compression/constriction of the sixth nerves as they exit the brainstem by the anterior inferior cerebellar and internal auditory arteries and transient nuclear or neural ischemia due to poor blood-flow associated with spontaneous, periodic rises of intracranial pressure (Lundberg a waves) (11-13).

Because of the frequent association of abducens palsy and increased intracranial pressure with or without an intracranial mass, it has been called a “false localizing sign.”

The mechanism of other cranial nerve palsies due to idiopathic intracranial hypertension remains even more speculative. Nevertheless, a popular theory for any cranial nerve palsy associated with idiopathic intracranial hypertension invokes pressure-induced intracranial structural shifts which pull and stretch the extra-axial segment of the involved cranial nerve. At sites where cranial nerves traverse bone and arteries, it is hypothesized that progressive stretching of the nerve fibers might temporarily paralyze axonal function. Once intracranial pressure is lowered, the cranial nerves can “relax” and function is immediately restored.

For example, Kiwak and Levine (8) detailed the clinical course of a patient with idiopathic intracranial hypertension and bilateral facial palsies which remitted and recurred as intracranial pressure was lowered then rose again. Permanent recovery of the facial diplegia was secured after lumbarperitoneal shunt placement. These authors hypothesized a mechanism of stretch injury to the facial nerves within the facial canal as there was no concomitant eighth nerve dysfunction. The rapidity of recovery implied that significant demyelination or axonal disruption did not occur. However, we have seen a patient with idiopathic intracranial hypertension who had persistent facial diplegia with eventual facial nerve synkinesis from aberrant regeneration despite lowering of her intracranial pressure with medical and surgical modalities.

Our patient is noteworthy because her initial symptom of idiopathic intracranial hypertension was hemifacial spasm, not facial palsy. We believe her hemifacial spasm was a manifestation of intra-

cranial hypertension because of the close temporal relationship between its appearance and the onset of more typical symptoms of elevated intracranial pressure (headache, tinnitus, visual field loss). Furthermore, there was no reemergence of our patient's hemifacial spasm when carbamazepine was stopped and the other symptoms of idiopathic intracranial hypertension were being effectively controlled with acetazolamide.

A well-documented mechanism for some cases of hemifacial spasm is compression of the facial nerve by an adjacent blood vessel as the nerve emerges from the brainstem (14). The utility of noninvasive neuroimaging in identifying dolichoectasia of the vertebrobasilar artery as a cause of hemifacial spasm was recently investigated in 37 patients with hemifacial spasm by Bernardi and colleagues (15). Routine magnetic resonance imaging was positive for abnormal brainstem vasculature in 27% of patients, 51% with magnetic resonance angiography and 65% with magnetic resonance tomographic angiography. Our patient had routine magnetic resonance imaging that did not show evidence of vertebrobasilar artery tortuosity or ectasia. Why compressive injury sometimes acts to irritate a nerve and at other times acts to disable a nerve is not clear. We propose that elevated intracranial pressure shifted our patient's facial nerve to a more intimate position against adjacent brainstem vascular structures, which triggered an irritative, hyperexcitable response from the nerve. Perhaps if left untreated, eventual facial nerve palsy would have developed from progressive compression and stretching of the facial nerve.

REFERENCES