Reversible facial pain due to hydrocephalus with trigeminal somatosensory evoked response changes

Case report

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A patient with hydrocephalus due to aqueductal stenosis suffered facial pain which was relieved after the insertion of a ventriculoperitoneal shunt. The trigeminal somatosensory evoked response (TSER) of the affected side showed lower amplitudes and longer latencies as compared to the unaffected side. Following surgery, the waves regained higher amplitudes and shorter latencies. An episode of shunt malfunction was accompanied by recurrent facial pain and impairment of the TSER. Both improved after revision of the shunt. The possible etiology of facial pain in patients with hydrocephalus is discussed.

KEY WORDS: aqueductal stenosis, facial pain, trigeminal nerve, somatosensory evoked response, hydrocephalus

Since the introduction of trigeminal somatosensory evoked response (TSER) monitoring as a routine test in various neurological conditions in our service, many hydrocephalic patients have been examined. In most, the TSER showed low amplitudes with no significant abnormal changes in the latencies of the waves (unpublished data). However, in one of the hydrocephalic patients, who also presented with facial pain, the TSER from the affected side of the face showed changes in both amplitudes and latencies of waves. A description of this case follows.

Case Report

This 42-year-old woman was admitted to the neurosurgical service in June, 1981, with the chief complaint of chronic right-sided retro-orbital and facial pain which was constant and throbbing in nature. There were neither trigger points nor accompanying autonomic system signs. She also experienced two episodes of sudden dizziness followed by stupor, lasting for 2 hours, with spontaneous recovery. There were no convulsions.

On examination, the patient seemed to be in good general condition. The blood pressure was 120/70 mm Hg, pulse rate was 78/min, and the entire physical examination was within normal limits. The neurological examination, however, revealed bilateral optic disc pallor. There was facial hypesthesia over the areas innervated by the first and second trigeminal divisions on the right side. Corneal reflexes were prompt and equal. There was mild gait ataxia, and the deep tendon reflexes were increased. There were no other neurological findings. Skull films revealed mild separation of sutures and erosion of the dorsum sellae. Computerized tomography (CT) showed a huge symmetrical dilatation of the lateral ventricles, with mild enlargement of the third ventricle. The aqueduct and the fourth ventricle could hardly be visualized.

The TSER of the second trigeminal division on the right side, obtained by electrical stimulation of the upper lip, showed reduced amplitudes and longer latencies of the waves, as compared to the TSER obtained from the left side of the same lip (Fig. 1 upper traces). Following the insertion of a ventriculoperitoneal shunt, the patient improved, the ataxia disappeared, and the facial pain was relieved. One week later, the TSER showed increased amplitudes and shorter latencies of the waves (Fig. 1 lower traces).

The patient did well for a period of 6 months, when she was readmitted with symptoms of shunt dysfunction, namely nausea, unsteadiness, and double vision. She also complained of right facial pain. Examination
FIG. 1. Trigeminal somatosensory evoked responses (TSER). Upward deflection is negative. S = stimulation. Upper traces: The waves obtained from the affected side (broken line) prior to the insertion of a ventriculoperitoneal shunt have lower amplitudes and longer latencies, as compared to the TSER obtained from the unaffected side of the face (solid line). Lower traces: Following the insertion of the shunt, the TSER of both sides has equal amplitudes and latencies.

disclosed a patent, well functioning shunt system. A CT scan, however, showed severe hydrocephalus although the ventricular catheter was positioned correctly. The TSER again showed distorted waves obtained from the right lip, as compared to the TSER of the left side.

At a shunt revision procedure, the shunt system was found to be patent. However, the distal opening pressure was 140 mm H2O. Subsequently, the pressure system was changed to achieve adequate drainage of cerebrospinal fluid (CSF). Following surgery, clinical improvement was again accompanied by higher amplitudes and shorter latencies in the TSER.

Discussion

Maurice-Williams and Pilling reported three instances of hydrocephalus associated with paroxysmal facial pain and numbness, where relief of hydrocephalus by a CSF shunt led to remission of pain. They postulated that the paroxysmal pain was due to stretching of the trigeminal sensory root by the displacement of the brain stem secondary to hydrocephalus. The authors suggested that short-circuiting the electrical impulses between fibers of a similar size in the stretched sensory root might have been responsible for the trigeminal pain. Higher than usual petrous ridges may, according to these authors, have an additive stretching effect on the trigeminal nerve.

Tucker, et al., also reported two patients in whom unilateral trigeminal neuralgia was the only presenting symptom of hydrocephalus due to aqueductal stenosis. The pain disappeared following the insertion of a CSF shunt, but recurred with shunt dysfunction. They mentioned three additional cases reported by Seeger, in which trigeminal neuralgia was associated with hydrocephalus; one of these cases also had aqueductal stenosis. Tucker, et al., also related the neuralgia to stretching of the trigeminal sensory root due to a shift of the brain stem as a result of pressure gradients between supra- and infratentorial compartments.

The fact that trigeminal pain is rarely encountered in hydrocephalic patients may, however, shed doubts on the stretching theory, which also would not explain the unilaterality of the pain.

The TSER in our patient showed reduced amplitudes and delayed latencies of the waves obtained from the affected side of the face whenever the intracranial pressure (ICP) was elevated, either before the insertion of a shunt or due to shunt dysfunction. The amplitudes increased in size and the latencies shortened following relief of elevated ICP. Interestingly, similar electrophysiological findings of lower amplitudes and longer latencies have been reported by Stöhr, et al., and by Bennett and Jannetta in patients with idiopathic trigeminal neuralgia. They found these results indicative of pressure on the trigeminal nerve root in the posterior fossa, which leads to local demyelination resulting in trigeminal pain. It is possible that the facial pain in hydrocephalic patients is also caused by local compression and irritation of the trigeminal nerve root by an adjacent aberrant or atherosclerotic blood vessel. Thus, relief of ICP will enable this blood vessel to move away from the nerve root, with subsequent relief of pain. Shunt malfunction with concomitant raised ICP will again cause a shift of the vessel toward the nerve root, with recurrence of pain.

The relative rarity of aberrant blood vessels in the vicinity of the trigeminal nerve root may account for the small number of hydrocephalic patients who also present with unilateral facial pain. Thus, pressure over the trigeminal sensory root, rather than stretching of the nerve fibers, may account for the unilateral facial pain encountered in hydrocephalic patients. This is further supported by the TSER monitoring.

References

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