Abnormal movements with hydrocephalus

Case report

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The author reports the case of a pituitary dwarf with choreoathetoid movement of the tongue and both arms as results of a shunt malfunction. He had undergone craniotomy for excision of a suprasellar craniopharyngioma, and 13 years later complete cerebral angiography and Conray ventriculography showed massive hydrocephalus with no evidence of tumor recurrence. Revision of the shunt was followed by complete recovery of the movement disorder.

KEY WORDS • hydrocephalus • abnormal movements • dyskinesia

Several secondary manifestations of regional brain damage in severe hydrocephalus have been described. I wish to report a case in which shunt malfunction led to the development of an abnormal movement disorder that was reversible by revision of the shunt. In review of the literature, I did not encounter a similar case report.

Case Report

Medical History. In July, 1956, a 4-year-old boy presented with headaches. A tentative diagnosis of craniopharyngioma was made owing to the presence of calcification in the suprasellar area. In January, 1957, he developed bilateral papilledema. A ventriculoatrial shunt was carried out subsequent to a pneumoencephalogram. The shunt has required multiple revisions.

In February, 1960, he was examined because of sudden onset of lethargy. Funduscopic examination showed bilateral optic atrophy. Ventriculography was followed by aspiration of approximately 65 cc of oily material from the tumor. Bifrontal craniotomy established the clinical diagnosis and confirmed the presence of a large cyst in the left frontal lobe as well as solid tumor in the third ventricle. A gross total removal was performed. Postoperatively he developed transient diabetes insipidus and since then he has been on maintenance doses of cortisone and thyroid.

In September, 1961, a right-sided focal seizure evolved into a grand mal convulsion. At this time his shunt was not functioning and it was revised. Since then he has been on anticonvulsants and no subsequent seizures have been reported.
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Present Admission. In October, 1973, the patient was readmitted with a 3-week history of lethargy, frontal headaches, and occasional vomiting.

Examination. His stature was that of a 6- to 8-year-old boy. Ophthalmoscopic examination showed occasional perpendicular and rotary nystagmus with bilateral optic atrophy. There was a trombone-like movement of the tongue, two to three per second, and a choreoathetoid movement of the left and right hand when extended. He had 3+ hyperreflexia on both sides symmetrically. Plantar reflexes were flexor. His gait was broad-based and unsteady.

Among pertinent laboratory studies, Dilantin and phenobarbital in the serum were below toxic level. The T-3 and T-4 vertebrae were normal. Conray shuntgram showed delayed emptying (greater than 9 minutes). Cerebrospinal fluid could be aspirated from the valve and there were no obvious breaks in the continuity of the shunt tubes. Right carotid arteriogram with cross compression showed symmetrical hydrocephalus. Conray ventriculogram showed enlarged lateral and third ventricles (Fig. 1).

Operation. Although the ventricular end of the shunt was patent, it was revised along with the valve, which was felt to be incompetent. There was steady improvement of the neurological dysfunction in the postoperative period. The tongue movement improved in 24 hours and within 48 hours all abnormal movement had disappeared.

He continues to do well with no recurrence of movement disorders when last seen in March, 1976.

Discussion

This case represents an unusual manifestation of shunt dysfunction. The tremor of the tongue was two to three per second, and increased when the patient tried to protrude his tongue. The tremor could be stopped on command but it would return in 5 to 10 seconds. The dyskinesia in our patient was similar to the “bobble-head doll syndrome” which presents a flexion-extension movement of the head and neck on the trunk at a rate of two to three per second. This movement disorder has been reported to be associated with third ventricular dilatation from a cyst or hydrocephalus.6-9 In obstructive hydrocephalus the suprapineal recess of the third ventricle may become markedly dilated and fill the quadrigeminal cistern, and may produce difficulties with upward gaze.5,10,11 Endocrine abnormalities in hydrocephalus presumably arise from compression of hypothalamus.1

The shuntgram was valuable in recognizing the faulty valve in the absence of any obvious disconnection, kink, and/or sleeve formation. Recently Dewey, et al.,3 discussed the value of the shuntgram in recognizing shunt dysfunction.

The dorsomedial nucleus of the thalamus occupies a large paraventricular area that could be compressed with a dilated third ventricle. The reports of stimulation studies have shown a somatotropic motor pattern in this nucleus, with legs most rostral and lateral and the head and neck areas most caudal and medial.7,8 It has been suggested that abnormal impulses in the bobble-head syndrome originate from the medial aspect of the dorsomedial thalamic nucleus, relay to the basal ganglion, and from there to motor centers.6,7 It is possible that the movement disorder in our patient may have resulted from the massive enlargement of the third ventricle precipitating a similar mechanism.
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References


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