Sialorrhea from fourth ventricle hydrocephalus

Case report

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The case of a 50-year-old man with tetraventricular hypertensive hydrocephalus is presented, remarkable for fourth ventricle dilatation. This patient showed a significant sialorrhea as the main symptom, which is quite unusual. This condition was successfully treated by cerebrospinal fluid diversion. The uncommon features of this case are summarized.

Key Words: hydrocephalus · sialorrhea · fourth ventricle

Symptoms of hydrocephalus in adults are generally related to intracranial hypertension, mental deterioration, and gait and sphincter disorders. The intriguing case presented here involved massive sialorrhea due to hypertensive hydrocephalus, with prominent fourth ventricle enlargement. The condition resolved after cerebrospinal fluid (CSF) diversion.

Case Report

This 50-year-old man had been suffering from sialorrhea for 1 year, and gait instability and sporadic vomiting had occurred during the month before presentation. No head trauma, meningitis, hemorrhage, tumor, or other central nervous system disorder was evident in his medical history. The only remarkable diseases reported were brucellosis and typhus during his childhood. There was nothing noteworthy in his family history.

Examination. Neurological examination revealed unsteadiness on eye closure only. Routine laboratory profiles were normal. Magnetic resonance imaging showed tetraventricular hydrocephalus (Fig. 1 left). The fourth ventricle was especially enlarged, with impingement on its floor and dilatation of the lateral recesses. No expansive lesions were demonstrated in the posterior fossa.

Treatment and Course. The patient underwent external ventricular drainage for 1 day. Intracranial pressure recorded during the first hours ranged between 18 and 22 mm Hg. Cytological and chemical studies of the CSF were normal. Immediately after drainage was discontinued, the sialorrhea and ataxia vanished.

Two months later, sialorrhea recurred. This time it was more serious and disabling than before treatment, and was associated with vomiting, hiccupping, headache, and ataxia. A definitive ventriculoperitoneal shunt was placed, producing rapid resolution of all symptoms.

At his 1-year follow-up evaluation, the patient was symptom-free. Magnetic resonance imaging demonstrated that the hydrocephalus had resolved and a normal anatomical pattern of the fourth ventricle had been restored (Fig. 1 right).

Fig. 1. Left: Magnetic resonance image demonstrating tetraventricular hydrocephalus. The fourth ventricle is particularly enlarged and shows a significant “scalloping” of its floor. Right: Image obtained 1 year after insertion of a ventriculoperitoneal shunt showing resolution of the hydrocephalus, with normalization of the fourth ventricle.
Discussion

Tetraventricular hypertensive hydrocephalus with prominent fourth ventricle enlargement is a rare condition in adults. To the best of our knowledge, the occurrence of this condition in association with a dysautonomic sign, such as sialorrhea, is unique and puzzling.

In the case presented, the etiopathogenesis of hydrocephalus is not clearly defined. We could not identify a congenital etiology or an acquired condition, such as neoplastic, traumatic, or hemorrhagic disease. We also discarded an infective origin (such as brucellosis), an association that has never been described in the literature. We postulate that a membranous obstruction of the foramina of Luschka and Magendie may have produced a subcritical reduction of the CSF flow from the fourth ventricle. Eventually, the decrease in CSF was accompanied by manifest symptoms. There was also a suggestive association between the impingement on the fourth ventricle floor and the underlying salivary nuclei of the facial and glossofaryngeal nerves (Fig. 2). It appears that the patient’s unusual symptom of sialorrhea was associated with a state of irritation of the salivary nuclei, although the pathogenesis of the latter is not clear, and was produced by “scalloping” of the fourth ventricle floor.

We deemed it possible to restore the CSF flow via temporary external drainage, with a good short-term (2 months) effect. At that time, sialorrhea recurred and was treated successfully and definitively with insertion of a medium-pressure ventriculoperitoneal shunt. The fourth ventricle returned to its normal size and the sialorrhea resolved.

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References


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