CASE REPORT

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Hydrocephalus: A Fatal Late Consequence of Mumps Encephalitis

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ABSTRACT: Common and usually self-limited diseases may occasionally have fatal consequences. Hydrocephalus is a very rare complication of mumps, with just a few cases reported in the literature. Here we report a fatal case of hydrocephalus presenting 19 years after mumps encephalitis. The long latency period between encephalitis and hydrocephalus-associated symptoms makes this case particularly interesting.

KEYWORDS: forensic science, hydrocephalus, encephalitis, mumps, autopsy, headache

Headache is one of the most common neurological symptoms. In most cases headaches are benign, but differential diagnoses lists include more than 300 different causes (1). In Western countries, it is considered that 80% of the population have frequent headaches, and the most frequent causes are migraine (52%) and tension (32%). Other common disorders include trigeminal neuralgia, postinjury headache, cluster headache, and subaracnoid haemorrhage.

Hydrocephalus, the abnormal enlargement of the cerebral ventricles, accompanied by an increase in cerebrospinal fluid (CSF) volume, is an uncommon cause of headache. In fact, hydrocephalus-related headaches represent only 0.06% (2). Most cases are due to obstruction of the CSF flow or a decrease in its absorption in the arachnoid villi. In infants and young children hydrocephalus may produce an enlarging head, tense fontanelles, and distended scalp veins. After suture closing, clinical manifestations may include lethargy, vomiting, headache, papilledema, blurring vision, diplopia, tinnitus, and gait apraxia. The hallmark for the diagnosis of hydrocephalus is enlargement of the ventricular system, best demonstrated by MRI or CT scanning.

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Case Report

A 30-year-old male went into the emergency service complaining of headache for the past few days. Nineteen years earlier he had been hospitalized because of mumps complicated with meningoencephalitis. His past medical history was otherwise unremarkable. The general physical exam was normal. The neurological exam showed mild horizontal nistagmus, hyperreflexia and slightly unstable gait. There were no sensory or strength deficits, nor papilledema. A skull X-ray was normal. He was given acetaminophen, and a brain CT scan and a follow-up visit at the neurology outpatient clinic were scheduled. However, four days later he suffered a cardiac arrest at home. He was taken again to the emergency service, but resuscitation maneuvers were unsuccessful. A postmortem examination was performed the next morning. Doctors involved were reported by the family for a possible medical malpractice.

At autopsy, the lungs showed diffuse oedema and focal intraalveolar hemorrhages. There were no other abnormalities in the thorax or the abdomen. The meninges appeared normal. The brain was swollen, with marked enlargement of the lateral and third ventricles (Fig. 1). The Sylvius aqueduct was slightly dilated; the fourth ventricle was not enlarged. There were no signs of herniation. The microscopic exam showed subependimal gliosis in IV ventricle's floor (Fig. 2), residual lymphocyte meningitis (Fig. 3) and minimal hemorrhage in locus coeruleus.

It was concluded that death was due to hydrocephalus with intracranial hypertension, probably related to previous encephalitis.

Discussion

Although it is not possible to obtain definitive proof of a causal relationship between past mumps infection and the hydrocephalus, this seems to be the more likely explanation in the present case. After the report by Dandy in 1920 (3) suggesting the role of inflammatory processes in the pathogenesis of hydrocephalus, several animal and clinical studies have given support to this concept (4–6). The role of the mumps virus in the pathogenesis of acquired aqueductal stenosis was demonstrated by Johnson in animals inoculated with virus (7). Mumps virus seems to have a special tropism for ependymal cells, as has been shown in some animal models (8). After the first case reported by Timmons and Johnson (6), several ad-

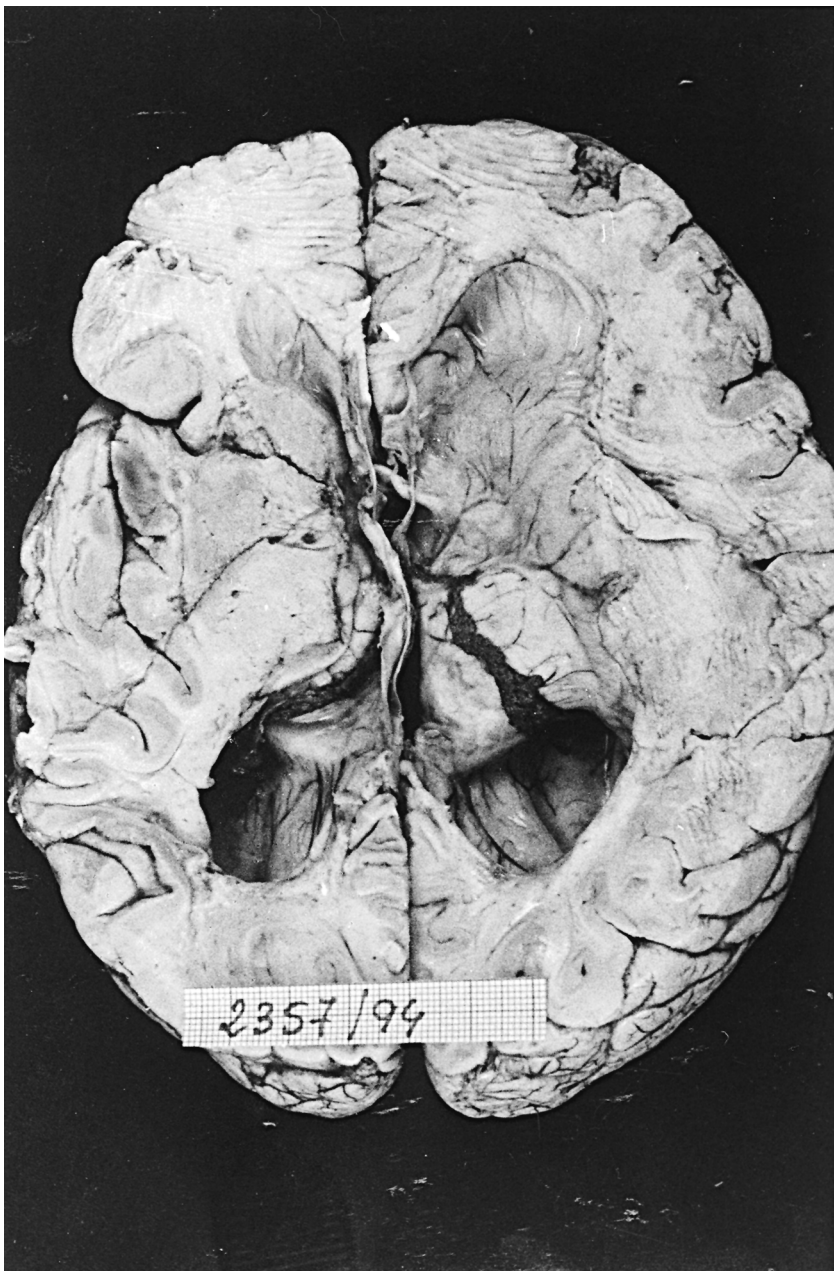

FIG. 1—*Section of the brain showing marked enlargement of lateral ventricles.*

ditional cases have been published describing the association between hydrocephalus due to aqueductal stenosis and mumps (4–6,9–11). Thus, Oran et al. found 13 reports in the literature (12). The time interval between the clinical mumps infection and the diagnosis of the hydrocephalus ranged from 3 weeks to 4 years. However, Viola et al. (11) recently reported a case of acute hydrocephalus developing immediately after a clinically evident mumps infection. The long interval between mumps infection and hydrocephalus manifestations is an interesting peculiarity of the present case.

Although most patients with headaches have benign, and often self-limited, conditions, the subtle abnormalities found in the neurological examination of this patient might have suggested the existence of a structural disorder. However, there were no objective signs of intracranial hypertension, thus a CT scan was scheduled on a nonurgent basis. Overruling of the malpractice report was based on the widely accepted idea that urgent CT is not routinely indicated in young subjects with headache (13,14). Nevertheless, a high index of suspicion remains mandatory in patients with recent-onset headache in order to prevent fatal outcomes. This is exemplified by the present case, which also illustrates how long may be the latent period between mumps encephalitis and clinically evident hydrocephalus.

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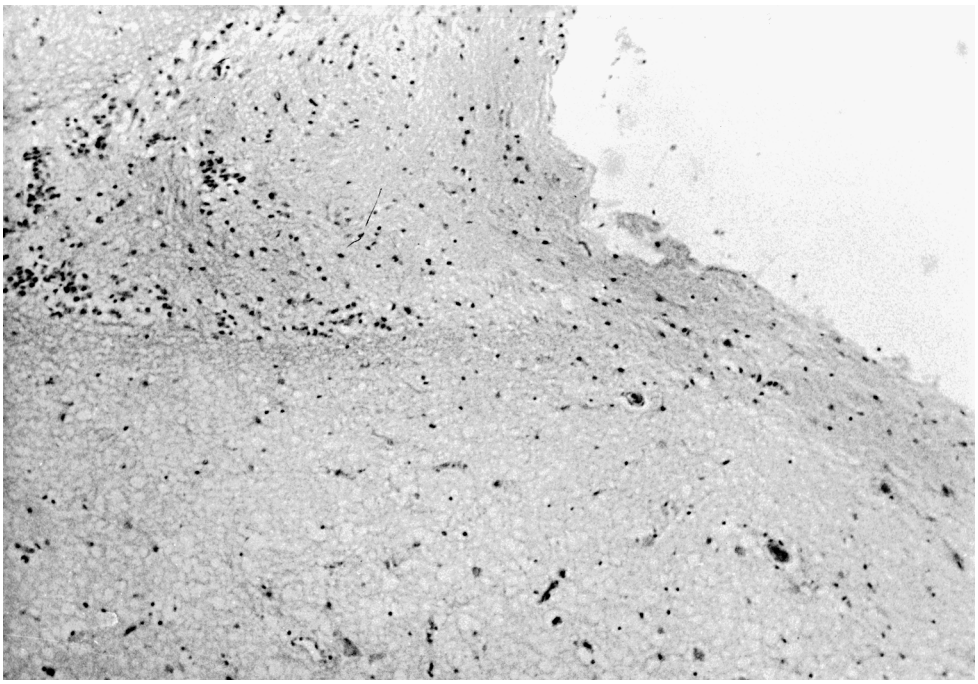

FIG. 2—*Subependymal gliosis at IV ventricle floor (H-E, ×10).*

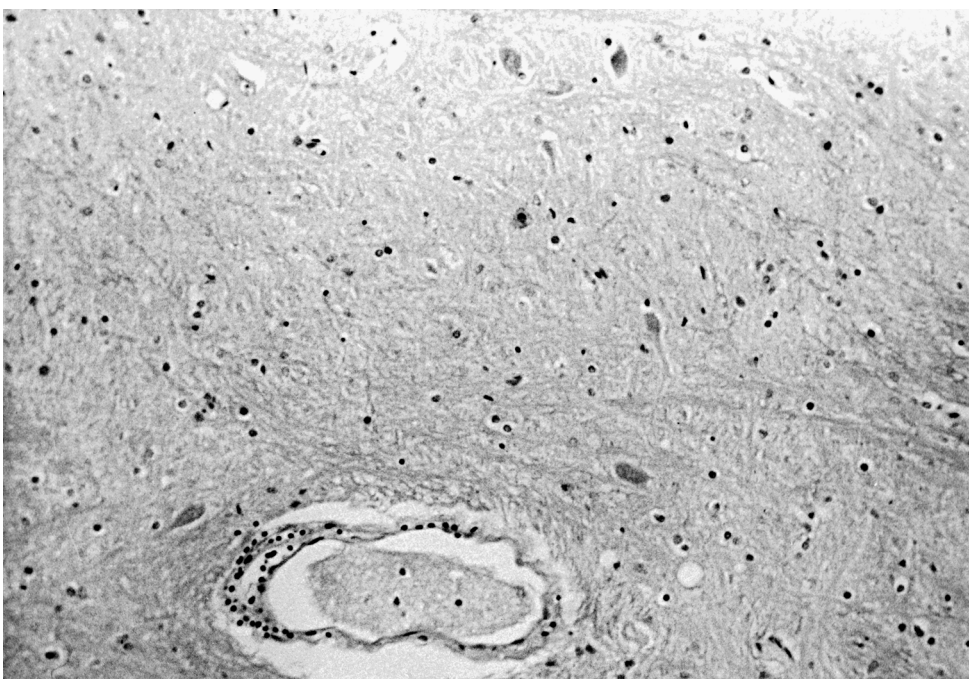

FIG. 3—*Residual perivascular lymphocytic infiltration (H-E, ×10).*

References

1. Evans RW. Diagnostic testing for the evaluation of headaches. Neurol Clin 1996;14:1–26.
2. Rasmusen BK, Jensen R, Scroll M, Olessen J. Epidemiology of headache in general population—a prevalence study. J Clin Epidemiol 1991; 44:1147–57.
3. Dandy, WE. The diagnosis and treatment of hydrocephalus resulting from strictures of the aqueduct of Sylvius. Surg Gynecol Obstet 1920;31:340–8.
4. Bray, PF. Mumps: a cause of hydrocephalus? Pediatrics 1972;49:446–7.
5. Thompson JA. Mumps: a case of acquired aqueductal stenosis. J Pediatr 1979;94:923–4.
6. Timmons, GD, Johnson KP. Aqueductal stenosis and hydrocephalus after mumps encephalitis. N Engl J Med 1970;283:1505–7.
7. Johnson RT, Johnson KP, Edmonds CJ. Virus-induced hydrocephalus: development of aqueductal stenosis in hamsters after mumps infection. Science 1967;157:1066–7.
8. Uno M, Takano T, Yamano T, Shimada M. Age-dependent susceptibility in mumps—associated hydrocephalus: neuropathologic features and brain barriers. Acta Neuropathol (Berl) 1997;94:207–15.

9. Lahat E, Aladjem M, Shiffer J, Starinsky R. Hydrocephalus due to bilateral obstruction of the foramen of Monro: a 'possible' late complication of mumps encephalitis. Clin Neurol Neurosurg 1993;95:151–4.
10. Spataro RF, Lin SR, Horner FA, Hall CB, McDonald JV. Aqueductal stenosis and hydrocephalus: rare sequelae of mumps virus infections. Neuroradiol 1976;12:11–3.
11. Viola L, Chiaretti A, Castorina M, Tortorolo L, Piastra M, Villani A, et al. Acute hydrocephalus as a consequence of mumps meningoencephalitis. Pediatr Emerg Care 1998;14:212–4.
12. Oran B, Ceri A, Yilmaz H, Kabakus N, Aycicek A, Erkul I. Hydrocephalus in mumps meningoencephalitis: case report. Pediatr Infect Dis J 1995;14:724–5.
13. Sanchez-Precioso S, García-Canto E, Villaescusa O, Barbero P, Moreno JA, Mulas F. A retrospective study of infant headache. Rev Neurol 1995;23:764–8.
14. Nevo, Y, Kramer, U. Rieder-Groswasser I, Harel, S. Clinical categorization of 312 children with chronic headache. Brain Dev 1994;16:441–4.

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