Mumps hydrocephalus ameliorated with external drainage

Güler Kanra[1], Metehan Özen[1], Ateş Kara[1], Serhan Küpeli[1], İlker Devrim[1]
A. Bülent Cengiz, Nejat Akalan[2]
Departments of [1]Pediatrics, and [2]Neurosurgery, Hacettepe University Faculty of Medicine, Ankara, Turkey

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Mumps is still one of the most common childhood diseases in areas where national immunization has not been implemented. Although central nervous system manifestation is not so uncommon, hydrocephalus secondary to mumps is very rare. In this report, we present a toddler who developed severe hydrocephalus during mumps infection, which resolved via timely cerebral spinal fluid (CSF) drainage. We would like to emphasize early intervention for CSF external drainage.

Key words: mumps, hydrocephalus, external drainage.

Mumps is still one of the most common childhood diseases in developing countries where national immunization has not yet been implemented. Central nervous system (CNS) involvement is one of the most common manifestations of the infection[1]. In experimental animal studies, hydrocephalus secondary to the aqueductal stenosis in mumps infection has been very well documented[2-4], whereas only a limited number of human case reports are available in the literature[5,6]. Anti-edema therapy, aqueductal cannulation and ventriculo-peritoneal (VP) shunt are the interventions to overcome hydrocephalus. In this report, we present a toddler who developed severe hydrocephalus during mumps infection, which resolved via timely cerebrospinal fluid (CSF) drainage. To the best of our knowledge, this is the first case of mumps hydrocephalus ameliorated via external drainage.

Case Report

A four-year-old previously healthy boy presented with a two-day history of fever, vomiting and tendency to sleep and was hospitalized in a local hospital. Ceftriaxone was started for the presumptive diagnosis of bacterial meningitis and continued for six days. He developed stupor and lethargy in following days, and cranial computerized tomography (CT) scan on 4th day disclosed no pathology. He was referred to our clinic because of deterioration under antibiotic therapy.

On admission (hospital day 6), he was lethargic and disoriented. Meningeal irritation findings like neck stiffness and Kernig's and Brudzinski's signs were all positive, whereas deep tendon reflex and light reflex were normal. Complete blood count, serum biochemistry other than amylase, serum NH_3, and urinary and serum amino acid chromatography were within normal limits. CSF examination revealed pleocytosis with lymphocyte predominance [11 polymorphonuclear leukocytes (PMNLs) and 33 lymphocytes per mm^3], protein 285 mg/dl and glucose 49 mg/dl (simultaneous blood glucose was 102 mg/dl). Gram stain, acid-fast staining and latex agglutination indicated no microorganism, and culture remained sterile. Polymerase chain reaction (PCR) studies of CSF for herpes simplex and Coxsackie viruses, and *M. tuberculosis* were all negative. Detailed medical history revealed that the mother had symptomatic mumps infection four weeks before the onset of the patient's complaints. Serum anti-mumps IgM and IgG and CSF anti-mumps IgG were found positive.

The ceftriaxone therapy was continued on admission, and empiric acyclovir was started. He gradually evolved into unconscious state

(hospital day 11), and was responding only to painful stimulation by withdrawal. Afterwards, he developed spastic quadriparalysis. On the same day, cranial magnetic resonance imaging (MRI) disclosed findings compatible with meningoencephalitis and triventricular hydrocephaly (Fig. 1). Upon measuring CSF opening pressure as 210 mm CSF, external drainage was implanted into lateral ventricle since the patient's hemodynamics were too unstable for general anesthesia. His neurologic examination improved rapidly within two days, and external drainage was extracted after six days, as he was conscious enough to tolerate mild daily activities. A VP-shunt was replaced on the 19th day of hospitalization. He was discharged after eight days of its implantation. The patient survived well with VP-shunt and hydrocephalus regressed markedly in his cranial MRI after one month (Fig. 2). At one year of clinical follow-up, the patient had a normal neurologic examination.

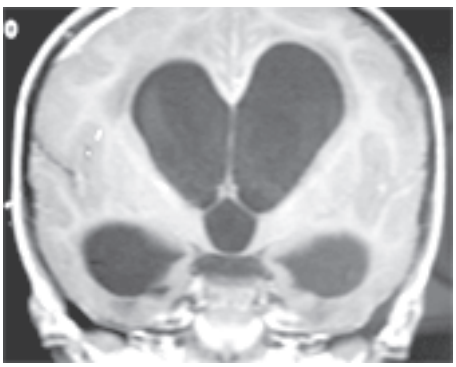

Fig. 1. Triventricular hydrocephaly on computerized tomography scan at 11th day of hospitalization.

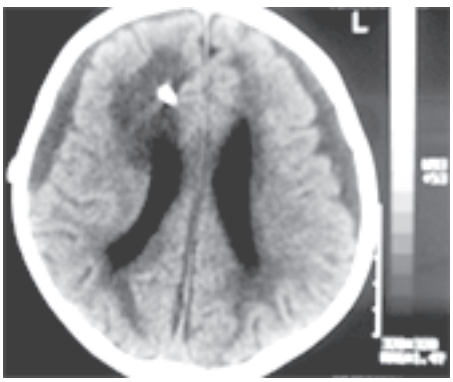

Fig. 2. Computerized tomography scan after ventriculo-peritoneal shunt.

Discussion

Mumps is an acute contagious disease and has predilection for glandular and nervous tissue. Even though some authors still suggest that CNS involvement is a complication of the infection, it is well known to be a manifestation rather than complication[1]. In a study by Bang and Bang[7], 62% of symptomatic mumps cases have an increased number of cells upon CSF examination, while only 28% have CNS symptoms. Mumps meningoencephalitis is usually encountered 3-10 days after parotitis, and characterized by fever, vomiting, nuchal rigidity and change in sensorium. Convulsions and Kernig's and Brudzinski's signs can be rarely elicited.

However, mumps meningoencephalitis may precede or even occur in absence of glandular involvement. This case had no sign of parotitis, but recent mumps infection of the mother and relevant antibody positivity in blood and CSF were suggestive of an active disease.

Hydrocephalus is extremely rare following mumps meningoencephalitis[8]. In addition to various animal studies reporting hydrocephalus secondary to aqueductal stenosis, some recent articles have shown that hydrocephalus in mumps infection is intimately related to the maturity of brain barriers[9]. A limited number of human cases in the literature report occurrence of hydrocephalus particularly secondary to the aqueductal stenosis in mumps infection. We demonstrated triventricular hydrocephalus, which might suggest aqueductal stenosis.

Regarding the treatment modalities, anti-edema therapy, aqueduct cannulation and replacement of VP-shunt are documented interventions. If untreated or if medical intervention is delayed, this complication may end in fatality[10,11]. We preferred external drainage implantation because of the poor general condition of the patient. He responded very well to the drop in his CSF pressure and improved rapidly. We extracted it five days after intervention. However, the patient required a VP-shunt later in his clinical course, which helped him to survive without sequelae. The external drainage enabled this patient to achieve a more stable hemodynamic condition and undergo further medical approaches.

In conclusion, mumps meningoencephalitis is usually a self-limited condition, but occasionally may yield serious neurologic sequelae. Unusual

neurologic findings must alert the physician to possible neurologic complications. We would like to emphasize the importance of cranial MR imaging in follow-up of severe mumps meningoencephalitis cases, even though initial imaging is within normal limits. External drainage should be preferred in selective cases until such time when the VP-shunt, the gold standard for increased CSF pressure secondary to aqueductal stenosis, is implanted.

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