Case Report

Encephalomyelitis following mumps

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Study design: A case report with review of literature since 1961.

Setting: Departments of Neurology and Radiology, Faculty of Medicine, Zonguldak Karaelmas University, Zonguldak, Turkey.

Method: A 22-year-old male patient diagnosed and treated as mumps encephalomyelitis is described. The clinical findings of the patient and the difficulties in differential diagnosis are discussed with the help of previously reported eight adult cases.

Results: Increased T2 signals in the spinal cord from C4 to C6 and T5 to T10 were seen by magnetic resonance imaging. This was the largest spinal involvement in the reported mumps cases. He was treated with supportive therapy oral steroids and early rehabilitation. On the sixth month, his neurological examination revealed bilateral 2/5 paresis in lower extremities.

Conclusion: Encephalomyelitis following mumps is an uncommon but serious event in adults. *Spinal Cord* (2005) 43, 441–444. doi:10.1038/sj.sc.3101735; Published online 8 March 2005

Keywords: mumps; myelitis; encephalomyelitis

Introduction

Mumps virus is a common cause of meningitis. A viremia disseminates the virus to parotid glands and occasionally to submaxillary glands, testes, ovaries, mammary glands and leptomeninges. Nervous system involvement is uncommon and usually presents as meningitis or encephalitis. Myelitis, polyradiculitis and cranial neuritis are rarely reported.¹

Acute transverse myelitis (ATM) is an intramedullary spinal cord disease characterized by abrupt onset of rapidly progressive weakness of the lower extremities accompanied by a loss of sensation and sphincter control. Association with a large variety of infectious agents has been described. These include herpes simplex virus, Epstein–Barr virus, hepatitis B virus and cytomegalovirus.^{2,3} The myelitis syndromes may be subdivided into meningitis with myelitis, meningitis with anterior horn syndrome and myelitis without meningitis.¹ Mumps was seldom reported in association with myelitis. In this study, we present a case with diagnosis of the most rare neurological manifestation of mumps; encephalomyelitis.

Case report

A 22-year-old male was referred to our neurology intensive-care unit because of alteration in conscious-

ness and respiratory distress. His family reported progressive weakness and difficulty in walking with urinary retention beginning 4 days earlier. Behavioral changes (agitation and irritability) occurred on the third day and respiratory distress and deterioration in consciousness developed on the fourth day. He was diagnosed as mumps with parotitis and orchitis 2 weeks earlier. There was a family history of pulmonary tuberculosis in his father 10 years ago. The patient was not immunized for mumps.

Physical examination revealed neck stiffness and difficulty in respiration. His temperature was 38°C, pulse 100 per minute and respiration 24 per minute. Blood pressure was 110/70 mmHg. There was clouding of consciousness. Pupils were equal in size and reactive to light. There was bilateral downbeat nystagmus with right ptosis. Motor examination revealed 2/5 paresis of right arm and 0/5 paresis of legs. Tendon reflexes were suppressed in legs and plantar reflexes were absent. The patient had to be catheterized because of urinary retention.

On admission blood tests including full blood count and urinary examinations were normal. Sedimentation rate was 55 mm/h. Lumbar puncture showed normal opening pressure. Cerebrospinal fluid (CSF) contained 160 leukocytes/mm³ (100% mononuclear cells), 100 red blood cells/mm³, raised protein (78.7 mg/dl) and low CSF–blood glucose ratio (36, 102 mg/dl). No micro-organism was detected on Gram stain or subsequent

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culture. Cranial computerized tomography (CT) on admission was considered normal. Electroencephalogram showed generalized moderately slow activity. As a result of his poor general condition and respiratory distress, magnetic resonance imaging (MRI) could not be performed on the first 3 days. He was diagnosed as encephalomyelitis secondary to mumps. However, because of the CSF findings with low blood/CSF glucose ratios, presumptive diagnosis of tuberculous meningitis was also considered. He was treated with antituberculosis drugs, intravenous fluids, steroids (1 g/kg, orally), catheterization and frequent aspiration from tracheotomy. His consciousness and pulmonary distress was better on the third day. Cranial MRI (Philips Gyroscan, S15-ACS, 1.5 T) revealed high signal intensity in pons and superior medulla oblongata on T2-weighted and flair images and cervical–thoracic MRI revealed increased T2 signals in the spinal cord from C4 to C6 and T5 to T10. Axial T2-weighted MRI verified the intraspinal signal location (Figures 1 and 2a). Leptomeningeal enhancement in all thoracic segments was seen after gadolinium administration (Figure 2b).

Blood tests including C-reactive protein, vitamin B₁₂, folate and antibody panel for collagen vascular diseases (ANA, anti-sDNA, ENA, pANCA, cANCA) were all within normal limits. Anti-HIV antibody with ELISA method was negative. CSF examination, repeated on the 10th day of admission, revealed 20 leukocytes/mm³ (all mononuclear), 103.6 mg/dl protein and 38 mg/dl glucose (blood glucose = 102). Serological examination in blood and CSF for HSV-type 1–2, anti-CMV, anti-EBV, anti-Rubella Ig M and G, *Treponema pallidum*, *Mycoplasma pneumonia* and *Borrelia burgdorferi* were negative.

Figure 1 Axial plane T2-weighted MR image of thoracic spine revealed increased signal within the spinal cord

Figure 2 Sagittal MRI of the spine. An intramedullary high signal lesion is present from 5th to the 10th thoracic vertebrae on T2-weighted image (a). Leptomeningeal enhancement in all thoracic segments was seen after gadolinium administration on T1-weighted image (b)

Serological testing of CSF on the 21st day showed negative IgM and positive IgG response to mumps virus.

On the 7th day of treatment, he was conscious and talking properly. His motor examination revealed 2/5 paresis of right arm and 1/5 paresis of legs. A comprehensive rehabilitation program was given, his right arm improved on the second month, but the legs did not improve. Despite aggressive wound care and supportive therapy, decubitus ulcers on both buttocks developed. He had reconstructive surgery on the fifth month. On the sixth month, his neurological examination revealed bilateral 2/5 paresis of legs with bladder and bowel disturbance.

Literature review

We reviewed the foreign and English-language literature from 1961 to January 2004 with use of MEDLINE and secondary references. The key words used were mumps (cross-linked with myelitis and encephalomyelitis) and central nervous system (CNS) (cross-linked with mumps). This review yielded 26 more cases presenting with mumps-related myelitis. Of these, 18 were children and eight were adults. Of these cases, 17 (10 ≤ 17 years and seven > 17 years) were reported in Scheids' series in 1961.⁴ We also found three more patients diagnosed as acute transverse myelitis and Guillain–Barre overlap syndrome.^{5,6}

Discussion

Of all patients with mumps parotitis, at most 10% develop clinical meningitis. Much less frequently, mumps may be complicated by acute encephalitis, which is generally mild, without focal signs, and with low mortality and few sequelae. A still more rare but more severe encephalitic syndrome presenting as a form of

Table 1 Reported adult transverse myelitis patients following mumps viremia

Authors	Case no.	Sex	Age (years)	Days following mumps	Level of ATM	CSF findings			Therapy	Outcome
						Cells	Protein (mg%)	Glucose (mg/dl)		
Scheid (1961)	1	F	24	25	T4	NM	NM	NM	NM	Spastic gate (4 years)
	2	F	39	11	T5	5	NM	NM	NM	Paraparesis (4 months)
	3	F	37	3	Weakness of lower extremities	2	140	NM	NM	Death
	4	M	37	3	C?	15	51	NM	NM	Death
	5	M	19	4	None	25	NM	NM	NM	Recovery
	6	F	29	120	T4 and cranial nerve (VII) paresis	30	NM	NM	NM	Recovery
	7	M	19	10	Paraparesis and areflexia	10	NM	NM	NM	Recovery
Vanketasubramanian (1997)	8	M	38	21	T7	23 ^a	55	52	0.5g/d metpred (5 days)	Recovery (2 weeks)
Present case	9	M	22	10	C4+ encephalitis	260 ^b	78	36	1 mg/kg prednisolone/orally	Sequela 2/5 Paraparesis (sixth month)

Vanketasubramanian N has reported hyperintensity from T7 to T11

^aCSF cell types: mainly lymphocytes

^bCSF cell types: 160 lymphocytes, 100 red blood cells

NM = not mentioned; ATM = acute transverse-myelitis; pred = methylprednisolone

immune-mediated postinfectious encephalomyelitis follows an estimated 1 in 6000 cases and develops 7–15 days after parotitis.⁵ A high percentage of these patients exhibit seizures, hemiparesis, severe obtundation and a mortality of 10%.⁴

While well recognized among children, ATM following mumps in adults has been poorly reported. Scheid⁴ has reviewed the neurological manifestations of mumps and discussed the 17 myelitic cases, including suspected ones, between 1914 and 1961. Seven adult cases from his series are presented in Table 1. The diagnosis was made according to clinical findings and mumps in history and the treatment regimen used was not mentioned. We could find only one more adult case diagnosed as myelitis following mumps since then⁷ (Table 1).

The oldest patient was a 39-year-old female and was reported in Scheid's review. The time interval between clinical symptoms of mumps and myelitis varied between 1–2 days and 60 days, mostly in the first 2 weeks. All reported cases presented with neurological findings suggesting acute development of transverse-myelitis without encephalitis. This patient in this particular case represents a further diagnostic complexity in that he had features of ascending myelitis symptoms followed by encephalitis on the fourth day.

A history of progressive ascending myelitis with meningitis and CSF examination with hypoglycorrhachia, high protein and lymphocytic predominance suggests tuberculosis is still a public health issue in our country and CNS tuberculosis needs early diagnosis and treatment. Although a recent history of parotitis was supporting the diagnosis of mumps-related encephalomyelitis, we urgently began antituberculosis therapy. In the follow-up, no tuberculosis-related laboratory result was found. Repeated CSF examination on the 10th day revealed less lymphocytic cells. In CNS tuberculosis with successful treatment, the CSF slowly returns to normal. CSF cell counts are mostly reduced by 50% during the first month.⁸ Hypoglycorrhachia, as well as a left-shifted CSF pleocytosis, may be more common with mumps meningitis than other viral meningitides.⁴ Good recovery in CSF findings in our patient was also compatible with viral infection. However, none of the reported mumps cases had low CSF/blood glucose ratio in the literature.

The histological changes in mumps myelitis include dilatation of vessels, perivascular demyelination and proliferation of glial cells, degeneration of anterior horn cells and perineural edema.⁷ In ATM, the neurological changes are prominent at the thoracic or lumbar regions and the higher levels reveal more edema and demyelination. The findings of focal cord edema and increased T2 signal on MRI are typical of myelitis. Gadolinium enhancement is not always seen. The wide availability of MRI allows rapid and accurate diagnosis of myelitis, and excludes other conditions mimicking it such as spinal cord compression or intramedullary lesions such as syringomyelia, hematomyelia or tumor. In most of the previously reported cases, myelitis was localized in

the thoracic region. The presented case is the second one with the involvement of cervical and thoracic regions. Extension of signal changes in MRI have been found to be correlated with the severity of myelitis and its outcome.⁹

Although the advantage of intravenous bolus of methylprednisolone has been illustrated in transverse-myelitis due to multiple sclerosis, there are also studies that do not suggest a beneficial role of methylprednisolone on the outcome of ATM.¹⁰ However, the rarity of disease prevents the design of randomized studies that demonstrate the beneficial effect of corticosteroids in mumps myelitis. Steroid therapy was used in only three reported mumps myelitis cases and they recovered fairly quickly.^{7,11,12} We preferred oral corticosteroids in our patient. We believe that the antiedema and anti-inflammatory effects of corticosteroids were an important part of our therapy. However, the possibility of natural recovery cannot be entirely excluded. The supportive therapy and comprehensive rehabilitation programs are also very important.

In conclusion, encephalomyelitis following mumps is an uncommon event in adults. This virus should always be considered among possible etiologic agents causing such neurological syndromes.

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