

Bilateral Neuroretinitis Associated With Mumps

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Background: Involvement of the optic nerve is a rare complication of mumps infection.

Objectives: To report a case of bilateral neuroretinitis complicating a mumps infection and to review 5 previously reported cases.

Design: Case report and literature review.

Setting: Tertiary hospital.

Patient: A 7-year-old girl had sudden-onset blindness due to bilateral neuroretinitis. Approximately 3 weeks prior to the initial examination, she developed a self-limited febrile illness with parotid swelling and subsequent meningoencephalitis.

Results: Mumps was determined to be the underlying cause of the meningoencephalitis and bilateral optic neuritis because of the exposure history in this nonvaccinated child, the typical clinical signs and symptoms, and the positive serologic test results. Recovery of visual function was gradual but nearly complete.

Conclusions: Physicians should be aware that optic nerve involvement may be a manifestation of mumps infection. The delayed onset of optic neuritis, the bilateral involvement, and the near complete recovery suggest an immune-mediated pathogenesis.

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MUMPS (epidemic parotitis) is an acute communicable disease caused by a paramyxovirus, characterized by painful enlargement of the salivary glands, particularly the parotid glands.¹ Neurologic and ocular manifestations of mumps are well documented²⁻¹³ but seldom appear concomitantly.^{6,8,9,11,12} Meningitis and mild meningoencephalitis are the most frequent complications of mumps in children.¹ Optic neuritis is a rare neurologic complication following mumps infection and is manifested clinically by decreased visual acuity, decreased color perception, and visual field defects.^{5,7} Funduscopic examination may show marked edema of the head of the optic nerve (papillitis) and retina (neuroretinitis) or an entirely normal appearance (retrobulbar neuritis).^{5,6,8,9,11,12}

We describe a 7-year-old girl in whom meningoencephalitis was followed by bilateral neuroretinitis secondary to a mumps infection. We also review the clinical spectrum, response to therapy, and visual se-

quelae of previously reported pediatric cases of optic neuritis complicating mumps.

REPORT OF A CASE

A 7-year-old girl from Mumbai, India, was admitted to our hospital with a 1-day history of sudden-onset blindness. Three days prior to admission, she was discharged from a local hospital after a 2-week stay for management of acute bacterial meningitis. Her clinical signs and symptoms included severe headache, vomiting, and altered mental status. There was no history of trauma, seizure, or skin rash. Therapy with intravenous ampicillin (200 mg/kg per day) and cefotaxime (300 mg/kg per day) for 2 weeks and mannitol for the initial 3 days of hospitalization resulted in complete recovery. The cerebrospinal fluid (CSF) profile was consistent with viral meningitis rather than bacterial meningitis. The CSF analysis revealed a white blood cell count of $15 \times 10^3/\mu\text{L}$ (20% neutrophils and 80% lymphocytes), glucose level of 71 mg/dL (3.94 mmol/L), and pro-

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Magnetic resonance image of the brain (T2-weighted image) showing signal hyperintensity in the left optic radiation (arrow).

tein level of 46 mg/dL. The gram stain was negative for bacteria and the CSF culture was sterile. A detailed history from her parents revealed that the patient had developed a febrile illness with bilateral submandibular swelling 1 week prior to the earlier hospitalization. On evaluation by a family physician, a diagnosis of submandibular adenopathy secondary to pharyngitis was made, and the patient had received an oral antibiotic (drug unknown). On further probing it emerged that while our patient was being treated for meningitis in the hospital, her older sibling had developed mumps characterized by fever and bilateral parotid enlargement. Both children were unimmunized against mumps.

Physical examination findings revealed a conscious but irritable child with a temperature of 36.7°C, heart rate of 110 beats/min, respiratory rate of 22/min, and a blood pressure of 110/74 mm Hg. Examination of visual acuity revealed no perception of light in both eyes. Both pupils were fixed, dilated, and not reactive to light. The anterior chambers appeared normal. Fundusoscopic examination showed bilateral optic disc edema with obliteration of the disc margins. There were scattered hemorrhages against a hyperemic background. The ocular findings were confirmed by an ophthalmologist. There were no signs of meningeal irritation and no focal neurologic deficits.

In view of the exposure history in a nonvaccinated child with subsequent development of self-limited febrile illness with parotid swelling, mumps was suspected as the underlying cause of meningoencephalitis and bilateral optic neuritis. Pertinent diagnostic evaluation included a repeated CSF examination, which was unremarkable and showed complete resolution of the pleocytosis. The CSF gram stain was negative for organisms and the culture was sterile. A polymerase chain reaction test for *Mycobacterium tuberculosis* on the CSF specimen was negative. Results of a chest radiograph were

unremarkable. Magnetic resonance imaging (MRI) of the brain showed a focal linear hyperintense lesion in the posterior part of the left optic radiation (**Figure**). The rest of the study results were reported as normal. A mumps-specific IgM antibody titer measured by enzyme-linked immunosorbent assay was positive at .125 mg/dL (1.25 g/L) (normal reference range, 54-222 mg/dL [<0.9 g/L]), confirming recent mumps infection. No specific therapy was offered and the patient was discharged to home after the prognosis was explained to the parents along with a plan for close follow-up. A follow-up specimen for mumps serologic analysis was not obtained. A repeated ophthalmologic evaluation performed 2 weeks later showed improvement of visual acuity, with presence of perception and projection of light in both eyes. Both pupils were normal in size but reacted slowly to light. Follow-up at the end of 6 weeks revealed a visual acuity of 6/60 OU. Direct and consensual reactions to light were normal, with no evidence of rapid afferent pupillary defect in either eye. Fundusoscopic examination showed residual pallor of optic discs, with resolution of bilateral retinal hemorrhages. Gradually, her vision improved, and after 3 months the visual acuity was 20/60 OD and 20/40 OS. There was, however, residual pallor of both optic discs. Findings from a visual evoked potential examination were normal and a repeated MRI study showed resolution of the hyperintense lesion along the left optic radiation.

COMMENT

The incidence of neurologic involvement in mumps is difficult to estimate, but CSF pleocytosis has been reported in 56% of patients with parotitis, while 10% have symptoms of meningitis.² In typical cases, mumps meningitis follows parotitis by 3 to 10 days,¹⁴ but neurologic symptoms may precede or even occur in the absence of salivary gland involvement. The illness is characterized by fever, headache, nausea, vomiting, signs of meningeal irritation, and only rarely, seizures. The outcome is generally benign, and the CSF usually shows pleocytosis, with a predominance of lymphocytes, and an elevated protein level, which in about 10% of cases is accompanied by hypoglycorrhachia.¹⁵ True encephalitis is rare, developing in about 0.2% of cases, although it is responsible for most of the central nervous system sequelae, including behavioral disturbances, persistent headache, seizure disorder, deafness, and optic atrophy.¹⁶

Ocular manifestations in mumps are rare but well recognized.⁷ Various parts of the ocular apparatus may be involved, such as dacryadenitis, optic neuritis, keratitis, iritis, conjunctivitis, and episcleritis.⁵⁻¹³ Optic nerve involvement ranks next to that of the lacrimal gland as the most common ocular manifestation of mumps^{7,14} and is usually a complication of a central nervous system infection.¹¹ The optic neuritis is usually bilateral, and may occur in 3 forms: papillitis, neuroretinitis, and the retrobulbar type.⁷ Papillitis is characterized by hyperemia of the optic disc, with edema. The nerve fiber layer of the retina is involved in neuroretinitis. Visual loss is the predominant clinical manifestation and may be accompanied by visual field defects, decreased color perception, and painful globe motion. In retrobulbar neuritis,

Clinical Characteristics and Outcome of Optic Neuritis Associated With Mumps*

Case No./ Age, y/Sex	Onset After Parotitis, d	Associated CNS Signs	Visual Acuity	Optic Disc	Therapy	Outcome Visual Acuity	Outcome Optic Disc	Year
1/11/F	35	None	Normal OD LP OS	Normal OD Edema OS	None	CF OS Normal OD	Atrophy OS	1907 ⁵
2/10/M	13	Encephalitis	NLP OU	Edema OU	None	20/20 OU	Pallor OU	1933 ⁶
3/7/F	14	Meningitis	HM OU	"Gray" OU	Dexamethasone	20/20, OU	Pallor OU	1974 ⁹
4/15/M	14	Meningitis	20/200 OD Normal OS	Edema OD Normal OS	Methylprednisolone	20/40 OD Normal OS	Edema OD Normal OS	1990 ¹¹
5/5/M	...	Encephalitis	Normal	Normal	Prednisolone	Asymptomatic	Asymptomatic	1991 ¹²
6/7/F	21	Meningitis	NLP OU	Edema OU	None	20/60 OD 20/40 OS	Pallor OU	2000†

*CNS indicates central nervous system; LP, light perception; CF, counting fingers; NLP, no light perception; HM, hand movements; and ellipses, not stated.

†Current study.

the optic disc is spared and only the axial portion of the optic nerve is involved. The clinical signs and symptoms include central scotoma, leading to visual loss, and normal ophthalmoscopic examination results. The retrobulbar type of neuritis is less common than the neuroretinitis type.^{5,7}

A review of the English-language literature revealed 5 well-described pediatric case reports of optic neuritis complicating mumps^{5,6,9,11,12} (**Table**). Ocular manifestations were limited to papillitis in 1 case, neuroretinitis in 3 cases, and retrobulbar neuritis in 2 cases. There were 3 boys and 3 girls, with ages ranging from 7 to 15 years. Five cases had severe visual loss, including sudden onset blindness in 2 cases. One patient was asymptomatic. The onset of ocular symptoms began between 2 and 5 weeks after parotitis. Funduscopic examination results were unremarkable for 2 cases, and bilateral involvement occurred in 3 patients. Accompanying neurologic manifestation included meningitis in 3 cases and encephalitis in 2 cases. One patient had no visual symptoms, and an MRI study with short inversion time recovery sequences in conjunction with visual evoked potentials proved useful in the detection of asymptomatic optic neuritis.¹² Four cases had near complete recovery of visual function, indicating a temporary block in conduction of action potentials and not structural disruption of optic nerve axons. Pallor of the optic discs was a common sequela. One case developed secondary optic atrophy.⁵ Our patient was nonvaccinated, had history of exposure to mumps, and subsequently developed clinical findings consistent with mumps, including acute parotitis followed by aseptic meningitis. Three weeks after the onset of parotitis, she developed bilateral neuroretinitis with serologic evidence of mumps infection.

Meningoencephalitis followed by parainfectious optic neuritis has been reported in several other viral infections, including measles, rubella, varicella, Epstein-Barr virus, and other pathogens, such as *Coxiella burnetii*, toxoplasmosis, cat-scratch disease, and Lyme disease.^{17,18} The pathogenesis of parainfectious optic neuritis is not well understood, but an immunologic basis has been postulated.¹⁷ The delayed onset of optic neuritis in mumps infection, bilateral involvement of the optic nerves, and profound loss of visual function with often complete recovery support a systemic auto-

immune demyelination process rather than random viral invasion of each optic nerve.¹⁷ Additionally, the temporal framework and immunopathologic features of postinfectious encephalitis closely resemble those of experimental models of virus-induced demyelination of the central nervous system.¹⁹

The role of corticosteroids is controversial. A randomized controlled trial of intravenous methylprednisolone followed by oral prednisone in acute optic neuritis in the adult population showed quicker recovery from visual loss and better vision at 6 months.²⁰ However, it is unclear whether corticosteroids affect the natural history of the disease in children, and this merits further investigation.²¹ The experience with corticosteroids in mumps is anecdotal and limited to a few case reports.^{9,11,12}

Although regarded as a benign infectious disease, physicians should be aware of optic neuritis as a potentially serious ocular complication of mumps infection. In resource-poor countries, mumps immunization should be included in the routine vaccination program.

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