MR Findings in Listerial Rhombencephalitis

Gülay Alper, Laurie Knepper, and Emanuel Kanal

Summary: We describe a case of listerial rhombencephalitis in a previously healthy 40-year-old man. The diagnosis was based on the clinical findings, results of cerebrospinal fluid analysis, blood culture, and MR imaging findings. The treatment was started before culture results were available, and the patient had a full clinical recovery.

Index terms: Brain, infection; Encephalitis; Rhombencephalon

Listeria monocytogenes is an anaerobic, gram-positive bacillus. Infection with this organism occurs primarily in immunocompromised hosts and is uncommon in the general population. We present a case of listerial rhombencephalitis in an otherwise healthy person in whom diagnosis was based on the clinical findings, blood culture, cerebrospinal fluid (CSF) results, and magnetic resonance (MR) findings, all of which were obtained early in the course of the disease. In this case, treatment was started before culture confirmation on the basis of clinical and MR findings, which were consistent with listerial infection.

Case Report

A 40-year-old right-handed man came to the local emergency department with rhinitis and fever and was started on an antibiotic. Blurred vision, dysphagia, drooling, hoarseness, hiccuping, and headache developed, and he was admitted to a psychiatric facility because his symptoms were presumed to be psychogenic. His condition worsened, and ceftriaxone therapy was begun after lumbar puncture and MR imaging produced abnormal findings. The patient was then transferred to the university hospital.

At the time of admission, the patient was alert with excessive salivation and frequent hiccuping. On examination, he had right-sided upbeating rotational nystagmus, decreased right-sided corneal reflex, a right-sided central facial paresis, bilateral absent gag reflex, and marked bilateral cerebellar ataxia, more prominent on the right side. He was subsequently intubated because of bilateral vocal cord paralysis and airway obstruction. CSF findings included leukocyte count of 330 mm^3 (47% neutrophils, 52% lymphocytes, 1% monocytes), protein level of 114 mg/dL, and normal glucose level. CSF cultures were negative for *Listeria* as well as other bacterial, viral, and fungal microorganisms. The spinal fluid VDRL and the results of tests for Lyme disease antibody, toxoplasmosis titers, and cryptococcal antigen were negative. Results of a serum human immunodeficiency virus test were also negative.

Findings at cranial computed tomography (CT) were normal. Initial MR imaging showed abnormal lesions of the brain stem, cerebellar peduncles, and cerebellum (Fig 1A). There was abnormal widening of the medulla associated with patchy regions of enhancement after intravenous administration of MR contrast agent. Similar areas of enhancement were also seen in the right cerebellar hemisphere (Fig 1B and C).

Listerial infection was suspected and trimethoprim-sulfamethoxazole (TMP-SMX) was begun after blood cultures were obtained, because the patient had an allergy to penicillin. Blood cultures grew *L monocytogenes*. The patient continued to improve clinically. Within 2 weeks of beginning TMP-SMX he was able to walk with a mildly ataxic gait, able to swallow, and had normal functioning of vocal cords by laryngoscopy. Repeat CSF examinations showed remarkable improvement.

A follow-up MR examination was performed after 14 days of therapy. Concomitant with considerable clinical improvement at this time, this examination revealed a noticeable decrease in the extent of the lesions. These lesions were somewhat smaller, more consolidated, and had considerably less contrast enhancement (Fig 1D).

Three weeks after beginning TMP-SMX, the patient was discharged to home with a mild residual right-sided dysmetria. Findings on examination were completely normal when he was seen as an outpatient 1 month after discharge.

Discussion

L monocytogenes can cause meningitis, meningoencephalitis, or abscess formation in the

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From the Department of Pediatric Neurology, Children's Hospital of Pittsburgh (Pa) (G.A.), and the Departments of Neurology (L.K.) and Neuroradiology (E.K.), University of Pittsburgh (Pa) Medical Center.

Address reprint requests to Gülay Alper, MD, Department of Pediatric Neurology, Children's Hospital of Pittsburgh, 3705 Fifth Ave at DeSoto St, Pittsburgh, PA 15213.

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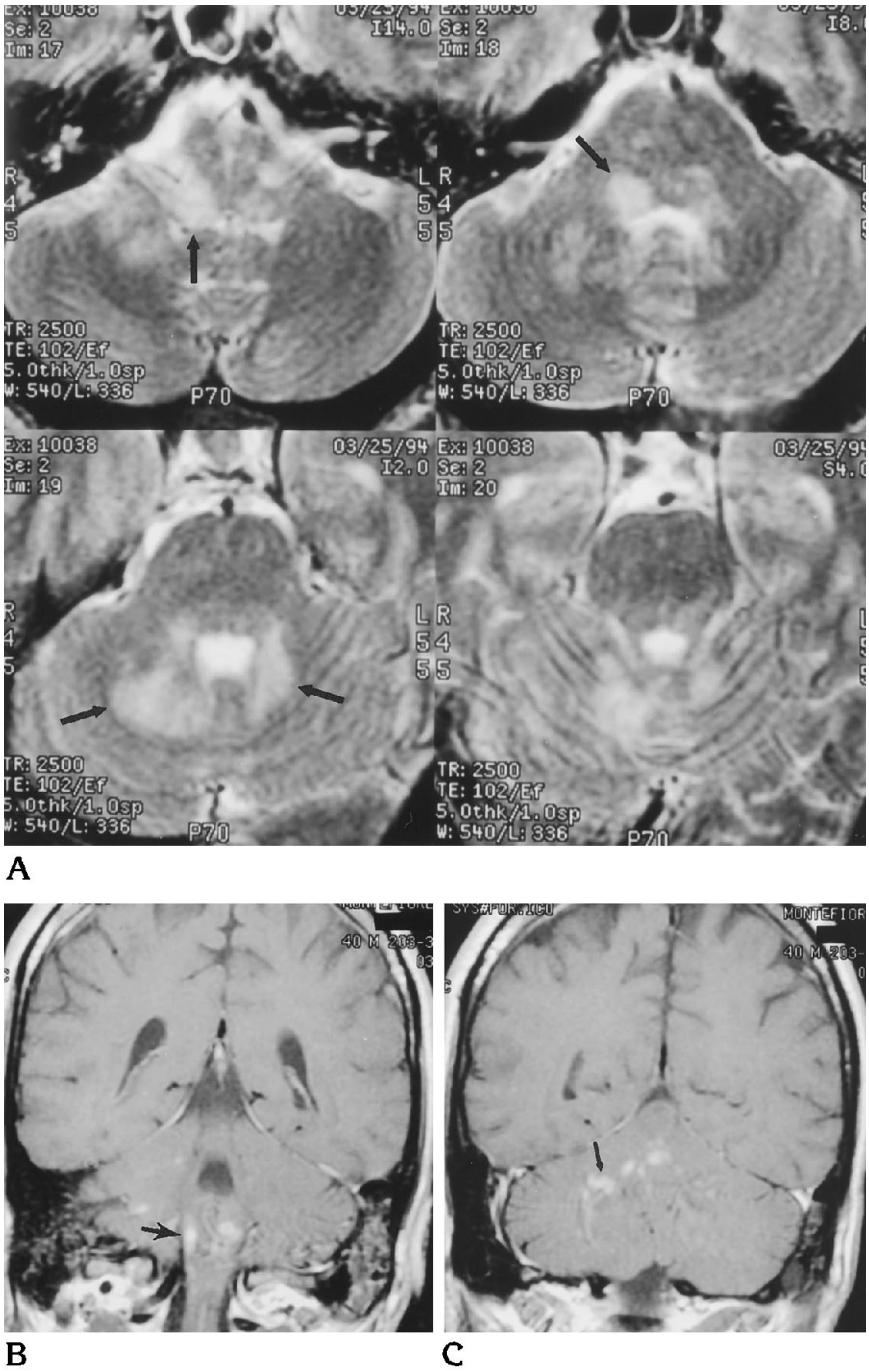


Fig 1. *A*, Axial T2-weighted MR images (2500/102/1 [repetition time/echo time/excitations]) show bilateral patchy regions of abnormal signal in the medulla, posteroinferior pons, cerebellar peduncles, and cerebellar hemispheres (*arrows*), with the right side more involved than the left.

B and C, Coronal T1-weighted contrast-enhanced MR images (500/19/2) show patchy regions of enhancement in the medulla (*arrow* in B) with extension interiorly to the uppermost aspect of the cervical spine and in the cerebellum (*arrow* in C).

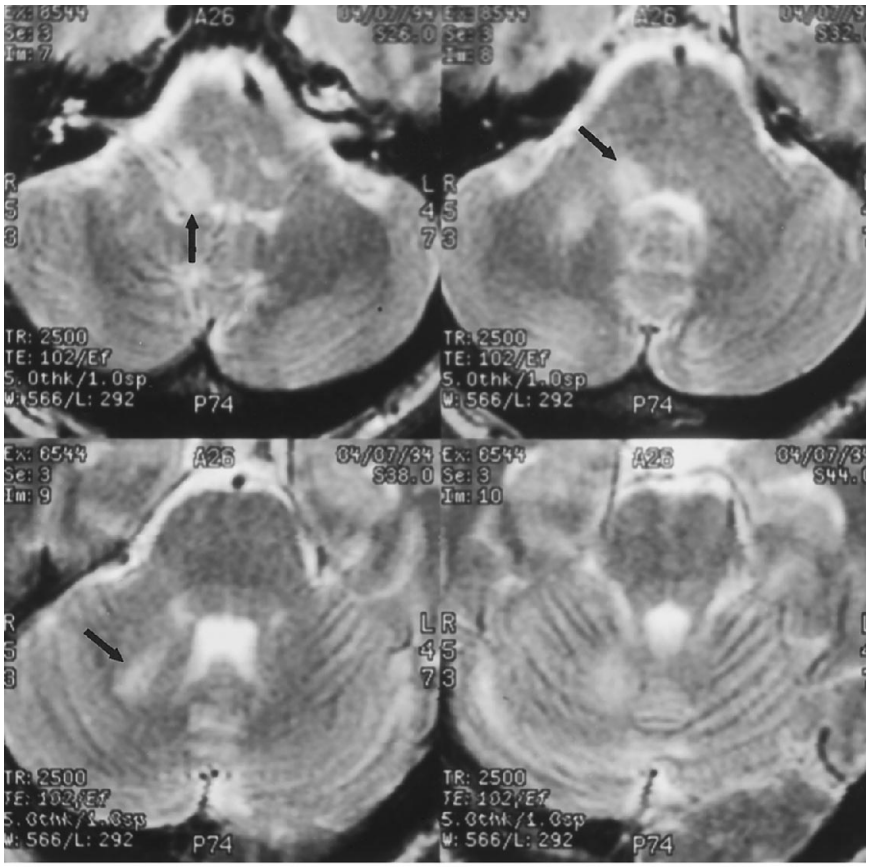
Figure continues.

central nervous system. Rhombencephalitis is a particular form of listerial encephalitis that affects primarily the brain stem and cerebellum (rhombencephalon). Listerial infection is often misdiagnosed initially because the prodromal symptoms are nonspecific and meningeal signs are uncommon. Brain stem encephalitis should be considered upon progression of such symptoms as nystagmus, gaze palsy, facial numbness, vertigo, dysphagia, persistent hiccuping, and respiratory failure, suggesting multiple cranial nerve involvement. Early diagnosis of brain stem involvement is essential, as respiratory failure can lead to death if untreated.

The CSF in listerial infection typically reveals an increased leukocyte count, usually with predominance of polymorphonuclear cells, increased protein, and normal glucose levels. Because *L monocytogenes* is sensitive only to certain antibiotics, it is important to establish an

Fig 1, *continued.*

D, Axial T2-weighted MR images (2500/102) show patchy regions of increased signal in the middle cerebellar peduncle and cerebellum restricted to the right side (*arrows*) 2 weeks after treatment with trimethoprim-sulfamethoxazole.



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early microbiological diagnosis. *L monocytogenes* is difficult to isolate from the CSF but is often readily cultured from blood.

In addition to the microbiological diagnosis, MR imaging is extremely important in demonstrating the predilection of listerial infection for the brain stem and cerebellum. These regions can also be involved in viral encephalitis, lymphoma, vasculitic diseases (systemic lupus erythematosus, Behçet disease), neurosarcoidosis, tuberculosis, multiple sclerosis, and acute disseminated encephalomyelitis. Viral encephalitis causes primarily a lymphocytic predominance in the CSF with negative blood cultures. Tuberculous meningitis can be distinguished by the fact that it causes primarily leptomeningeal involvement with low CSF glucose levels. Vasculitic and granulomatous diseases have associated systemic findings. In acute disseminated encephalomyelitis, lesions are mostly widespread, involving the subcortical hemispheric regions not restricted to the brain stem and cerebellum. Although similar MR findings may be seen in multiple sclerosis, the clinical and CSF findings of this patient would not be typical for this diagnosis.

Neuroimaging before the advent of MR imaging was not generally helpful in supporting the diagnosis of listerial meningitis, because visualization of the brain stem and cerebellum on CT scans is limited by bony artifacts. Most of the reports of listerial rhombencephalitis have described normal findings on CT scans (1–4). Some abnormal CT findings in documented listerial infection have included widening of the brain stem, hydrocephalus, brain stem or cerebellar abscess, and vermian hemorrhage (4, 5). There are only a few cases of listerial infection in which central nervous system lesions were documented by MR imaging (6–9). King and Jeffree reported abnormal MR findings of the cervical spine in a patient with listerial infection (10). Fatal cases of listerial rhombencephalitis have been reported with neuropathologic findings of numerous intracellular and extracellular bacilli in the areas of the brain stem and cerebellar white matter (1, 9, 11).

Our patient had normal findings on a CT scan, but MR images obtained 1 day later showed extensive pontomedullary and cerebellar lesions. Antibiotic therapy covering *Listeria* organisms was started immediately after review of the MR findings and spinal fluid analysis but before culture confirmation, and the patient had a full recovery. The overall mortality rate for listerial meningitis exceeds 50% (9). Mortality is higher in patients who are immunosuppressed or in whom the treatment of listerial infection is delayed. Although *L monocytogenes* is an uncommon pathogen in previously healthy persons, it can cause fatal CNS infection in people with no underlying predisposing factors.

One should always suspect listerial rhombencephalitis in the presence of an acute onset of progressive cranial nerve dysfunction and ataxia, CSF leukocytosis with polymorphonuclear cell predominance with normal glucose levels, and MR findings of brain stem (primarily pontomedullary) and cerebellar lesions. MR imaging performed early in the course of the disease would strongly support listerial rhombencephalitis in conjunction with spinal fluid results and is essential in choosing the appropriate antibiotic treatment while awaiting culture confirmation. Early treatment can decrease the morbidity and mortality of this rare pathogen.

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