Bulbar Poliomyelitis: MR Findings with Pathologic Correlation

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Summary: We report a case of bulbar poliomyelitis in which postmortem pathologic tissue sections reflect the changes seen on MR scans of the midbrain and medulla oblongata.

Index terms: Nervous system, infection; Viruses; Neuropathology

Case Report

This 17-month-old female with severe combined primary immunodeficiency was admitted for gastrostomy tube placement. Upon admission she had a fever of 38.6°F mononcytosis, and an abnormal interstitial pattern on chest radiograph. Nine days later, the patient developed fever spikes, torticollis, and generalized decreased muscle tone. A computed tomography (CT) scan was performed that was suspicious for a pontine mass. A lumbar puncture was performed that revealed 53 white cells (92% polymorphonuclear), normal glucose, and slightly elevated protein. The patient was treated with antibiotics for possible bacterial meningitis. One day later, a magnetic resonance scan showed a normal pons and increased signal intensity in the midbrain and medulla oblongata on T2-weighted images. The patient became ventilator dependent. Enterovirus was cultured from the stool and the spinal fluid obtained on the day prior to the magnetic resonance scan. The enterovirus was identified as poliovirus type II.

With supportive therapy, the patient's neurologic condition improved. Facial muscle movements and spontaneous respirations returned. The patient subsequently became septic, developed respiratory distress, and suffered a cardiac arrest which was refractory to treatment. The patient died 5 weeks after admission.

At autopsy, examination of the central nervous system (CNS) revealed numerous asymmetric discrete foci of cystic necrosis in the brain stem and anterior horns of the spinal cord (Figs. 1 and 2). Microscopic examination revealed neuronal loss with extensive macrophage infiltration (Fig. 2). The pathologic findings in combination with the isolation of poliovirus were felt to be diagnostic of poliomyelitis. The patient had received two doses of oral polio vaccine; the last dose was 8 months prior to the onset of this illness. Virus typing by the Centers for Disease Control revealed the virus to be a vaccine strain.

Discussion

The poliovirus is an enterovirus of the family of Picornaviruses. It has a single strand of RNA that replicates in the host cell cytoplasm and is released by cell lysis. Most cases of polio do not involve the CNS with only 0.1%–1% progressing to paralysis (1).

Poliomyelitis was sporadic prior to the late 19th century when epidemics swept Europe and North America. In 1952, there were 20,000 reported cases in the United States. From 1980–1984, there were an average of 10 cases per year, all of which were vaccine related (2). There is a greater predisposition for infection in adults than in children. Exercise, trauma, and prior injections all seem to increase the likelihood of limb paralysis. The involvement of the spinal cord motor neurons lead to the name "poliomyelitis," from the Greek for "gray marrow."

The mechanism of CNS infection by poliovirus is thought to be either by neural spread or direct infection during the viremic phase (1). The pathway of neural spread from the gut is probably via the vagus nerve, allowing infection of the nucleus solitarius. However, there may be direct infection of the area postrema of the brain stem during the viremic phase of the illness, because it has no blood brain barrier. When poliovirus infects the

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Fig. 1. A, T2-weighted axial image (2200/90/1) demonstrates abnormal increased signal intensity in the cerebral peduncles (arrow).

B, Gadolinium-DTPA-enhanced T1-weighted image (600/15/1) of the same section reveals (curved arrow) decreased signal intensity without abnormal enhancement.

C, Pathologic section shows areas of necrosis (arrows) in the region of the substantia nigra corresponding to the areas of abnormal signal intensity in A.

Fig. 2. A, T2-weighted axial image (2200/90/1) demonstrates abnormal increased signal intensity in the medulla oblongata (arrow).

B, Pathologic section demonstrates necrosis (arrows) of the reticular formation corresponding to the area of abnormal signal intensity in A.

C, Photomicrograph showing cystic necrosis in the reticular formation surrounded by an area of macrophage infiltration.
brain, the brain stem is affected first and is virtually always associated with meningitis.

Paralytic poliomyelitis may present with spinal cord symptoms, brain stem symptoms (bulbar) or both. Only 10%-15% of paralytic polio is bulbar, with the reticular formation most severely affected. Cranial nerve involvement is common, with cranial nerves VII, IX, and X most often involved. Patients may experience difficulty breathing or swallowing. Occasionally, there is autonomic dysfunction as well with hypo- or hypertension and cardiac arrhythmias.

Pathologically, there is early chromatolysis that may go on to cell membrane lysis. Mononuclear cell perivascular cuffing and neuronophagia occur. In severe cases, there may be cavitation of the involved brain tissue (3).

From 1973–1984 there were 14 cases of paralytic polio reported in immunodeficient patients (2). In such patients, there may be persistent CNS infection with enterovirus, usually vaccine polio, but sometimes echovirus. These patients are usually hypo- or agammaglobulinemic, with normal cellular immunity. However, this may occur in patients with combined immunodeficiencies, as in this case. Symptoms may occur months after the vaccination and usually progress to a fatal conclusion. The findings in this case were of abnormal high signal in the midbrain and medulla without enhancement or mass effect. There is one other report of similar MR findings in a patient with herpes brain stem encephalitis (4). In that case the involvement was limited to the pons. This allows at least one differential feature from polio that does not involve the pons. Other diagnostic considerations would include demyelination. Abnormal myelination was a consideration in this case since the anterior horn of the internal capsule, usually myelinated at eight months of age, was not myelinated in this 17-month-old child. However, the febrile presentation and positive spinal fluid viral cultures suggested an infectious process.

This case demonstrates the pathologic basis of magnetic resonance (MR) imaging. The MR findings are not specific, and both disorders of myelin and encephalitis should be considered when abnormal signal is seen without associated mass effect. However, in the clinical setting of an immunocompromised child with brain stem involvement that spares the pons and flaccid paralysis, the diagnosis of bulbar poliomyelitis should be entertained.

References