Rhombencephalitis due to *Listeria monocytogenes*

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**Abstract**

Brain stem encephalitis is an unusual form of CNS listeriosis that is associated with a high mortality. This is a case of a 46-year-old male with a history of dermatomyositis on methotrexate who presented with fever, headache, asymmetrical cranial nerve palsy and right hemiparesis. MRI showed a ring-enhancing lesions in medulla oblongata. CSF cultures grew *Listeria monocytogenes*. Despite treatment with ampicillin and gentamicin, the patient developed three rare manifestations of neurolisteriosis namely rhombencephalitis, hydrocephalus and brainstem hemorrhage and did not survive. Early detection and treatment is vital in preventing adverse outcomes.

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**Case report**

**Background**

*Listeria monocytogenes* is a food borne pathogen that can cause a spectrum of CNS infections including meningitis, encephalitis and brain abscesses. While immunocompromised patients, pregnant women, neonates and elderly patients are at the highest risk of developing listeria meningitis; infections can also be seen in immunocompetent patients.

**Case presentation**

A 46-year-old male presented to the emergency room with subjective fevers, severe global headache, right-sided upper and lower extremity weakness, blurred vision, difficulty swallowing and unsteady gait. Fever and headache began 1-week prior. He initially went to an urgent care clinic, was diagnosed with sinusitis and given a prescription for oral azithromycin. A worsening headache and neurological symptoms prompted a visit to the emergency room where he developed respiratory failure and was emergently intubated.

His past medical history was significant for dermatomyositis for which he received weekly methotrexate. His most recent dose of methotrexate was 3 weeks prior to presentation. He was employed as a coach for a local high school football team and had no history of recent travel.

Pertinent findings on physical exam included a temperature of 100.8°F, heart rate of 115, pinpoint pupils, deviation of left eye upwards and laterally, deviation of right eye downwards, left facial palsy and right hemiparesis. Speech and gait was not tested. Laboratory findings included leukocytosis of 15,000 with 88% neutrophils, mild elevation of alanine aminotransferase (ALT) at 82 U/L and aspartate aminotransferase (AST) at 87 U/L. HIV test was negative. Blood cultures remained negative for bacterial growth. Magnetic resonance imaging (MRI) of the brain showed a ring enhancing lesions in the lateral medulla (Fig. 1). Lumbar puncture revealed an opening pressure of 20 cm of water. CSF analysis showed glucose of 44 mg/dl, protein of 115 mg/dl, 67/mm³ white blood cells (48% polymorphonuclear cells) and 13/mm³ red blood cells. The patient was empirically treated for meningitis with intravenous (IV) vancomycin, (IV) ceftriaxone, (IV) ampicillin and (IV) acyclovir. CSF cultures grew *Listeria monocytogenes*. Antibiotic therapy was modified to (IV) ampicillin and (IV) gentamicin. Despite appropriate antibiotic therapy, the patient’s condition continued to worsen. Repeat MRI of the brain on fourth day of admission showed progression to multiple ring enhancing lesions in the brain stem (Fig. 2). The patient became unresponsive on eighth day of admission. Computed tomography (CT) of the head showed extensive brainstem hemorrhage with resultant hydrocephalus (Fig. 3). He underwent ventriculostomy without significant improvement. He was transitioned to comfort care measures and died on the tenth day, following terminal extubation.

**Discussion**

While extremes of age, pregnancy, immunosuppression and malignancies are thought to major risk factors for developing CNS listeriosis, rhombencephalitis is known to primarily affect healthy adults [1,2]. The term rhombencephalitis refers to the initial
rhombencephalon or hindbrain. Rhombencephalitis or brainstem encephalitis is an unusual form of CNS listeriosis first described in 1957 [3]. It has a characteristic biphasic course consisting of a prodrome of headache, nausea, vomiting and fever lasting for several days followed by progressive asymmetrical cranial-nerve palsies [4]. MRI is crucial for early diagnosis and mortality remains high at 51% [5]. Hemorrhage and hydrocephalus are also rare complications of CNS listeriosis [6].

Treatment of choice for CNS listeriosis is ampicillin or penicillin G with the addition of an aminoglycoside such as gentamicin. For patients with penicillin allergies, meropenem or trimethoprim-sulfamethoxazole may be used [7].

This patient developed all three rare manifestations of neuro-listeriosis namely rhombencephalitis, hydrocephalus and brainstem hemorrhage and did not survive despite initiation of antimicrobial therapy within hours of hospitalization. While this patient was on weekly methotrexate, the perceived risk of serious infections related to methotrexate therapy is seen to be lower than previously thought to be [8–10].

Declaration of Competing Interest

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Consent

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Author contribution

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CRedit authorship contribution statement

Neha Paranjape: Conceptualization, Data curation, Investigation, Visualization, Writing - original draft, Writing - review & editing.

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