**Listeria rhomboencephalitis**

Mohamed Ramadan, Nicole M McGrath

A previously fit 78-year-old electrical linesman described a 1-week history of gradual non-specific deterioration in his general health without headache, and progressive generalised weakness. He became unsteady 3 days before presentation. The next day he developed difficulty swallowing, poor appetite and dry retching, then left-sided facial weakness on the day of admission. He had known chronic atrial fibrillation on warfarin, treated hypertension and was a current smoker.

At presentation he was afebrile with no meningism. Neurological examination revealed rotatory nystagmus in the primary gaze, left lower motor neurone facial weakness, dysarthria, right palatal weakness with profound dysphagia, right tongue deviation and truncal ataxia.

His peripheral white cell count was $14.2 \times 10^6 \text{mmol/litre}$ with a neutrophil predominance; INR 3.4. A CT head scan showed moderate vascular calcification and mild cerebral atrophy only.

At admission, the patient was thought to have had a vertebrobasilar territory stroke and was given aspirin. That night, the patient’s breathing deteriorated acutely, progressing rapidly to respiratory arrest. The patient was resuscitated, intubated and mechanically ventilated, then transferred to the Intensive Care Unit. The following morning he was alert and able to communicate fully via hand signals and writing. Limb strength was normal.

Two days after admission he developed a temperature spike of 38.4°C. He was commenced on cefuroxime and blood cultures were taken that subsequently grew *Listeria monocytogenes*, identified initially on Gram stain and confirmed by culture of typical colonies, and use of Oxford media and API Coryne. An MRI brain scan revealed brain stem and upper spinal cord small ring enhancing lesions (Figures 1–4).

The patient was changed to intravenous amoxycillin and gentamicin therapy 3 days after admission, when listeriosis was first identified. However there was no improvement in his respiratory function. Several attempts at extubation over the next few days were unsuccessful.

After discussion with the patient and the family, while the patient was still conscious, a decision of discontinuing treatment and extubation was made. The patient was declared dead 9 days post admission. A subsequent public health investigation concluded that the source of *Listeria* was “unknown, most likely an unidentified food”.  

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Figure 1. Axial MRI, T1 window showing two ring enhancing lesions in the midbrain

Figure 2. Sagittal MRI T1 Window showing a column of ring-enhancing lesions extending up the brain stem
Figure 3. T2 Axial MRI showing homogenous opacity in brain stem and left cerebellar hemisphere

Figure 4. Coronal T1 MRI film showing multi-enhancement lesions extending up the brain stem mainly on the left
**Discussion**

Listeriosis is very uncommon, with a notification rate of 0.6 cases per 100,000 in New Zealand.\(^1\) It can present in sporadic or foodborne outbreaks. Unlike most other foodborne organisms, *Listeria* can multiply in refrigerated food that is contaminated.

It mainly affects pregnant women, neonates, immunocompromised personnel and the elderly. Central nervous system (CNS) involvement occurs in 47% of cases\(^2\) and there are three distinct forms of CNS infection.\(^3\)

Meningoencephalitis, the most common, and cerebritis that can extend to cerebral abscess, are usually found in immunocompromised patients. Rhombencephalitis (brainstem infection) accounts for 9% of CNS listeriosis cases and can affect healthy elderly.\(^3\)

Patients with *Listeria* rhomboencephalitis typically present with a prodroma of headache, nausea, fever and malaise for several days followed by progressive asymmetrical cranial-nerve lesions, cerebellar signs, hemiparesis or hypoaesthesia, and impairment of consciousness. 41% develop respiratory failure.\(^4\)

At presentation, 85% of patients have fever and 90% have cranial nerve deficits.\(^2\) There is often only a mild CSF lymphocytosis, and in 22% there is no CSF pleocytosis at all. *Listeria* is more commonly isolated in blood culture (61% of cases\(^4\)) than cerebrospinal fluid (CSF) (41%\(^4\)). MRI with contrast is much more sensitive and specific than CT brain and is the preferred examination in patients with brainstem signs.\(^3\)

The treatment of choice is penicillin with or without an aminoglycoside for at least 2 weeks or until blood cultures and MRI are negative.\(^3\) Cotrimoxazole is an effective alternative, or meropenem if there is no history of an IgE-type immediate hypersensitivity, in penicillin allergic patients. Overall mortality is 51% and 61% of survivors have permanent neurological deficits.\(^4\)

This case illustrates that, in the absence of fever, headache or meningism at presentation, CNS infection, particularly listeriosis, can be overlooked. In fact, the striking absence or only subtle symptoms and signs of meningitis relative to brainstem symptoms and signs in *Listeria* rhomboencephalitis previously inspired the term “ameningitic encephalitis”.\(^2\) Focal neurological signs in older immunocompetent patients are often attributed to stroke. However, prodromal illness is not typical of stroke and an alternative aetiology, particularly infection, needs to be considered, even in the absence of fever at presentation.\(^2\)

An urgent MRI brain scan is indicated for patients with progressive disabling brainstem signs.\(^4\) As in this case, the MRI findings may raise the possibility of CNS infection and, in particular, *Listeria* rhomboencephalitis. Blood cultures are more likely to lead to a diagnosis than CSF culture but ideally and if possible, both should be performed.\(^4\)

Empiric antibiotic treatment that includes IV amoxycillin to cover the possibility of *Listeria* rhomboencephalitis is advised in the elderly, pregnant women, neonates and immunocompromised patients who present with the febrile-brainstem syndrome.\(^2\) However, prodromal fever is not universal and, as in our case, vague systemic
symptoms preceding progressive brainstem symptoms and signs should also alert the clinician to the possibility of *Listeria* rhomboencephalitis.

**Author information:** Mohamed Ramadan, Medical Registrar; Nicole McGrath, Physician; Whangarei Hospital, Whangarei, Northland

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**Correspondence:** Nicole M McGrath, Department of Medicine, Whangarei Hospital, Private Bag 9742, Whangarei, New Zealand. Fax: +64 (0)9 4304117; email: Nicole.mcgrath@northlanddhb.org.nz

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