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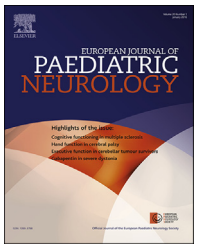

Case Study

An unusual presentation of paediatric *Listeria* meningitis with selective spinal grey matter involvement and acute demyelinating polyneuropathy

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ABSTRACT

Background: Paediatric *Listeria* meningitis is rare, especially in immuno-competent children, but associated with significant mortality and morbidity and frequent complications.
Methods: We report an unusual case of *Listeria* meningitis in a previously healthy 35 month-old girl with selective spinal grey matter involvement and demyelination in neurophysiological studies. Despite adequate antibiotic treatment, the case was initially complicated by ventriculitis, hydrocephalus and tonsillar herniation through the foramen magnum, requiring external ventricular drainage and subsequent ventriculoperitoneal shunt insertion. Paucity of movements, hypotonia, areflexia and bladder dysfunction then became evident.
Results: Electromyogram and nerve conduction studies showed acute inflammatory demyelinating polyneuropathy and the patient received intravenous immunoglobulin followed by corticosteroids. MRI scans with contrast revealed extensive whole cord selective grey matter signal changes. She required extensive neurorehabilitation, making gradual (but incomplete) recovery.
Conclusion: Spinal cord involvement is rare in neuro-listeriosis and there no previous paediatric reports of *Listeria*-related myelitis or demyelinating polyneuropathy. The mechanism behind these presentations is unclear but an auto-immune response to the infection might be considered.

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1. Introduction

Listeria monocytogenes causes infections mainly at the extreme ends of life (newborns and the elderly), in the immunocompromised and in pregnant women.[1] *Listeria* meningitis is rare in the paediatric population and exceedingly so after 3 months of life,[2] especially in the absence of immunodeficiencies. However, some cases of neurolisteriosis in immunocompetent children have been reported.[3] Although there is scarcity of paediatric reported cases, neurolisteriosis in children can be associated with severe morbidity and mortality.[4] Paediatric *Listeria* meningitis is often complicated by cerebral oedema and hydrocephalus.[4] Other central nervous system (CNS) associations of neurolisteriosis are ventriculitis, brain abscess formation, rhomboencephalitis, spinal cord abscess formation, spinal arachnoiditis and syringomyelia.[5–9] *Listeria*-related myelitis is extremely rare[10] but, to date, there is no relevant available literature in paediatrics. In this study, we describe the first reported paediatric case of neurolisteriosis with selective grey matter involvement in the spinal cord and features of peripheral demyelination on neurophysiological studies.

2. Case study

An afro-caribbean 3 year old girl born to non-consanguineous parents, with unremarkable past medical history and neurodevelopmental trajectory, presented acutely with pyrexia, back-arching and lethargy preceded by 3 days' history of fever and coryza. At presentation, irritability and neck stiffness were present, with no focal neurological signs, elicitable (not brisk) reflexes and equal and reactive pupils. Systemic examination was otherwise unremarkable. Empirical intravenous (IV) treatment for meningoencephalitis (ceftriaxone and acyclovir) was commenced. Initial cerebrospinal fluid (CSF) sampling 24 h later (day 2) showed pleocytosis (white cell count, WBC, 1350 $\times$ 10^6/L, neutrophils 5%, lymphocytes 90%), high protein (2.8 g/L, range 0.15–0.45 g/L) and low glucose (0.4 mmol/L, blood glucose 8.4 mmol/L). Computed tomography (CT) brain imaging on day 2 was unremarkable. Despite antibiotics, irritability and lethargy persisted. CSF cultures confirmed *L. monocytogenes* (serotype 1/2a), therefore antibiotics were changed to amoxicillin and gentamicin and, subsequently, to ampicillin and amikacin. On day 5, diminishing levels of consciousness, signs of raised intracranial pressure and equal but sluggish pupils were noted. Hence, intubation and mechanical ventilation was required. A repeat CT brain revealed hydrocephalus with tonsillar herniation through the foramen magnum; a right trans-frontal External Ventricular Drain (EVD) was inserted. Additionally, marked hypotonia, areflexia, paucity of movements and absence of response to stimuli were noted; these persisted long-term, even without concurrent sedative or muscle relaxant administration. Therefore, an electroencephalogram was performed on day 6, demonstrating generalised slowing and no alteration after tactile and auditory stimulation. Somatosensory Evoked Potentials (following bilateral median nerve stimulation) were unremarkable, indicating intact somatosensory pathways to the brain above the foramen magnum level. Flash Electroretinogram (ERG)/Visual Evoked Potentials (VEP) were also normal. Head magnetic resonance imaging (MRI) on day 7 showed marked ependymal and leptomeningeal enhancement, consistent with meningitis and ventriculitis. Non-enhancing parenchymal signal abnormalities were also shown, extending to the cervical cord. Electromyography/nerve conduction studies on day 8 showed widespread motor nerve compound muscle action potential reduction and absent F-waves but no sensory nerve abnormalities or axonal degeneration. Overall, findings were consistent with acute inflammatory demyelinating polyneuropathy (AIDP). Therefore, IV immunoglobulin (2 g/kg) was administered on day 11, followed by IV Methylprednisolone (30 mg/kg/day for 5 days); these resulted in no clinical improvement, with persistence of flaccidity and areflexia. Oral prednisolone (40 mg/day, 2.5 mg/kg/day) was then given for four weeks, subsequently weaned off gradually over a 4-week period. EVD was internalised on day 21 and a left trans-parietal ventriculoperitoneal (VP) shunt inserted. Repeat MRI head and whole spine with contrast (days 22 and 40) showed reduction in the size of the third and lateral ventricles, no leptomeningeal or ependymal enhancement, but extensive (and persisting) whole cord grey matter signal changes, associated with loss of volume (Fig. 1). Amikacin and ampicillin were discontinued after 2 (day 20) and 3 weeks in total (day 27), respectively. CSF WBC had previously

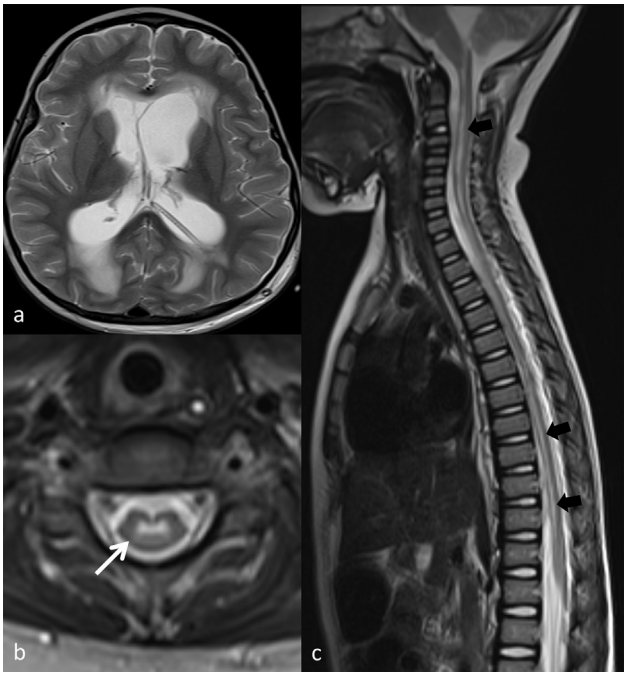

Fig. 1 – Magnetic resonance imaging findings. 1a: Axial T2-weighted brain imaging depicting hydrocephalus and a left trans-parietal ventriculoperitoneal shunt. 1b: Axial T2-weighted spinal cord imaging. An 'M-shaped' grey matter abnormal signal enhancement is shown (white arrow), suggestive of selective grey matter involvement. 1c: Sagittal T2-weighted spinal cord imaging, demonstrating the extensive, selective (non-enhancing) whole cord grey matter signal changes (black arrows).