A Case of Listeria Rhomboencephalitis with Misleading Imaging Presentation and Laboratory Findings

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Adrien De Minteguiaga: Revision of literature and manuscript writing
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Valeria Onofrj: Revision of literature and manuscript writing

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The authors of the present manuscript have nothing to disclose.

CONSENT
Did the author obtain written informed consent from the patient for submission of this manuscript for publication? yes

HUMAN AND ANIMAL RIGHTS
The authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

ABSTRACT

Listeria rhomboencephalitis is an uncommon, sporadically occurring form of listerial infection that progresses rapidly and is invariably fatal unless treated early . Cases are often undetected. Listeriarhomboencephalitis is characterised by a prodromal febrile illness followed by the abrupt onset of cranial nerve deficits associated with cerebellar ataxia and long-tract motor and sensory features. It can be a diagnostic challenge due to the multitude of possible differential diagnoses and also because the presentation, imaging findings, laboratory findings and course of the disease are also variable. Here we report a case with several misleading features.

CASE REPORT

BACKGROUND

This case report describes a rare case of listeria rhboencephalitis with an unusual clinical and imaging presentation highlighting the complexity of the diagnosis of listeria rhboencephalitis whose importance remains crucial for the treatment, which if timely, can be life-saving in most cases. Listeria rhomboencephalitis is a rare diagnosis, but its facets need to be known. Our case report adds to the current literature an instructive case of an important diagnosis with discussion on differential diagnosis.

CASE REPORT

Ethical statement

This case was performed according to the Declaration of Helsinki (1997) and subsequent revisions.

Case presentation

We report a case of an 81-year-old female presenting to the emergency unit with recent onset of cerebellar and right facial syndrome. Past medical history included a bulbo-medullary stroke several months before (Figure 1).

Imaging findings

Initial emergency assessment was performed with a non-contrast CT, which proved negative (not shown). An MRI was performed the following day (Figure 2).

Compared to a previous MRI performed at the time of the bulbo-medullary stroke, there was an increase in periventricular signal abnormalities, the appearance of signal abnormalities with diffusion restriction in the right paramedian bulbar region with foci of pseudonodular enhancement. The CSF sample was acellular with normal chemistry and microscopy.

The concomitant presence of multiple T2/FLAIR hypersignal foci in the supratentorial, mostly periventricular white matter and in the brainstem led us to hypothesize diffusion in space with dissemination of an inflammatory/demielinating process.

Due to the progression of symptoms, we realized a spinal cord MRI four days later, which showed an increase in size of the bulbo-spinal lesion (Figure 3).

Two days later, the patient had a rapid deterioration of symptoms, for which reason she was admitted to the intensive care unit, and we decided to carry out a new cerebral MRI (Figure 4) which showed a marked increase in the extent of lesions now involving the right cerebellum and middle cerebellar peduncle, the brainstem, the thalami and the right basal ganglia, characterized by predominantly confluent T2/FLAIR signal hyperintensities marked by multiple foci of diffusion restriction and ring-like enhancement, with a tendency to coalescence. These findings were compatible with a rhomboencephalitis with coalescing microabscesses.

Management

A 2nd lumbar puncture was performed, which came back positive for Listeria monocytogenes. Antibiotic therapy was started. However, after one week of intravenous amoxicillin 2 g 4-hourly and intravenous co-trimoxazole 960 mg 6-hourly, the patient died due to respiratory complications.

DISCUSSION

Etiology and demographics

Listeriosis may target the meninges resulting in the clinical picture of acute/subacute bacterial meningitis, occurring in 30% of Listeria monocytogenes infections, with a mortality rate of 15% [1]. LR occurs in upto 11 - 24% of patients with CNS listeriosis, a rare but pathognomonic form of central nervous system infection with a high mortality rate of around 51% [2].

Listeria monocytogenes is a gram-positive facultative intracellular bacterium, for which soil appears to be the chief environmental reservoir. Soft cheeses and undercooked meats are among the most common modes of transmission, although it was not the case for our patient [3]. After exposure, bacteria invades the brainstem by retrograde axonal migration along the oropharyngeal cranial nerves [1].

Factors predisposing to LR in humans have not been identified. Unlike listerial meningitis, where two-thirds of patients are immunosuppressed, LR has been reported to occur mainly in immunocompetent adults [4].

Clinical and imaging findings

LR typically begins with a prodrome of 4 days of headache (73%), fever (85%) and nausea or vomiting (64%), but these symptoms were absent in our patient. The prodrome is usually followed by the onset of focal signs of lower brainstem and cerebellar involvement [2,5,6]. Patients frequently present with asymmetrical cranial nerve dysfunction, since involvement of the nuclei of the cranial nerves is visible with a frequency of 78% for nerve VII (as in our patient), 74% for nerve VI and less for the other cranial nerves (V, IX, XI). Most patients have combined cranial nerve deficits and long-tract signs (81%), and 50% have cerebellar signs [6].

CSF abnormalities in LR are not typical of bacterial infections and 20% of patients may have normal CSF on initial sampling, such in our case [7]. In a review of patients with LR by Armstrong and Fung, CSF Gram stains were positive in only 10% of cases and CSF cultures were positive in 33 - 41% and blood cultures in 61% [8,9].

MRI is the radiological investigation of choice in the diagnosis of brainstem disease. Characteristic MRI changes include T2 and FLAIR hyperintensity in the brainstem and cerebellum, patchy diffusion restriction and ring enhancing lesions [1,10]. In our case, the lesions of the brainstem showed first focal diffusion retriction and pseudonodular enhancement, probably due to a relatively early stage of abscesses.

Treatment

Amoxicillin or ampicillin is generally considered as the most effective antimicrobial for LR. Gentamicin has been used in combination and appeared to be effective. Early treatment with ampicillin is associated with improved survival (76%) [8].

Differential diagnosis

The differential diagnosis of LR varies, depending on the age and geographical region. Non-infectious pathologies enter more frequently in the differential diagnosis , including multiple sclerosis, ADEM, Behcet's disease, paraneoplastic syndrome, and , very rarely, lymphoma. Infectious causes in adults include viruses, such as entero-, herpesviruses and Epstein-Barr virus and Streptococcus pneumoniae [1,6,10]. Tuberculosis and progressive multifocal leucoencephalopathy due to JC virus infection are also important to be considered [1].

TEACHING POINT

Listeria rhomboencephalitis is a rare but severe manifestation of Listeria infection of the central nervous system whic often occurs in healthy subjects , with a high mortality rate. The clinical and biological presentation might be misleading but the topography and morphology of the lesions at MRI is typical and should rise the suspicion of LR, in order to obtain early treatment and to improve survival.

QUESTIONS

Question 1: What is the pathogenesis of Listeria rhomboencephalitis?

Answers

1. Infectious
2. Neoplastic
3. Inflammatory
4. Demielinating

The correct answer is 1

Question 2: Does rhomboencephalitis affect:

Answers

1. The cerebral hemisphers
2. The hippocampus
3. The dural sinuses
4. The brainstem

The correct answer is 4

Question 3: Which cranial nerve is most often involved in Listeria Rhomboencephalitis?

Answers

1. III
2. IV
3. VII
4. V

The correct answer is 3

Question 4: Which pattern of enhancement is most often observed in Listeria Rhomboencephalitis?

Answers

1. Nodular
2. There is no enhancement
3. Diffuse and irregular
4. Patchy enhancement with ring enhancing lesions

The correct answer is 4

Question 5: Which of the following is not a differential diagnosis of Listeria Rhomboencephalitis?

Answers

1. Bechet disease
2. ADEM
3. Multiple sclerosis
4. Ependymoma

The correct answer is 4.

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FIGURES

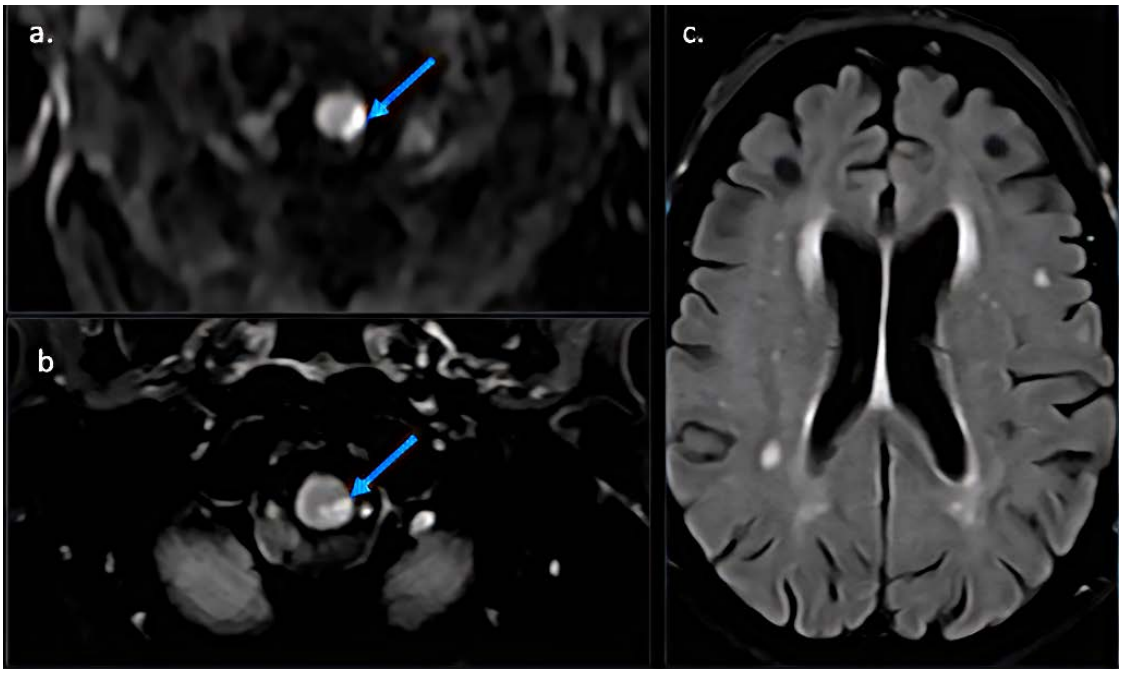

Figure 1: 81 years old female with ischemic bulbo-medullary lesion.

FINDINGS: a. And b. MRI axial images show respectivelly diffusion restriction (arrow) and FLAIR hyperintense signal (arrow) of a right sided ischemic bulbo-medullary lesion. c. Axial FLAIR image show multiple foci of signal hyperintensity

TECHNIQUE: 3T MRI. without contrast administration. a Axial DWI image: B0/B1000, TR/TE:4110ms/88.40ms, slice thickness:4mm. B. and c. FLAIR axial images: TR/TE: 3500ms/90ms, Inversion time: 1800ms. Slice thickness 4mm.

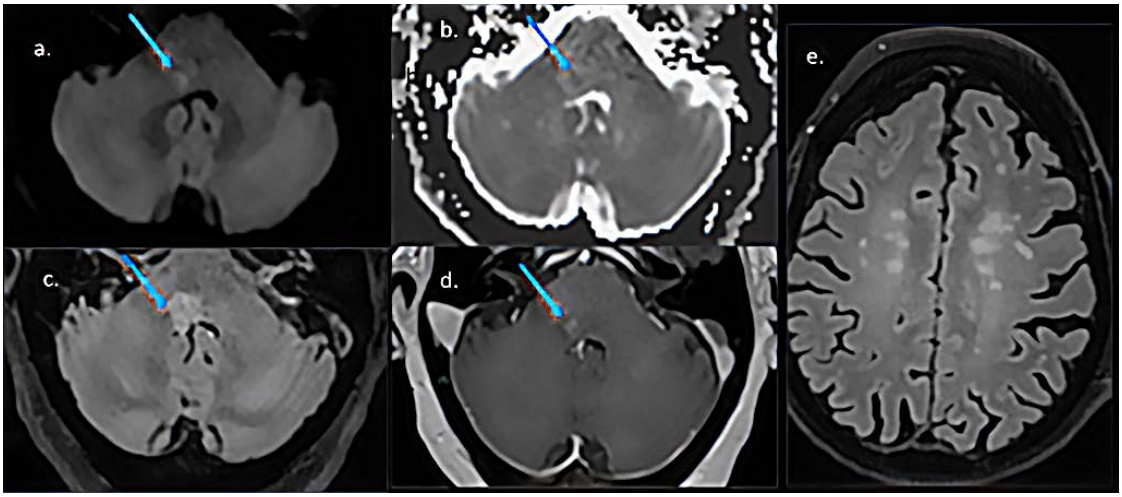

Figure 2: 81 years old female Listeria Rhomboencephalitis

FINDINGS: (a) Axial DWI, ADC map (b) axial FLAIR (c) and 3D-T1 VIBE after gadolinium injection (d) shows the appearence of a new lesion with moderate diffusion restriction and nodular enhancement in the left dorsal aspect of the pons (arrows). e. Axial FLAIR images show an increased number of white matter foci of signal hyperintensity, in keeping with the first diagnosis of demyelinating disease.

TECHNIQUE: 3T MRI. a. Axial DWI image: B0/B1000, TR/TE:4110ms/88.40ms, slice thickness:4mm. and (b.) ADC map. c. and e. FLAIR 3D axial images on axial plane: TR/TE: 3700ms/100ms, Inversion time: 1800ms. Slice thickness 1mm. D. 3D-T1 VIBE image, TR/TE: 2140 ms/3.4 ms, slice thickness

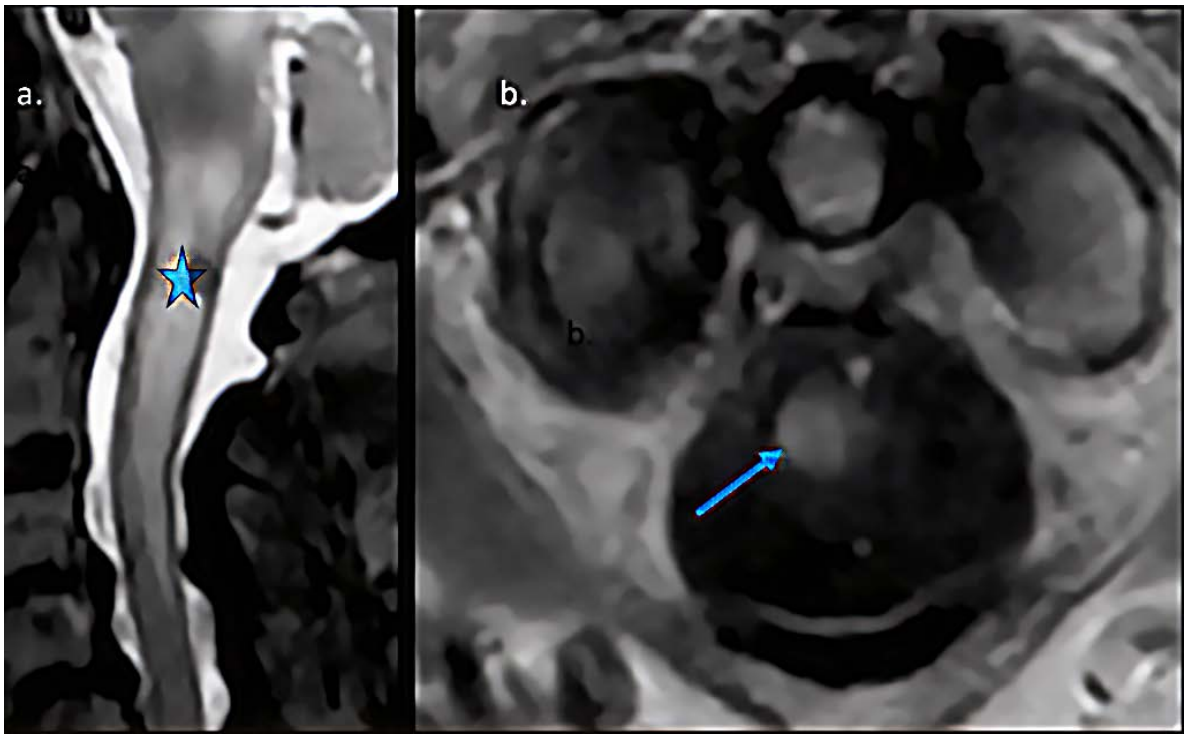

Figure 3: 81 years old female with Listeria Rhomboencephalitis.

FINDINGS: Sagittal STIR show the appearence of an extensive right sided bulbo-spinal lesion (star) with enhancement after contrast injection (arrow), as shown on the axial 3D-T1 vibe image (b)

TECHNIQUE: 3T MRI. a. Sagittal STIR (TR/TE: 2700ms/70ms. Slice thickness 3mm. b. 3D-T1 VIBE image (TR/TE: 2140 ms/3.4 ms, slice thickness: 1mm) after administration of Dotarem (0.2 ml/Kg).

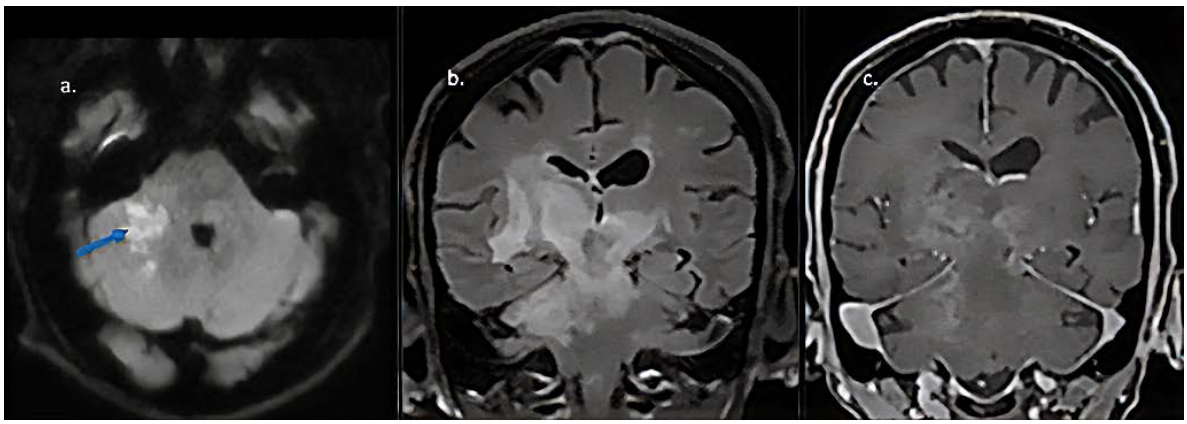

Figure 4: 81 years old female with Listeria Rhomboencephalitis.

FINDINGS: a.Axial DWI shows patchy diffusion restriction in the right cerebellum and medial cerebellar peduncle (arrow). b. Coronal FLAIR image shows extensive signal abnormality in the brainstem, cerebellum and right basal ganglia, with patchy enhancement on coronal 3D-T1 VIBE image (c) in keeping with the diagnosis of rhomboencephalitis with microabscesses.

TECHNIQUE: MRI 3T. a. Axial DWI image: B0/B1000, TR/TE:4110ms/88.40ms, slice thickness: 4mm. b. FLAIR 3D coronal images on axial plane: TR/TE: 3700ms/100ms, Inversion time: 1800ms. Slice thickness 1mm. c. 3D-T1 VIBE coronal image: TR/TE: 2140 ms/3.4 ms, slice thickness: 1mm, after administration of Doterem (0.2 ml/Kg).

KEYWORDS

Rhomboencephalitis, Listeria monocitogenes, Brainstem, Abscess, Magnetic Reseonance Imaging