Isolated Leptomeningeal Enhancement in Anti-N-methyl-D-aspartate Receptor Encephalitis: A case report

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Case report

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Abstract

Background Magnetic resonance imaging findings of anti-N-methyl-D-aspartate (anti-NMDA) receptor encephalitis have been reported to lack specificity. The hippocampus, frontal lobe, white matter, basal ganglia, and even spinal cord can be involved. However, isolated leptomeningeal enhancement is rare in anti-NMDA receptor encephalitis. Case presentation We report a 17-year-old boy who presented with a 2-week history of mild fever and progressive encephalopathy with behavioral and psychological abnormalities. Finally, a diagnosis of anti-NMDA receptor encephalitis was made. Initial magnetic resonance imaging results revealed diffuse contrast enhancement of the leptomeninges without parenchymal lesions. After treatment with high-dose intravenous methylprednisolone, plasma exchange, and intravenous immunoglobulin, the enhancement showed substantially decrease at the 3-week follow-up. Conclusions This case is unusual because isolated leptomeningeal enhancement is rare in anti-NMDA receptor encephalitis. The present case suggests that isolated diffuse leptomeningeal enhancement could be one of the imaging findings in patients with anti-NMDA receptor encephalitis.

Background

Magnetic resonance imaging findings of anti-N-methyl-D-aspartate (anti-NMDA) receptor encephalitis have been reported to lack specificity. The hippocampus, frontal lobe, white matter, basal ganglia, and even spinal cord can be involved [1]. Herein, we report an unusual magnetic resonance imaging finding in a 17-year-old patient with anti-NMDA receptor encephalitis. This case is unusual because isolated leptomeningeal enhancement is rare in anti-NMDA receptor encephalitis [1]. The present case suggests that isolated leptomeningeal enhancement could be one of the imaging findings in patients with anti-NMDA receptor encephalitis.

Case Presentation

A 17-year-old boy presented with a 2-week history of mild fever and progressive encephalopathy with behavioral and psychological abnormalities. According to the patient's history, he received a dog bite 7 years previously. After the attack, the patient was vaccinated against rabies. A neurological examination at admission revealed confusion, disorientation, and behavioral disturbance with agitation. Diagnostic studies of cerebrospinal fluid showed an increased number of white blood cells (32 × 106/L; 84% lymphocytes) and normal levels of protein, glucose, and chlorides. Additional tests, including high-throughput sequencing analysis of cerebrospinal fluid, showed no evidence of infection. Oligoclonal banding was positive in cerebrospinal fluid and negative in serum. Magnetic resonance imaging results revealed contrast enhancement of the leptomeninges without parenchymal lesions (Figure 1A). An additional movie file shows this in more detail (see Additional file 1). Anti- NMDA receptor antibodies were positive in serum (1:32) and cerebrospinal fluid (1:3.2) and a diagnosis of anti-NMDA receptor encephalitis was made. Tumor screening results were negative. High-dose intravenous methylprednisolone (1.0 g/d) with gradual tapering was initiated. Intravenous immunoglobulin (0.4 g/kg/d for 5 d) was administered after the patient completed a 5-d course of plasma exchange. The

patient showed gradual improvement after the treatment. Repeat magnetic resonance imaging after 3 week showed substantially decrease of the enhancement (Figure 1B). An additional movie file shows this in more detail (see Additional file 2).

Discussion

A previous study reported that 5 of 106 patients with anti-NMDA receptor encephalitis had meninges enhancement [2]; however, the authors did not report whether these 5 patients had concomitant parenchymal lesions. Furthermore, the pattern of leptomeningeal enhancement was not reported. The present case suggests that isolated diffuse leptomeningeal enhancement could be one of the imaging findings in patients with anti-NMDA receptor encephalitis.

Leptomeningeal enhancement has received renewed attention in other types of immune-mediated diseases of the central nervous system such as multiple sclerosis [3-5]. Histopathologic analysis has shown clustered inflammatory cells in the subarachnoid space in the area of in vivo leptomeningeal-compartment enhancement [3]. These cells are sometimes organized into ectopic follicle-like structures containing B and T cells, and they are often found around meningeal vessels; however, this produces non-diffuse enhancement of leptomeninges in multiple sclerosis, unlike the present case.
In studies on meningitis (including viral etiology), leptomeningeal enhancement has also been reported because of the disruption of the blood–brain barrier in the inflamed meninges [6]. Although no evidence of infection was found in the present case after a thorough test of cerebrospinal fluid, concurrent viral meningitis cannot be completely ruled out. However, the meningeal enhancement in the present case was not consistent with viral etiology because the grade of enhancement should be low and subtle in viral meningitis, particularly on contrast-enhanced images based on T1-weighted [6, 7].

Leptomeningeal enhancement based on T1-weighted imaging has various limitations, including excessive vascular enhancement, flat images, and inflow effect, which suggest that the observed enhancement might only be adjacent enhanced vessels rather than enhanced meninges [8]. However, repeat contrast-enhanced T1-weighted imaging using the same scan protocol suggested definite leptomeningeal enhancement in the present case.

Conclusions

This case was unusual because isolated diffuse leptomeningeal enhancement in anti-NMDA receptor encephalitis is not reported in the literature. More attention should be paid to the role of leptomeningeal enhancement in anti-NMDA receptor encephalitis.

Abbreviations

anti-NMDA: anti-N-methyl-D-aspartate

Declarations

Ethics approval and consent to participate

This study was approved by ‘Ethics committee of the First Affiliated Hospital of Zhengzhou University’ with written informed consent from patient's legal representatives.

Consent for publication

The legal representatives have consented to submission of this case report to the journal, and we have obtained a written informed consent.

Availability of data and materials

All data related to this case report are contained within the manuscript.

Competing interests

The authors report no conflict of interest.

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Authors’ contributions

PPN: drafting and revising the manuscript.

BS: drafting and revising the manuscript.

YMX: study concept and supervision; revising the manuscript.

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Figures

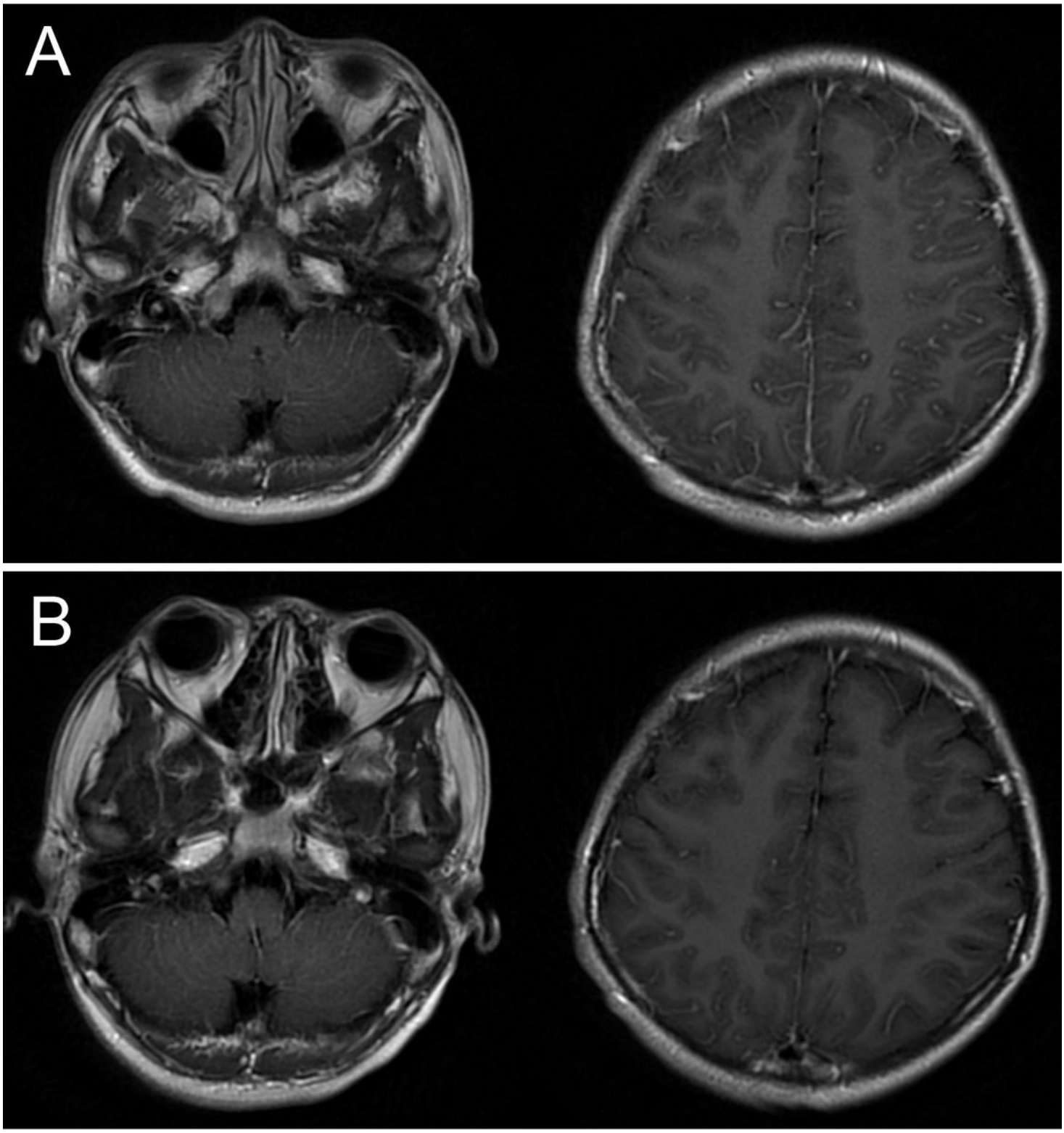

Figure 1

A: Contrast-enhanced T1-weighted magnetic resonance images showing diffuse leptomeningeal enhancement along the cortical sulci and cerebellar sulci. Notably, some of the hyperintensity in the sulcal spaces are vessels. B: Repeat contrast-enhanced T1-weighted magnetic resonance images showing substantially decrease of the enhancement.

Supplementary Files

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