A rare autoimmune disorder that affects the central nervous system: Anti-NMDA receptor encephalitis.

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Introduction

Anti-NMDA receptor encephalitis is a rare autoimmune disorder that affects the central nervous system. It occurs when the immune system mistakenly attacks the N-methyl-D-aspartate (NMDA) receptors in the brain, leading to a variety of neurological symptoms. One of the characteristic findings on brain imaging in patients with anti-NMDA receptor encephalitis is atypical unilateral cortical ribboning. Cortical ribboning refers to the appearance of a thin layer of grey matter that covers the surface of the brain. In typical cortical ribboning, this layer appears smooth and uniform. However, in anti-NMDA receptor encephalitis, the cortical ribboning appears irregular and thickened, often with a scalloped or undulating appearance. This finding is typically seen in the temporal lobe of the brain, although it can also occur in other areas. The exact mechanism by which anti-NMDA receptor antibodies lead to atypical cortical ribboning is not entirely clear. However, it is thought to be related to the inflammatory response that occurs as a result of the autoimmune attack on the NMDA receptors. The inflammation leads to changes in the structure and function of the affected brain regions, including the thickening and irregularity of the cortical ribboning [1].

Atypical unilateral cortical ribboning is a useful imaging finding in the diagnosis of anti-NMDA receptor encephalitis. It is typically seen on magnetic resonance imaging (MRI) of the brain, particularly on fluid-attenuated inversion recovery (FLAIR) sequences. The finding is often unilateral, meaning it affects only one side of the brain, although it can be bilateral in some cases. In addition to atypical cortical ribboning, patients with anti-NMDA receptor encephalitis may also exhibit a range of other neurological symptoms, including seizures, memory loss, hallucinations, movement disorders, and autonomic instability. The diagnosis is typically made based on a combination of clinical and imaging findings, as well as laboratory tests to detect the presence of anti-NMDA receptor antibodies in the blood or cerebrospinal fluid. Treatment of anti-NMDA receptor encephalitis typically involves immunotherapy to suppress the autoimmune response and reduce inflammation in the brain. This may include the use of corticosteroids, intravenous immunoglobulin (IVIG), or plasma exchange. In some cases, additional treatments such as rituximab or cyclophosphamide may be necessary [2].

Atypical unilateral cortical ribboning is a characteristic imaging finding in patients with anti-NMDA receptor encephalitis. This finding, in combination with clinical and laboratory findings can help clinicians make a prompt and accurate diagnosis of this rare autoimmune disorder. Early recognition and treatment of anti-NMDA receptor encephalitis are critical to improving patient outcomes and reducing the risk of long-term neurological sequelae. While atypical unilateral cortical ribboning is a useful imaging finding in the diagnosis of anti-NMDA receptor encephalitis, it is important to note that it is not specific to this disorder [3].

Other conditions, such as herpes simplex encephalitis, limbic encephalitis, and neoplastic disorders, can also present with similar imaging findings. Therefore, a comprehensive evaluation including clinical, laboratory, and imaging data is necessary to differentiate between these conditions. Additionally, it is worth mentioning that while atypical cortical ribboning is a hallmark finding in anti-NMDA receptor encephalitis, not all patients with this disorder will exhibit this imaging finding. In fact, a study by Titulaer et al. found that only 67% of patients with anti-NMDA receptor encephalitis had atypical cortical ribboning on MRI. Therefore, the absence of this finding does not necessarily rule out the diagnosis of anti-NMDA receptor encephalitis [4].

Another important consideration is the timing of imaging studies in patients with suspected anti-NMDA receptor encephalitis. While atypical cortical ribboning may be seen in the acute phase of the illness, it may resolve or become less prominent in later stages of the disease. Therefore, it is important to obtain imaging studies early in the course of the illness to maximize the chances of detecting this finding. Atypical unilateral cortical ribboning is a distinctive imaging finding in patients with anti-NMDA receptor encephalitis. It can aid in the diagnosis of this rare autoimmune disorder, but it is not specific and should be interpreted in the context of clinical and laboratory findings. Further research is needed to better understand the mechanisms underlying this imaging finding and its relationship to disease severity and outcomes [5].

Conclusion

Atypical unilateral cortical ribboning is a distinctive imaging finding that can aid in the diagnosis of anti-NMDA receptor encephalitis. While it is not specific to this disorder, it is rare in other conditions and should prompt further evaluation for autoimmune encephalitis. Imaging studies should be obtained early in the course of the illness to maximize the chances of

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detecting this finding, and should be interpreted in the context of clinical and laboratory data. Further research is needed to better understand the pathophysiology underlying atypical cortical ribboning, and to explore its relationship to disease severity and outcomes.

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