Uncommon Alliance : MOGAD and Behçet Disease

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INFLAMMATOIRE

Introduction

Myelin Oligodendrocyte Glycoprotein Antibody-Associated Disease (MOGAD) presents a broad spectrum of symptoms, including encephalitis and demyelination. Its relapses can be triggered by various factors, including previous immune conditions. This study reports on a case of MOGAD with longitudinally extensive transverse myelitis (LETM) occurring alongside Behçet's disease, marking a potentially significant correlation not previously documented.

Case

- A 40-year-old female, with Behçet's disease since 2014 (characterized by five annual episodes of bipolar aphthosis and inflammatory arthralgias with positive HLA B51), treated with Colchicine, presented with acute progressive weakness in the lower extremities and voiding dysfunction over three days. At the neurological examination, the patient was bedridden, presenting asymmetric paraplegia and a sensory level at D4. Babinski reflexes were positive on both sides. MRI revealed an extensive hyperintensity from D1 to D8, with gadolinium enhancement (Figure 1A), and displayed an 'H' sign in axial sequences (Figure 1B). The brain MRI was normal. CSF analysis was normal, with negative oligoclonal bands. * Serum MOG antibodies were positive, while anti-aquaporin-4 antibodies and other autoimmune tests were negative. Initial treatment involved steroids and plasma exchange, with significant improvement. However, relapse led to the initiation of long-term Mycophenolate Mofetil (MMF), stabilizing her condition with minimal assistance required for walking.

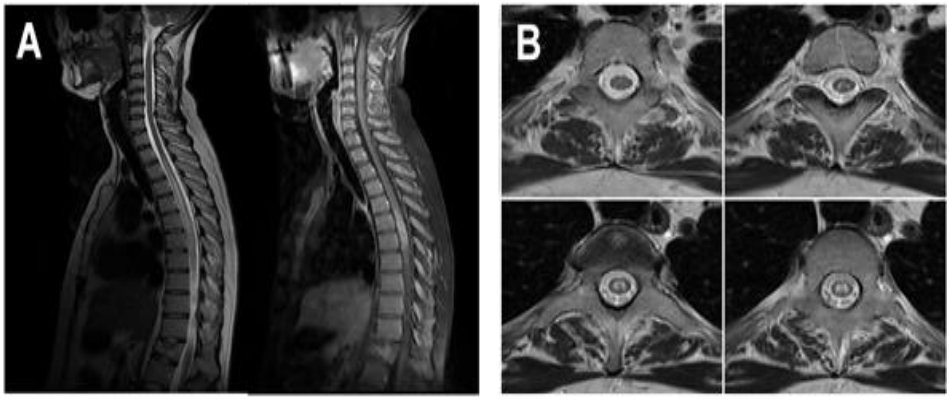

Figure : A :T2-weighted magnetic resonance imaging showing an extensive hyperintensity from D1 to D8, with gadolinium enhancement. B : Axial T2 weighed magnetic resonance image with an 'H' sign.

Discussion

This case illustrates the intersection of Behçet's disease and MOGAD, suggesting that underlying autoimmune dysregulation may predispose to both MOGAD and its relapses. China revealed that approximately one-third of MOGAD patients had a history of other immunological conditions, a factor significantly linked to the occurrence of relapses [1]. Unlike the common association between MOGAD and other autoimmune diseases, this case highlights the potential for unique immunopathogenic mechanisms influencing MOGAD's recurrent nature [2].

Conclusion

The co-occurrence of Behçet's disease with MOGAD may trigger relapses, emphasizing the need for long-term management strategies to reduce recurrence and disability. This case expands the understanding of MOGAD, suggesting that pre-existing immune dysregulation could play a crucial role in its recurrence and management.

References

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[2] Kunchok A, Chen JJ, Saadeh RS, Wingerchuk DM, Weinshenker BG, Flanagan EP, et al. Application of 2015 seronegative neuromyelitis optica spectrum disorder diagnostic criteria for patients with myelin oligodendrocyte glycoprotein IgG–associated disorders. JAMA neurology. 2020 Dec 1;77(12):1572-5.