






IgG4-related hypertrophic pachymeningitis with cerebral venous thrombosis

Paquimeningite hipertrófica relacionada à IgG4 associada a trombose venosa cerebral

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A 58-year-old female patient presented with a 4-year history of right-sided headache with tinnitus. She had been hospitalized previously due to dysfunction of multiple cranial nerves, such as diplopia, right hemiface hypoesthesia, right peripheral facial palsy, right hearing loss, dysphonia, and dysphagia, followed by venous thrombosis of the right sigmoid sinus (→ **Figure 1**), which was treated with warfarin. The cerebrospinal fluid yielded 90 leukocytes/mm³ (92% of lymphocytes) and 158 mg/dL of proteins. A magnetic resonance imaging (MRI) scan revealed tentorium-temporo-parietal hypertrophic pachymeningitis (→ **Figure 2**). A meningeal biopsy demonstrated dense lymphoplasmacytic infiltrate and storiform fibrosis (→ **Figure 3**), two of the three histopathological criteria.^{1,2} Therefore, the diagnosis was made, and rituximab^{3,4} was initiated, with a very important response in terms of symptoms and imaging exams (→ **Figure 4**).

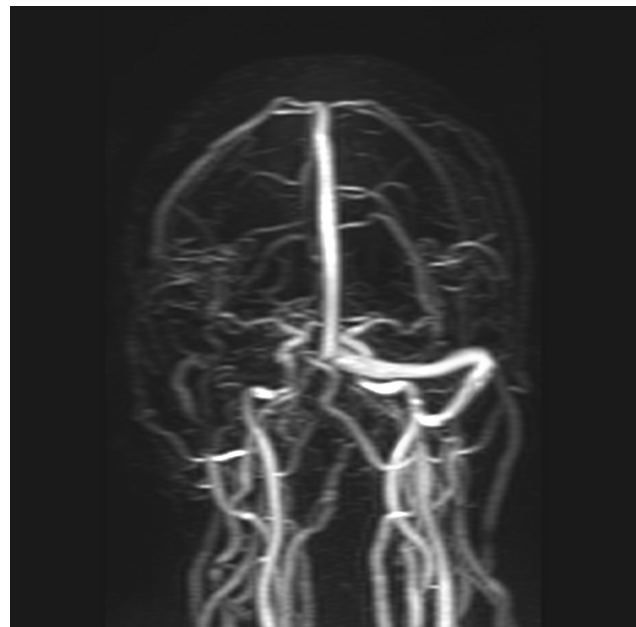


Figure 1 Venous magnetic resonance angiography showing venous thrombosis at the right side.

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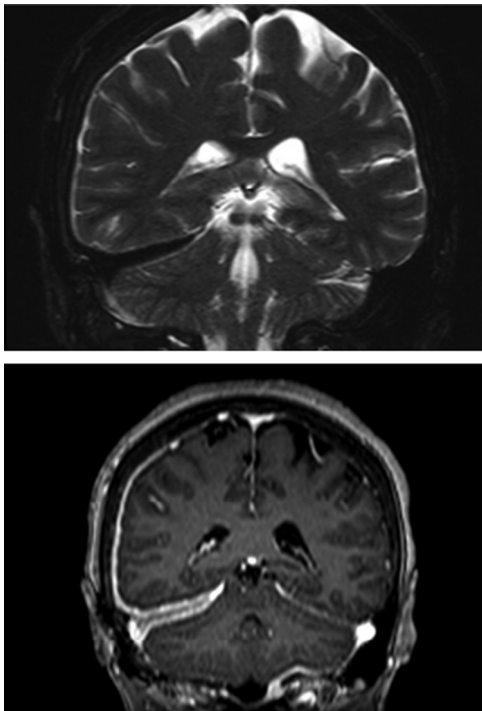


Figure 2 T2 (upper) and postcontrast T1 (bottom) weighted magnetic resonance imaging (MRI) scans, showing dural thickening on coronal view before treatment.

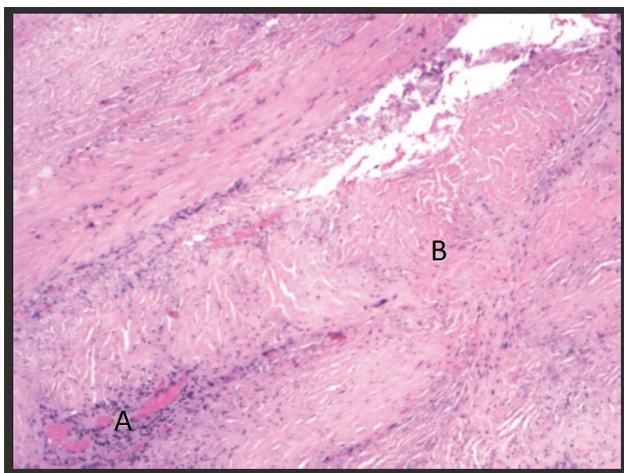


Figure 3 Meningeal biopsy in hematoxylin and eosin stain demonstrating dense lymphoplasmacytic infiltrate (A) and storiform fibrosis (B), two of the three histopathological criteria of IgG4-related disease.

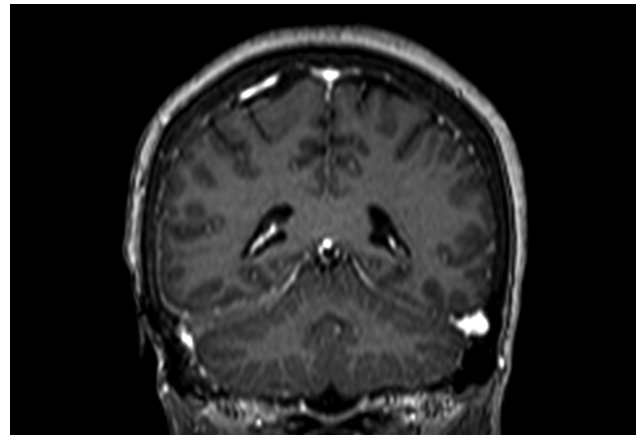


Figure 4 Postcontrast weighted MRI scan showing improvement in dural thickening at the right side on coronal view after treatment with rituximab.

Authors' Contributions

AVKC: writing of the original draft, writing – review and editing; ACA: writing of the original draft; SMCASMMG, AJR, and HRSN: supervision.

Conflict of Interest

AJR is an Associate Editor of this journal.

References

- 1 De Virgilio A, de Vincentiis M, Inghilleri M, et al. Idiopathic hypertrophic pachymeningitis: an autoimmune IgG4-related disease. *Immunol Res* 2017;65(01):386–394. Doi: 10.1007/s12026-016-8863-1
- 2 Deshpande V, Zen Y, Chan JK, et al. Consensus statement on the pathology of IgG4-related disease. *Mod Pathol* 2012;25(09):1181–1192. Doi: 10.1038/modpathol.2012.72
- 3 Lu LX, Della-Torre E, Stone JH, Clark SW. IgG4-related hypertrophic pachymeningitis: clinical features, diagnostic criteria, and treatment. *JAMA Neurol* 2014;71(06):785–793. Doi: 10.1001/jamaneurol.2014.243
- 4 Perugino CA, Stone JH. IgG4-related disease: an update on pathophysiology and implications for clinical care. *Nat Rev Rheumatol* 2020;16(12):702–714. Doi: 10.1038/s41584-020-0500-7