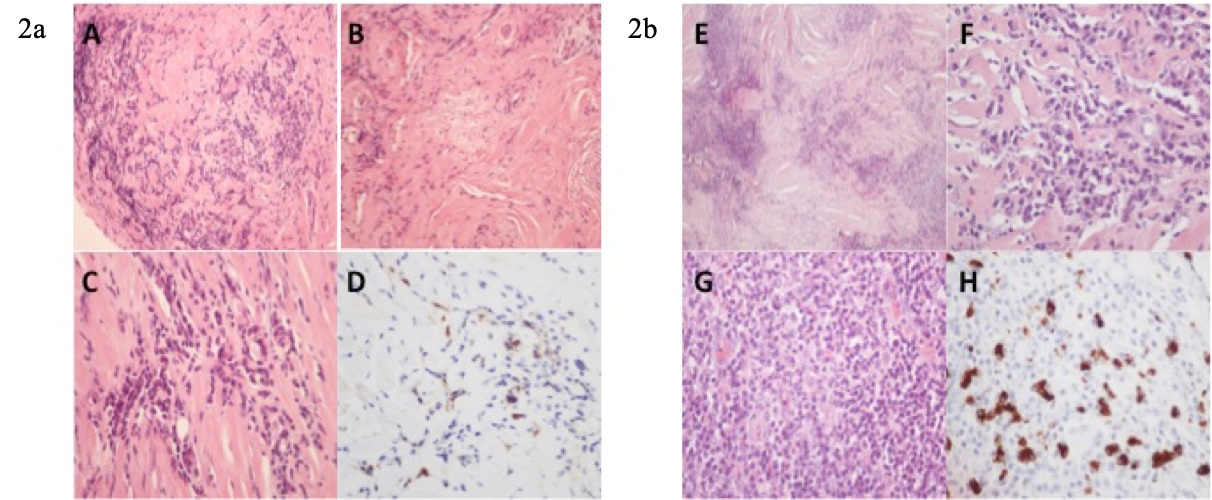
**Supplemental data**

**Figure e-1 : Histological findings of both IgG4-HP patients.**

****

**a)** A : Hematoxylin-Eosin-Saffron (HES) staining, x 200: fibrosis characterized by collagen keychains, intermingled with an inflammatory infiltrate. B: HES staining, x 200: storiform appearance fibrosis C: HES staining, x 400: inflammatory infiltrate rich in plasma cells. D: immunostaining anti-IgG4, x 400: numerous plasma cells (> 10 / HPF) expressing IgG4. **b)** E: HES x 40 staining: dissecting fibrosis with abundant inflammatory infiltrate producing layers or nodules. F: HES x 400 staining: inflammatory infiltrate rich in plasma cells, mixed with thick collagenic cysts. G: HES x 400 staining: in places abundant, polymorphous inflammatory infiltrate associating plasma cells, macrophage lymphocytes and neutrophils. H: anti-IgG4 x 400 immunostaining: numerous plasmocytes expressing IgG4 (> 10/HPF)

**Table e-1: Steroid therapy data**

|  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- |
|  | Total IgG4-HP | | Systemic IgG4-HP | | Isolated IgG4-HP | | Non specified IgG4-HP | |
|  | n | 60 | n | 18 | n | 25 | n | 17 |
| IV pulse steroid therapy  No IV pulse therapy | 39 | 20 (20%)  19 (60%) | 12 | 8 (28.6%)  4 (50%) | 23 | 10 (16.7%)  13 (63.6%) | 4 | 2 (0%)  2 (-) |
| Initial dose of oral steroids  < 30 mg/day  30-49 mg/day  > 49 mg/day | 27 | 2 (-)  6 (100%)  19 (52.6%) | 11 | 1 (-)  2 (100%)  8 (50%) | 15 | 1 (-)  4 (100%)  10 (60%) | 1 | 0  0  1 (-) |
| Duration of steroid therapy  < 6 months  6-12 months  > 12 months | 25 | 9 (100%)  6 (83.3%)  10 (42.9%) | 7 | 2 (100%)  0  5 (33.3%) | 14 | 5 (100%)  5 (100%)  4 (66.6%) | 4 | 2 (-)  1 (-)  1 (-) |

The percentage in parenthesis available for each box corresponds to the number of relapsing patients. The sign “-“ was used when the data was missing.

**e-References**

1. Takeuchi S, Osada H, Seno S, Nawashiro H. IgG4-Related Intracranial Hypertrophic Pachymeningitis : A Case Report and Review of the Literature. J Korean Neurosurg Soc [online serial]. 2014;55:300.
2. Ramirez L, D’Auria A, Popalzai A, Sanossian N. Bilateral Vision Loss Secondary to Pachymeningitis in a Patient with IgG4-Related Disease. Front Neurol [online serial]. 2014;5:192.
3. Liao B, Kamiya-Matsuoka C, Fang X, Smith RG. Refractory IgG4-related intracranial hypertrophic pachymeningitis responded to rituximab. Neurol - Neuroimmunol Neuroinflammation [online serial]. 2014;1:e41.
4. Lee Y-S, Lee HW, Park K-S, Park S-H, Hwang J-H. Immunoglobulin G4-Related Hypertrophic Pachymeningitis with Skull Involvement. Brain Tumor Res Treat [online serial]. 2014;2:87.
5. Kim SH, Kang Y, Oh SH, Paik S, Kim JS. Paraplegia in a Patient With IgG4-Related Sclerosing Disease: A Case Report. Ann Rehabil Med [online serial]. 2014;38:856.
6. Li L, Tse PY, Tsang FC, Lo RC, Lui W, Leung GK. IgG4-Related Hypertrophic Pachymeningitis at the Falx Cerebrii with Brain Parenchymal Invasion: A Case Report. World Neurosurg [online serial]. 2015;84:591.e7-591.e10.
7. Gu R, Hao P, Liu J, Wang Z, Zhu Q. Cervicothoracic spinal cord compression caused by IgG4-related sclerosing pachymeningitis: a case report and literature review. Eur Spine J [online serial]. 2016;25:147–151.
8. Lu Z, Tongxi L, Jie L, et al. IgG4-related spinal pachymeningitis. Clin Rheumatol [online serial]. 2016;35:1549–1553.
9. Williams T, Marta M, Giovannoni G. IgG4-related disease: a rare but treatable cause of refractory intracranial hypertension. Pract Neurol [online serial]. 2016;16:235–239.
10. Gospodarev V, Câmara J, Chakravarthy V, et al. Treatment of IgG4-related pachymeningitis in a patient with steroid intolerance: The role of early use of rituximab. J Neuroimmunol. 2016;299:62–65.
11. Ioannidis P, Parissis D, Bakirtzis C, Karayannopoulou G, Kanitakis J. Isolated IgG4-related hypertrophic pachymeningitis. Acta Neurol. Belg. 2017. p. 569–571.
12. Vanegas-Garcia AL, Calle-Lopez Y, Zapata CH, Alvarez-Espinal DM, Saavedra-Gonzalez YA, Arango-Viana JC. [Central nervous system in IgG4-related disease: case report and literature review]. Rev Neurol [online serial]. 2016;63:119–124.
13. Radotra BD, Aggarwal A, Kapoor A, Singla N, Chatterjee D. An orphan disease: IgG4-related spinal pachymeningitis: report of 2 cases. J Neurosurg Spine [online serial]. 2016;25:790–794.
14. Rice CM, Spencer T, Bunea G, Scolding NJ, Sloan P, Nath U. Intracranial spread of IgG4-related disease via skull base foramina. Pract Neurol [online serial]. 2016;16:240–242.
15. Hwang G, Jin S-Y, Kim H-S. IgG4-related disease presenting as hypertrophic pachymeningitis and compressive optic neuropathy. Jt Bone Spine [online serial]. 2016;83:601–602.
16. Schubert RD, Wood M, Levin MH, Perry A, Gelfand JM. The severe side of the IgG4-related hypertrophic pachymeningitis disease spectrum. Neurol Neuroimmunol neuroinflammation [online serial]. 2016;3:e197.
17. Maher M, Zanazzi G, Faust P, Nickerson K, T. Wong T. IgG4-related hypertrophic pachymeningitis of the spine with MPO-ANCA seropositivity. Clin Imaging. 2017;46:108–112.
18. Varrassi M, Gianneramo C, Arrigoni F, et al. Neurological involvement of IgG4-related disease: description of a case and review of the literature. Neuroradiol J [online serial]. Epub 2017.:197140091769817.
19. Massey J. IgG4-related hypertrophic pachymeningitis coexpressing antineutrophil cytoplasmic antibodies. Neurol. Neuroimmunol. NeuroInflammation 2017.
20. Waheed W, Skidd PM, Borden NM, Gibson PC, Babi MA, Tandan R. Metachronous Involvement, Diagnostic Imprecision of Serum Immunoglobulin G4 Levels, and Discordance Between Clinical and Radiological Findings in Immunoglobulin G4–Related Pachymeningitis. JCR J Clin Rheumatol [online serial]. 2017;23:215–221.
21. Lourenço EP, Nzwalo H, Sampaio MR, Afonso L. IgG4-related disease presenting with headache and papilloedema. BMJ Case Rep [online serial]. 2016;2016:bcr2016216435.
22. Della-Torre E, Passerini G, Furlan R, et al. Cerebrospinal fluid analysis in immunoglobulin G4-related hypertrophic pachymeningitis. J. Rheumatol. 2013. p. 1927–1929.
23. Utsuki S, Kijima C, Fujii K, Miyakawa S, Iizuka T, Hara A. Investigation of IgG4-positive cell infiltration in biopsy specimens from cases of hypertrophic pachymeningitis. Clin Neuropathol [online serial]. 2013;32:84–90.
24. Lipton S, Warren G, Pollock J, Schwab P. IgG4-related disease manifesting as pachymeningitis and aortitis. J. Rheumatol. 2013. p. 1236–1237.
25. Norikane T, Yamamoto Y, Okada M, et al. Hypertrophic Cranial Pachymeningitis With IgG4-Positive Plasma Cells Detected by C-11 Methionine PET. Clin Nucl Med [online serial]. 2012;37:108–109.
26. Shapiro KA, Bové RM, Volpicelli ER, Mallery RM, Stone JH. Relapsing course of immunoglobulin g4-related pachymeningitis. Neurology. 2012;79:604–605.
27. Della-Torre E, Bozzolo EP, Passerini G, Doglioni C, Sabbadini MG. IgG4-related pachymeningitis: Evidence of intrathecal IgG4 on cerebrospinal fluid analysis. Ann. Intern. Med. 2012. p. 401–402.
28. Lin C-K, Lai D-M. IgG4-related intracranial hypertrophic pachymeningitis with skull hyperostosis: a case report. BMC Surg [online serial]. 2013;13:37.
29. Tajima Y, Mito Y. Cranial neuropathy because of IgG4-related pachymeningitis; intracranial and spinal mass lesions. Case Reports [online serial]. 2012;2012:bcr2012006471-bcr2012006471
30. Yamashita H, Takahashi Y, Ishiura H, Kano T, Kaneko H, Mimori A. Hypertrophic Pachymeningitis and Tracheobronchial Stenosis in IgG4-related Disease: Case Presentation and Literature Review. Intern Med [online serial]. 2012;51:935–941
31. Choi SH, Lee SH, Khang SK, Jeon SR. IgG4-related sclerosing pachymeningitis causing spinal cord compression. Neurology. 2010;75:1388–1390.
32. Kosakai A, Ito D, Yamada S, Ideta S, Ota Y, Suzuki N. A case of definite IgG4-related pachymeningitis. Neurology. 2010;75:1390–1392.
33. Kim EH, Kim SH, Cho JM, Ahn JY, Chang JH. Immunoglobulin G4–related hypertrophic pachymeningitis involving cerebral parenchyma. J Neurosurg [online serial]. 2011;115:1242–1247.
34. Kato K, Tamano Y, Namioka A, et al. Hypertrophic pachymeningitis associated with autoimmune pancreatitis examined for IgG4 related disease: A case report. Neurol Surg. 2011;39:763–768.
35. Tsugawa J, Ouma S, Fukae J, Tsuboi Y, Sakata N, Inoue T. Multiple cranial neuropathies in a patient with IgG4-related hypertrophic pachymeningitis: A case report. Brain and Nerve. 2014;66:873–878.
36. Sakai T, Kondo M, Yoshii S, Tomimoto H. IgG4-related disease that presented cranial, cervical, lumbar and sacral hypertrophic pachymeningitis associated with infundibulo-hypophysitis. Rinsho Shinkeigaku [online serial]. 2014;54:664–667.
37. Chan S-K, Cheuk W, Chan K-T, Chan JKC. IgG4-related Sclerosing Pachymeningitis. Am J Surg Pathol [online serial]. 2009;33:1249–1252.
38. Tanboon J, Felicella MM, Bilbao J, Mainprize T, Perry A. Probable IgG4-related pachymeningitis: a case with transverse sinus obliteration. Clin Neuropathol [online serial]. 2013;32:291–297.
39. Hyun J-W, Kim S-H, Yoo H, Hong E, Huh S-Y, Kim HJ. Steroid-Resistant Relapsing IgG4-Related Pachymeningitis Treated With Methotrexate. JAMA Neurol [online serial]. 2014;71:222.
40. Imbergamo S, Campagnolo M, Manara R, Marino F, Adami F, Briani C. Teaching Neuro Images: Multifocal neurologic involvement as the only manifestation of IgG4-related disease. Neurology 2013.
41. Rodriguez-Castro E, Fernandez-Lebrero A, Lopez-Dequidt IA, et al. Hypertrophic pachymeningitis secondary to IgG4-related disease: case report and review of the literature. Rev Neurol [online serial]. 2015;61:308–312.
42. D. ZO, C. CO, M. B, S. C, M.C. Z. Pachymeningitis associated with IgG4 disease. Med [online serial]. 2017;77:242–244.
43. Della-Torre E, Campochiaro C, Cassione EB, et al. Intrathecal rituximab for IgG 4 -related hypertrophic pachymeningitis. J Neurol Neurosurg Psychiatry [online serial]. Epub 2017 Aug 17.:jnnp-2017-316519.
44. Kim EC, Lee SJ, Hwang HS, Kim J, Kim MS. Bilateral diffuse scleritis as a first manifestation of immunoglobulin G4–related sclerosing pachymeningitis. Can J Ophthalmol / J Can d’Ophtalmologie 2013;48:e31–3. doi:10.1016/j.jcjo.2012.11.006.
45. Wallace ZS, Carruthers MN, Khosroshahi A, et al. IgG4-Related Disease and Hypertrophic Pachymeningitis. Medicine (Baltimore) [online serial]. 2013;92:206–216.
46. Lindstrom KM, Cousar JB, Lopes MBS. IgG4-related meningeal disease: clinico-pathological features and proposal for diagnostic criteria. Acta Neuropathol [online serial]. 2010;120:765–776.
47. Makino S, Tanaka Y. Clinical &amp; Experimental Ophthalmology A Case of Hypertrophic Pachymeningitis with Elevated Serum IgG4. J Clin Exp Ophthalmol 2013;44172:2602155–9570. doi:10.4172/2155-9570.1000260.