BACKGROUND

Headache is the most common symptom in pachymeningitis from IgG4-related disease (IgG4-RD). Headache may also be present in extra-cranial IgG4-RD, for instance Mickulicz’s disease where lymph nodes, lacrimal glands and salivary glands are affected.

OBJECTIVE

• We report two patients with intra and extra-cranial IgG4-RD who presented with intractable headache to illustrate the challenges in diagnosing IgG4-RD.

CASE REPORT

A 63 year-old man with chronic headache presented with recurrent unilateral weakness and numbness associated with myoclonic jerks. MRI brain consistently showed right frontoparietal pachymeningeal thickening (Figure 1). Dural biopsy revealed non-specific inflammation. IgG4-RD was considered and the dural histology was reviewed when serum IgG4 was noted to be raised. Headache resolved with corticosteroid therapy. The MRI changes also improved (Figure 4).

Follow up MRI brain with contrast(Figure 4).
• Stable enhancing nodular thickening adjacent to the superior sagittal sinus.
• Stable gliosis of the tight high parietal lobe.
• No new enhancing intraparenchymal or extra-axial mass.

A 19 year-old man presented with acute onset of headache, painful lymphadenopathy, parotid (Figure 2) and lacrimal glands swelling (Figure 3). Submandibular gland biopsy showed inflammatory changes. IgG4-RD was considered when lymphadenopathy and glandular swellings worsened over a short time in the absence of infection or neoplasm. Review of submandibular gland histology confirmed IgG4-RD (Figure 5a-e). The patient did not respond to corticosteroids but to rituximab therapy.

• No intracranial abnormality especially that of pachymeningeal enhancement or pituitary abnormality detected in the MRI brain with contrast.
• CRP: 205mg/L.
  • Normal Range: 0-5mg/L.

DISCLOSURES / CONTACT INFORMATION

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Submandibular Gland Histology

CONCLUSIONS

• In both cases, accurate pathological diagnosis was the key. When the clinical suspicion of IgG4-RD is high, the characteristic features of IgG4-RD—namely, high population of IgG4 cells, lymphoplasmacytic infiltration, storiform fibrosis and obliterative phlebitis should be sought amidst the “non-specific” inflammation, an endeavour akin to “finding a black crow in a dark night”.

REFERENCES