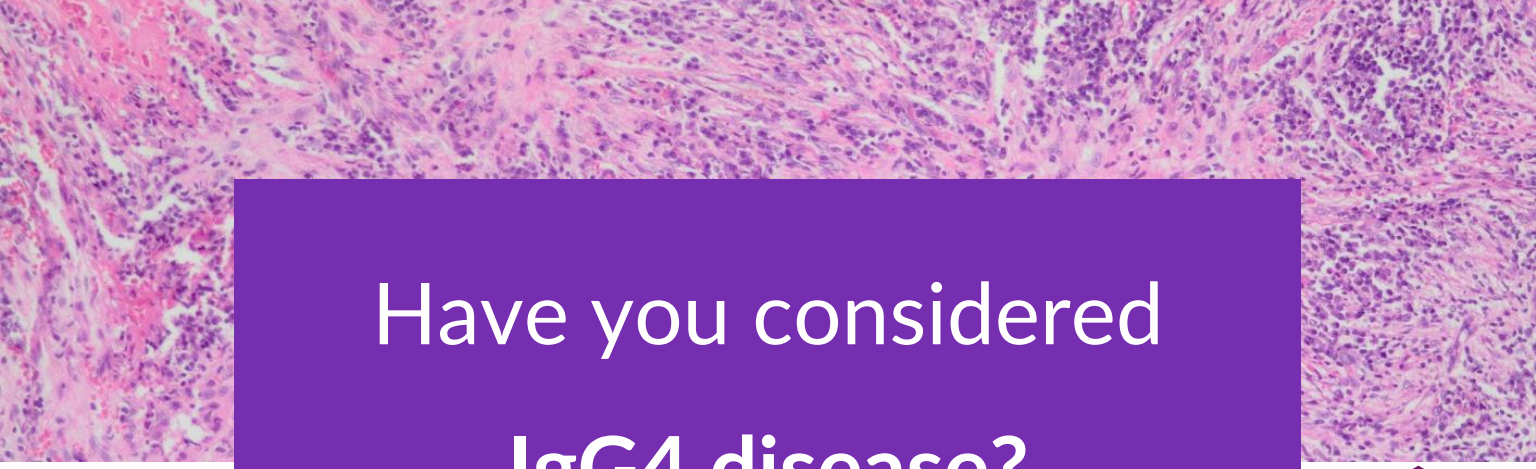
Have you considered IgG4 disease?



100X Low power magnification showing large areas of **storiform fibrosis** with a dense lymphoplasmacytic infiltrate

A Unique Presentation of IgG4 Disease with Ocular, Neurologic, and Mastoid Involvement

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A man in his twenties presented with headache and acute deterioration in visual acuity. Tinnitus and hearing loss followed. He was found to have panuveitis and raised intracranial pressure with associated papilloedema.

BACKGROUND

First described in 2001, immunoglobulin G4 (IgG4) disease, is a **multisystem inflammatory disorder** which typically presents as pancreatitis, retroperitoneal fibrosis, sclerosing cholangitis or interstitial lung disease.

- The central nervous system is rarely affected, with manifestations limited to pachymeningitis, hypophysitis and cranial nerve involvement
- This is the **first description of IgG4 disease presenting as panuveitis with associated mastoiditis and raised intracranial pressure**
- Two cases of IgG4 disease presenting with mastoiditis have been described, and both were associated with CNS involvement.

INVESTIGATIONS

- **MRI**: mastoiditis, thickened optic nerves.
- **Lumbar puncture**: raised opening pressure (>50 cm H_20), normal chemistry and cell count
- **Audiogram**: right sided moderately-severe to profound mixed hearing loss, and a left mild conductive hearing loss
- **Ophthalmology:** non-granulomatous panuveitis and papilloedema with retinal haemorrhages
- **Serology:** negative for infections, autoimmune diseases and sarcoidosis
- **CT chest**: no evidence of sarcoidosis
- Further investigations:
  - **FDG PET:** right mastoiditis
  - **Mastoid biopsy** (see histology).

DIFFERENTIAL

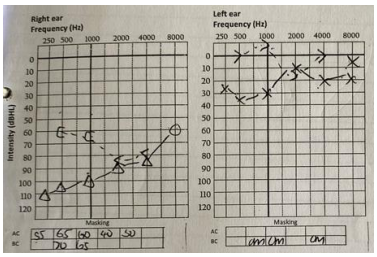
- **Idiopathic intracranial hypertension**
- **Mastoiditis and panuveitis:** infective, inflammatory, and non-benign causes of a systemic disorder with a diffuse meningeal process (TB, HIV, syphilis, sarcoidosis, autoimmune, malignancy etc.).

HISTOLOGY

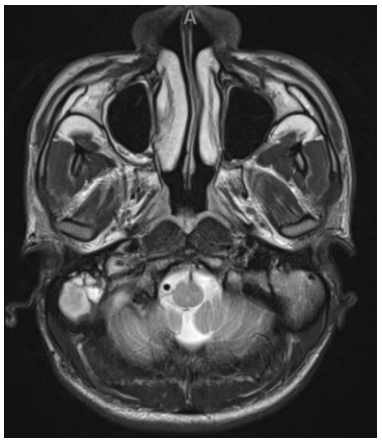
- Lymphoplasmacytic infiltration
- Raised plasma cells (IgG4+/IgG+ ratio >40%)
- Prominent storiform (cartwheel) pattern of fibrosis
- Serum IgG4 levels raised: 2.66 g/L (0.03-2.01).

MANAGEMENT AND OUTCOME

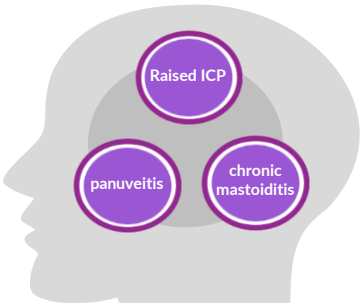
Symptoms improved with a combination of corticosteroids and long-term azathioprine. No progression of the disease.



Audiogram showing right sided moderately-severe to profound mixed hearing loss, and a left mild conductive hearing loss



Hyperintensity compatible with inflammation in the right mastoid on T2 sequence MRI brain



This clinical constellation has not been described in IgG4 disease, and the more commonly described systemic manifestations were not present in this patient

Histopathological diagnosis in this case was pivotal

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