

Diagnostic And Therapeutic Challenges in IgG4-Related disease in the Sphenoid Sinus

Omar Abu Suliman, MBBS, SB-ORL

Senior Registrar, otolaryngology Head & neck surgery

King Abdullah Medical City,

Makkah, Saudi Arabia

بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ





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Introduction

IgG4-related disease (IgG4-RD) is a newly recognized fibroinflammatory condition characterized by tumefactive lesions, a dense lymphoplasmacytic infiltrate rich in IgG4-positive plasma cells, storiform fibrosis, and often but not always, elevated serum IgG4 concentrations.

- **The disease was not recognized as a systemic condition until 2003, when extrapancreatic manifestations were identified in patients with autoimmune pancreatitis.**
- **Autoimmune pancreatitis had been linked to elevated serum IgG4 concentrations as early as 2001.**

- **IgG4-RD involves multiple organs, particularly exocrine organs.**
- **When it affects the head and neck, it commonly involves the salivary, lachrymal, and pituitary glands.**

- **It's quite rare to find such a lesion in the paranasal sinuses.**
- **We present a case of IgG4-RD in the sphenoid sinus.**

Case Presentation

A 38 years old female 3 months post delivery who started to have headache since 42nd day of post-delivery.

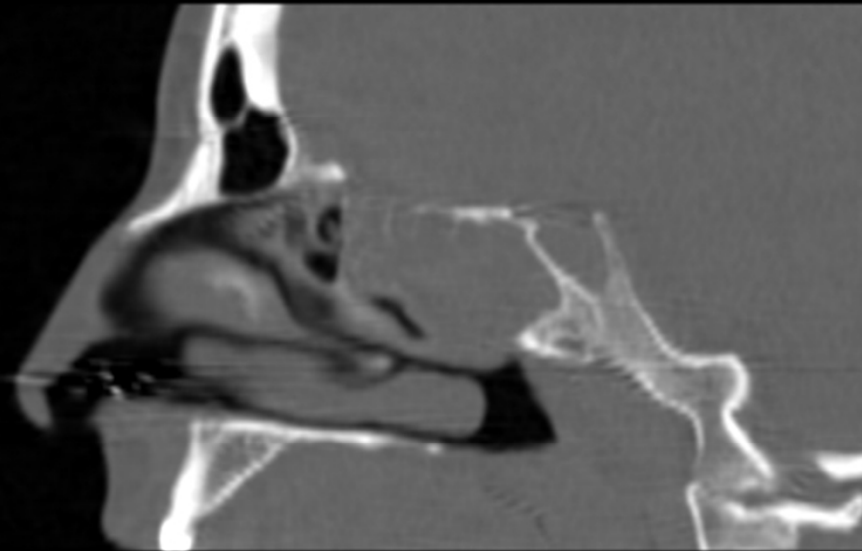
- **The headache was progressive and unilateral involving the whole left half of the head and resistant to analgesics.**
- **She was seen by neurologist who diagnosed her as a case of migraine and started anti migraine treatment.**

- **After getting anti migraine treatment for two weeks she experienced no benefit and headache became worse.**
- **Computed tomography (CT) brain scan done for her that showed normal brain study, but showed opacity in left sphenoid sinus.**
- **She approached the ENT clinic for further consultation where her examination was unremarkable and CT paranasal sinuses (PNS) was requested for her.**

- **One day later patient developed blurry vision and left 6th cranial nerve(CN).**
- **She was admitted as a case of complicated sphenoditis and intravenous antibiotics, i.e., ceftriaxone and vancomycin were started.**
- **CT PNS Done**

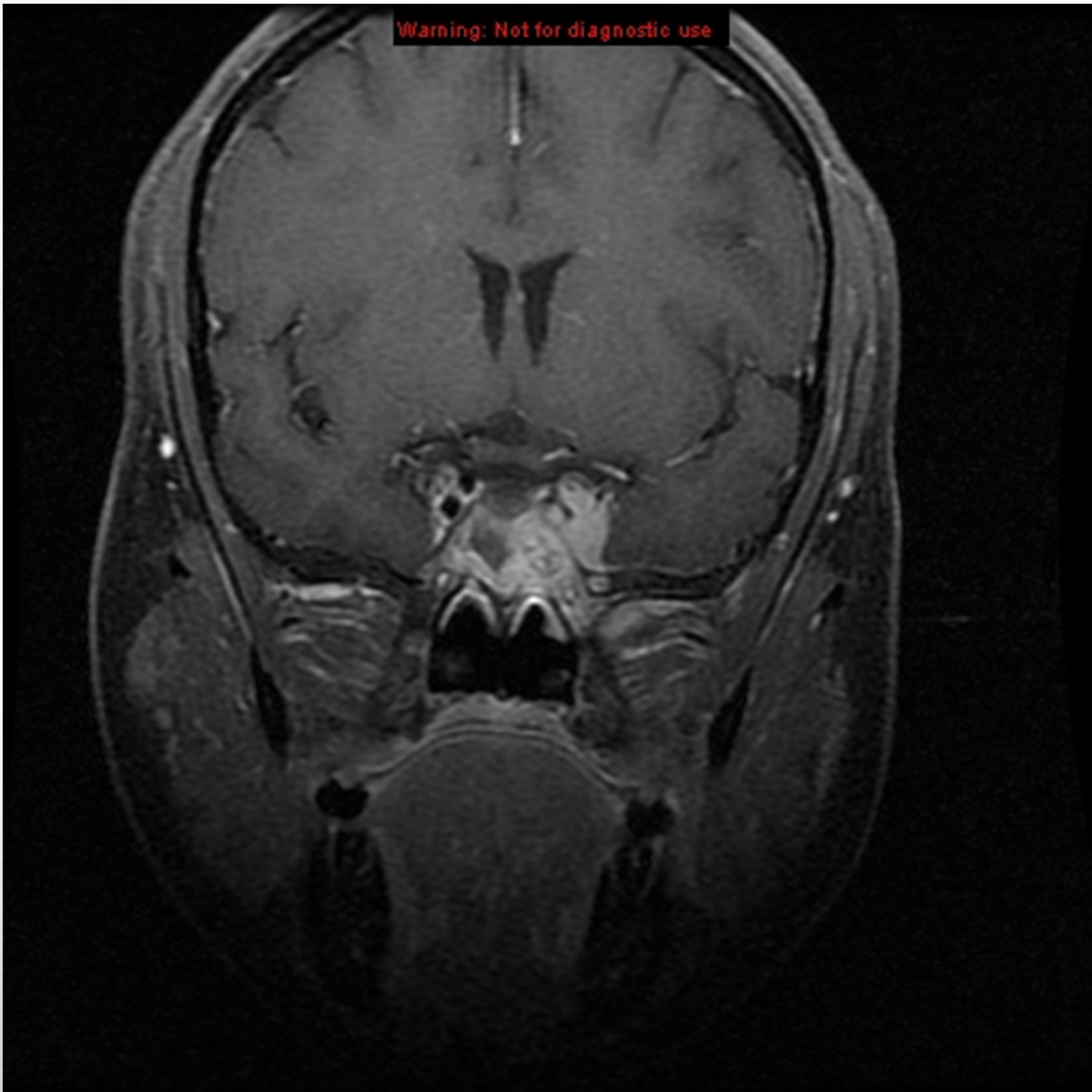
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○ **Magnetic resonance imaging (MRI) PNS done**

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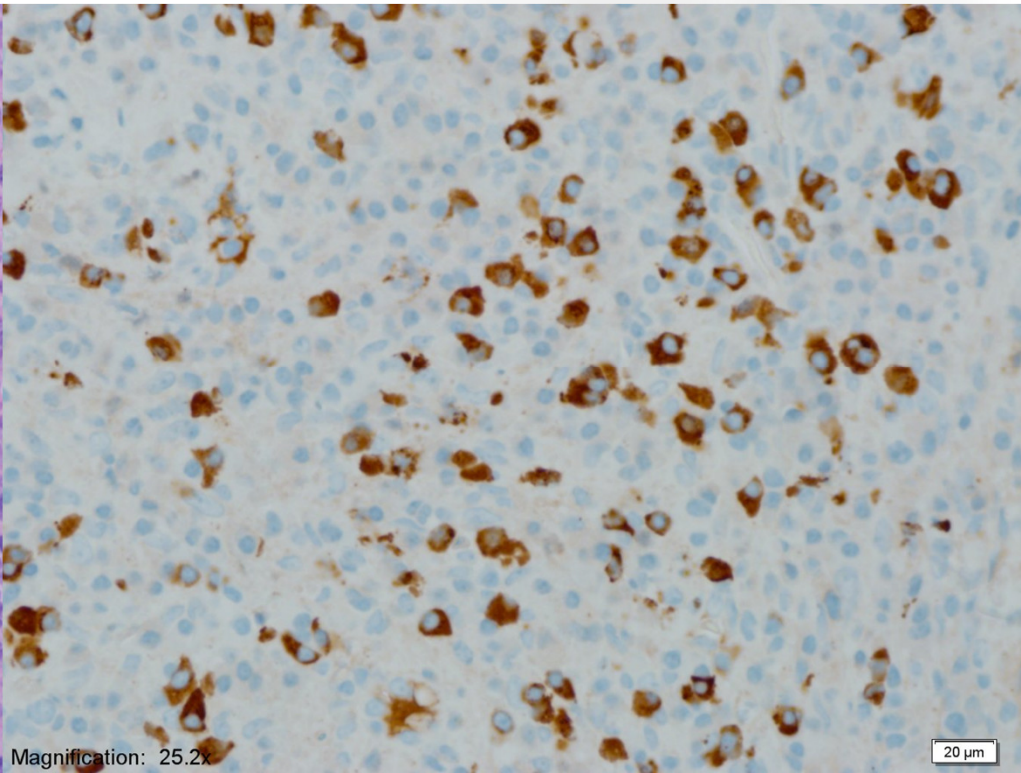
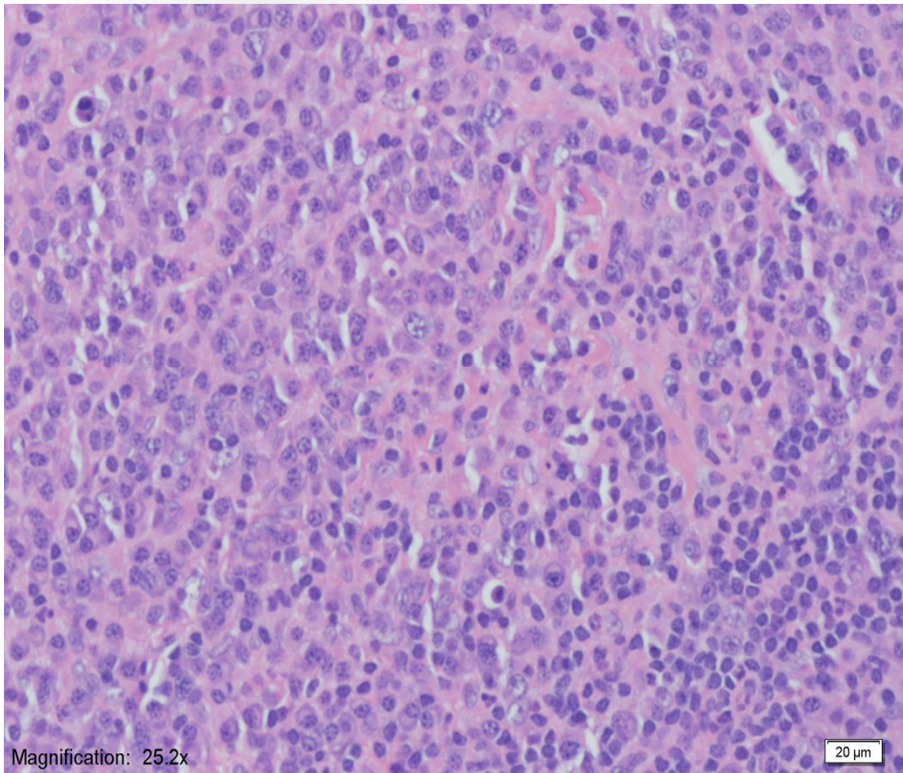
- **Functional endoscopic sinus surgery (FESS) was done under general anesthesia.**

Intraoperative, gritty whitish hard mass with minimal bleeding was noticed occupying the left sphenoid sinus extending to right sphenoid.

- **Mass was excised partially and sent for histopathology.**
- **Histopathology report showed non specific inflammation.**

- **The patient had uneventful post-operative period and was discharged.**
- **But 1 week later she presented with difficulty in left eye movement which revealed 3rd& 4th CN palsy in addition of 6th CN.**
- **Another biopsy was taken from deeper tissue.**

- **The Slides showed in Haematoxylin and Eosin (H&E) stain a moderately dense lymphoplasmacytic infiltration accompanied with fibrosis.**
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- **Immunostaining for IgG4 show significant increase in IgG4-positive plasma cells with IgG4-IgG ratio > 40% and >100 IgG4 positive plasma cells per High Field Focus (HPF).**



- **IgG4 serum level was elevated 0.53g/l that gave the diagnosis of IgG4-RD.**
- **Patient was started on steroid therapy dexamethasone 4mg IV(divided t.i.d for 3 weeks) then tapered over two weeks and then stopped.**
- **Afterwards, normal saline nasal irrigation and nasal steroid spray were started.**

Patient markedly improved with restored eye movement and vision, with no more headache or pain and was followed regularly for 9 months with no relapse.

Discussion

In our case the histological diagnosis is the main stay of diagnosing the disease.

- **Unawareness of such disease can lead to delay diagnosis.**

The diagnosis is made by histological and immunohistological findings.

- **There are varying criteria for the diagnosis of IgG4-RD; however, increased numbers of IgG4 plasma cells are required for the diagnosis and in cases with 50 IgG4 cells/HPF the reported specificity and sensitivity are 100%.**
- **The serum IgG4 titer can also be used to aid in diagnosis but is elevated in only 30% of patients with IgG4-RD and therefore not necessary for diagnosis.**

- **Glucocorticoids are typically the first line of therapy.**

A consensus statement from 17 referral centers in Japan suggested treating patients initially with prednisolone at a dose of 0.6 mg per kilogram of body weight per day for 2 to 4 weeks.

- **The authors suggested further that the prednisolone be tapered over a period of 3 to 6 months to 5.0 mg per day, and then continued at a dose between 2.5 and 5.0 mg per day for up to 3 years.**

- **Another approach has been to discontinue glucocorticoids entirely within 3 months.**

Glucocorticoids appear to be effective but disease flares are common.

Azathioprine, and methotrexate are used frequently as glucocorticoid-sparing agents or remission-maintenance drugs after glucocorticoid-induced remissions, but their efficacy has never been tested in clinical trials.

- **Jeremiah *et al.* reported the use of nasal steroid spray after sphenoidotomy with promising results.**

In our case we used nasal steroid spray along with normal saline nasal irrigation, which was efficient with marked results.

- **The aim of surgery is now mainly devoted to achieve a diagnostic biopsy and an adequate airway passage to pass local steroid.**

Conclusion

Although, IgG4-related disease of the sphenoid is extremely rare condition that rising diagnostic and therapeutic challenges.

It should be considered in the differential diagnosis of isolated sphenoid lesion with bone destruction.

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