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

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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 42\textsuperscript{nd} 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
Magnetic resonance imaging (MRI) PNS done
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.

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.
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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