

IgG4 in the head and neck- the great mimicker

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Introduction

Immunoglobulin G4-related disease is a chronic fibro – inflammatory condition characterized by enlargement of the affected organs, elevated serum

Immunoglobulin (Ig)-G4 levels, and abundant IgG4-positive plasma cell infiltration in the affected organs

Its spectrum of manifestations in the head and neck region remains relatively unrecognized in our country, thus presenting as a clinical as well as a radiological diagnostic dilemma.

The clinical and radiological manifestations can resemble neoplastic and other non-neoplastic pathologies in the head and neck region, hence a high index of suspicion for this disease entity can lead to an early diagnosis, initiate appropriate therapy and reduced healthcare costs.

Aims and objectives

To discuss the radiological manifestations of IgG4 related disease in the head and neck.

To highlight the importance of early and accurate diagnosis of IgG4 disease this is known to mimic malignancies in the head and neck.

Materials and methods

A single institutional retrospective review of cross-sectional imaging (Computed

Tomography and Magnetic Resonance Imaging) stored in the Picture Archiving and Communication System

Inclusion Criteria

Tumefactive lesions with imaging characteristics suggestive of neoplastic and other non-neoplastic pathologies without sufficient clinical / endoscopic corroboration.

A retrospective confirmed pathological diagnosis of Ig G4 disease was available.

Exclusion Criteria

Biopsy proven cases of malignant head and neck lesions. Biopsy proven cases of known granulomatous (tuberculosis / sarcoidosis) lesions in the head and neck.

Results

Nine cases were selected based on our inclusion criteria.

The following findings were obtained

Six of the patients selected were female and three were male.

All patients presented predominantly with ENT manifestations

1. Rhinitis and nasal blockage
2. Ear pain
3. Neck swelling
4. Periorbital swelling
5. Orbital symptoms i.e., vision loss, blurring of vision.

Results

The radiological findings included

Primary anatomical region		Frequency
1	Orbit and periorbital tissue	2
2	Skull base	2
3	Nasopharynx	2
4	Salivary glands	1
5	Pachymeninges	1
6	Miscellaneous (soft tissue)	1

- Computed Tomography (CT): Enlargement of the involved organ with diffuse hypoattenuation and homogenous enhancement.
- Magnetic Resonance Imaging (MRI): Low signal intensity on T1/T2/STIR Weighted imaging showing post-contrast homogenous enhancement. Lesions show restricted diffusion with low ADC values.
- Associated homogeneously enhancing cervical lymphadenopathy was seen in five out of nine cases.

Differential diagnosis		Frequency
1	Infective etiology	2
2	Low grade neoplasm	1
3	Sarcoidosis	1
4)	Lymphoma	1
5	Miscellaneous (orbital pseudotumor)	1

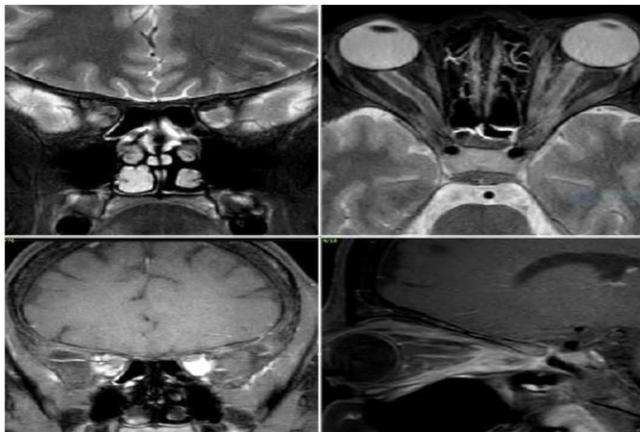


Figure 1: Orbit

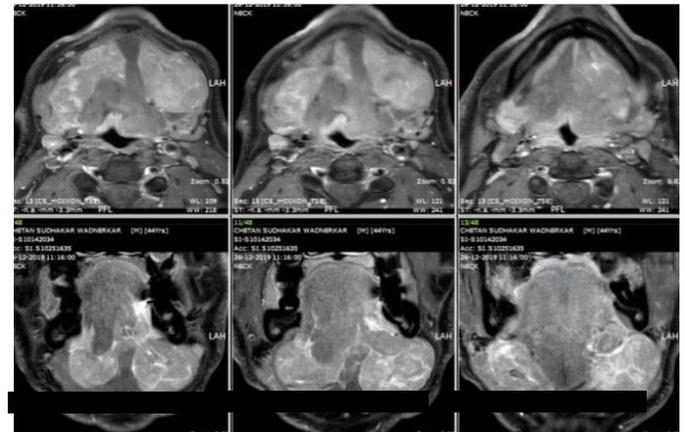


Figure 2: Sublingual Glands

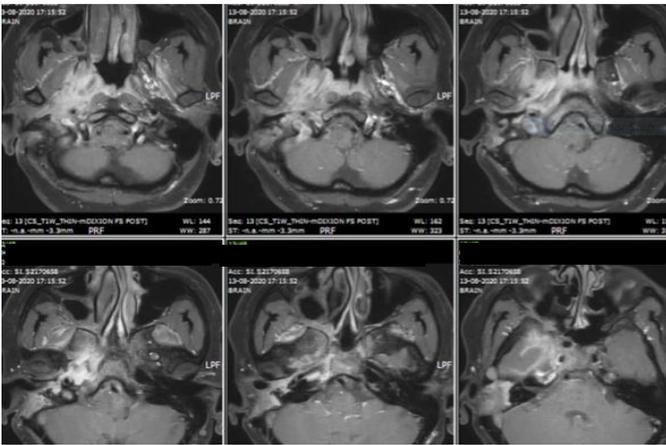


Figure 3: Lateral skull base/Temporal bone/MCF

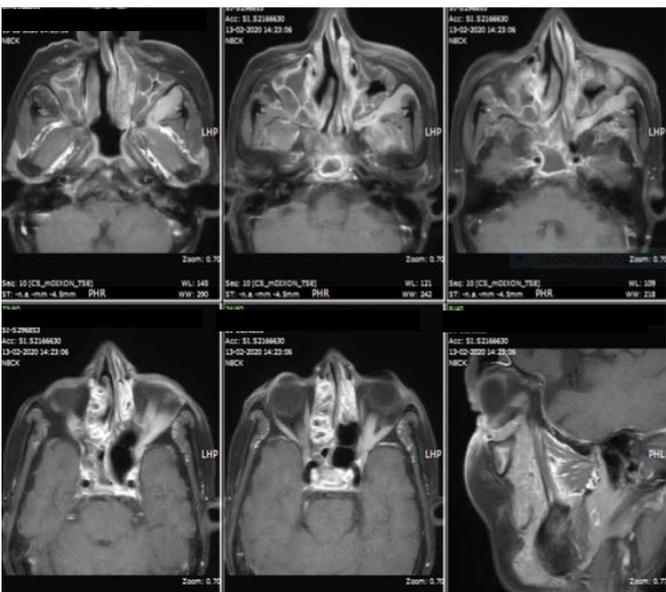


Figure 4: Masticator Space/ITF

Results

A confirmed pathological diagnosis was available in two cases which confirmed the classical storiform fibrosis (Basket weave pattern) of Ig G4 disease showing a dense lymphocytic infiltrate. These cases also show an elevated serum IgG4 plasma level.

Five of the nine cases showed no histopathological signs of Ig G4 disease however showed a raised serum Ig G4 level.

No histopathological or serological investigations were available for two patients.

All nine patients showed a marked resolution of symptoms post treatment with corticosteroids. Post treatment imaging was available in three patients which showed a significant reduction in disease process as compared to pretreatment imaging.

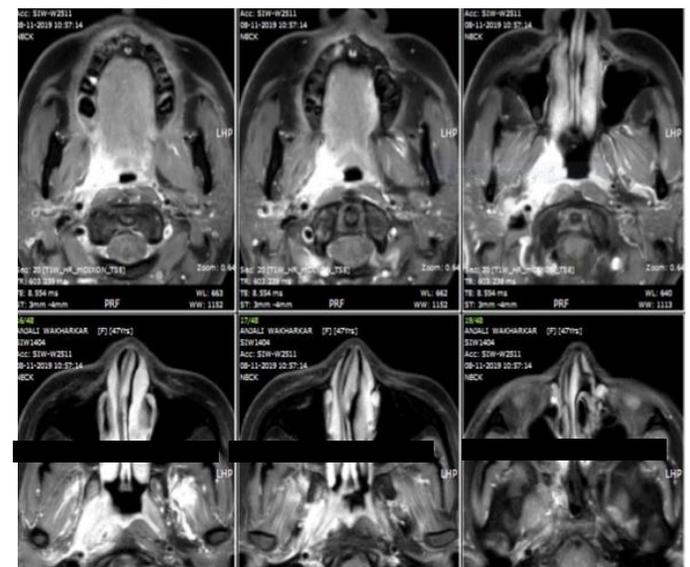
Discussion

IgG4 related disease is a well-known chronic fibro inflammatory condition with a tendency to form Tumefactive lesions in the affected organs.

A variety of head and neck inflammatory syndromes are now known to be associated with the Ig G4 related disease spectrum which include Mikulicz syndrome, Inflammatory pseudotumor of orbit, Kuttner tumor and Hashimoto's/Riedel's thyroiditis.

Diagnosis frequently involves biopsy of the lesion which remains the gold standard. Serologic analysis where elevated Serum IgG4 levels (30-40 times of normal upper limit) or Ig G4/Ig G ratio >40% can aid in the diagnosis.

Several radiological findings can help to raise the possibility of Ig G4 related disease.



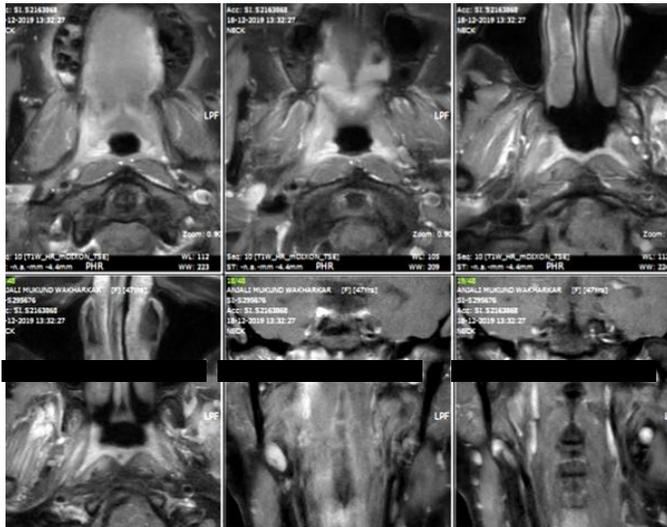


Figure 5: Nasopharynx Pre and post steroid therapy

Conclusion

It should be considered as a differential in a clinical setting of cases presenting with diffuse organ involvement in the head and neck and needs appropriate laboratory confirmation.

- Prompt and significant resolution of the disease following corticosteroid therapy can serve as a source of immediate relief to the patient and thus prevent any further invasive investigations.

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