Non-IgG4 Related Sclerosing Disease of the Maxillary Sinus: A Case Report

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Background

- Sclerosing disease of the sinuses is an exceedingly rare presentation of IgG4-related sclerosing diseases (RSDs), such as the more common IgG4+ autoimmune pancreatitis. It is characterized by benign, locally destructive, fibroinflammatory, and tumefactive lesions. [1]
- 2nd most common location of IgG4-RSDs is in the head and neck, however, the vast majority are located in the salivary, lacrimal, and pituitary glands. [1]
- Presentation is non-specific and may mimic symptoms of chronic rhinosinusitis such as: nasal obstruction/congestion, facial pain/swelling, headache, and epistaxis. [2]
- Consequently, delayed treatment can permit erosion of neighboring structures leading to neurological symptoms.
- Prognosis is good with treatment

Purpose: Expand upon the current, limited knowledge regarding sclerosing sinonasal disease and the importance of considering this rare disease in your differential.

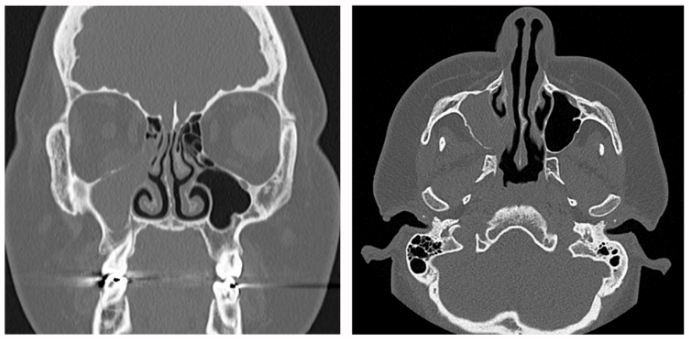

Fig. 1: CT Sinus coronal (left) and axial (right) views. Right maxillary sinus mass measuring approx. 1.8 x 3 x 3.9 cm. Evidence of bony erosion involving right pterygopalatine fossa, medial right ethmoidal air cell, and orbital floor involving inferior rectus muscle.

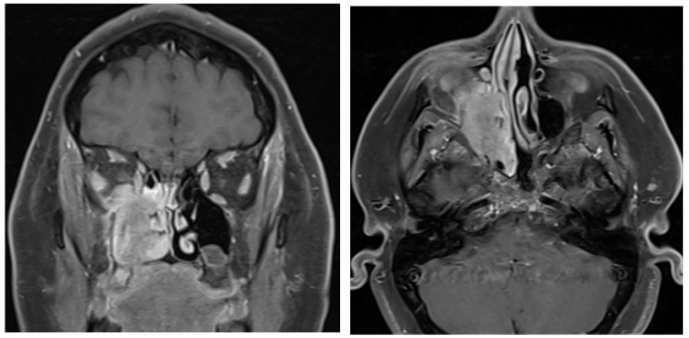

Fig. 2: MRI Orbits coronal (left) and axial (right) views. Confirmed findings. No extension of mass.

Discussion

- Formerly, sinonasal involvement was reported more frequently in males, >50 years old. More recent systematic reviews found the male:female ratio is closer to 1:1 with an average age of 47-49 years old,[1, 2, 5] more similar to the demographic of our patient.
- **Diagnosis:** Based on clinical presentation, imaging, and histopathological examination (gold standard). Criteria highly suggestive of IgG4-RSDs include:
  - Elevated ratio >40% IgG4+/IgG+ tissue plasma cells
  - Major histopathological features: dense lymphoplasmacytic infiltrate, fibrosis in a characteristic "storiform" pattern, and obliterative phlebitis. [3, 4, 5]
  - Despite our patient having a ratio of <40% IgG4+/IgG+ tissue plasma cells, the morphological features were highly suggestive of an IgG4-RSD and treated as such with efficacy.
- **Treatment:** Similar to other IgG4-RSDs.[5] High-dose oral corticosteroids with surgical debulking as needed. Antibiotics discontinued after negative cultures

Case Report

- 31-year-old female presented with 2-week history of right eye pain and headache. Denied any chronic sinus complaints or other significant PMH. Physical exam significant for diplopia on upward gaze and right hypertropia.
- CT sinus (fig. 1) revealed a right maxillary mass with bony erosion involving the pterygopalatine fossa, ethmoid air cells, and the orbital floor, involving the inferior rectus muscle. MRI of the orbits (fig. 2) confirmed the findings and found no extension of the mass.
- Empiric antibiotics and oral steroids initiated. Endoscopic sinus surgery performed for debulking and biopsy. Pathology revealed an inflamed, sclerosing lesion with fibrillar collagen consistent with the morphology of an IgG4-RSD. Less than a 10% ratio IgG+/IgG4+ tissue plasma cells were found which was less consistent with the disease. Serum IgG4 levels normal.
- Other studies: ALK-1 negative. Flow cytometry was non-contributory. Aerobic, anaerobic, AFB, fungal cultures all negative.

Conclusion

- After treatment, the patient exhibited significant improvement of her symptoms; however, still had residual diplopia.
- Our case did not match classic diagnostic criteria for IgG4-RSDs, but many features were suggestive of one. Perhaps high IgG4 levels are less important to sinonasal presenting sclerosing disease and thus deviate from typical criteria of IgG4-RSDs.
- Despite its rarity, greater awareness of sinonasal sclerosing disease is warranted to prevent advanced disease.

References

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