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IgG-4 related disease in Maxillofacial Region- A Rare case report

[1]Vivek Saxena, Senior Specialist, Oral and Maxillofacial Surgery, Army Dental Centre (R&R), New Delhi-110010, India.

[2]Narendra Babu BK, Classified Specialist, Army Dental Centre (R&R), New Delhi-110010, India.

[3]DK Gupta, Professor, Department of ENT & HNS, Army Hospital Research & Referral, New Delhi-110010, India.

[4]Akash Sehgal, Resident, Division of Oral and Maxillofacial Surgery, Army Dental Centre (R&R), New Delhi-110010, India.

Corresponding Author: Akash Sehgal, Resident, Division of Oral and Maxillofacial Surgery, Army Dental Centre (R&R), New Delhi-110010, India.

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Abstract

IgG-4 related disease is a chronic immune mediated disease that has fibro-inflammatory component affecting a variety of organ systems/or organs producing tumor like effects. While background pancreatitis and bilateral salivary gland enlargement may be a clinical guide in diagnosing the disease, the specific diagnosis clinically involves histopathological (HPE) examination revealing storiform fibrosis, obliterative phlebitis and tissue infiltration with lympho plasmatic infiltrates. We present a case of 74 yrs. old male patient with left pre-auricular swelling and pus discharge in whom initial diagnosis of left mandibular chronic suppurative osteomyelitis was made. After HPE examination, patient was diagnosed as a case of rare IgG4-related disease. Such lesions present a diagnostic challenge, but the outcome is very favorable. Corticosteroids remain the mainstay of the treatment along with surgical debridement of affected tissue.

Keywords: Corticosteroids, Immunohistochemistry lymphoplasmatic infiltrates, PMMC flap, Storiform fibrosis.

Case Report

A 74-year-old male patient reported to the Department of Oral and Maxillofacial Surgery, a tertiary care maxillofacial facility, at Army Dental Centre (Research & Referral), New Delhi. The patient presented with swelling and pus discharge from multiple sites in the left pre-auricular region (Figure-1) extending till the left angle of the mandible since three months from the day of reporting along with restricted mouth opening from the last one month.

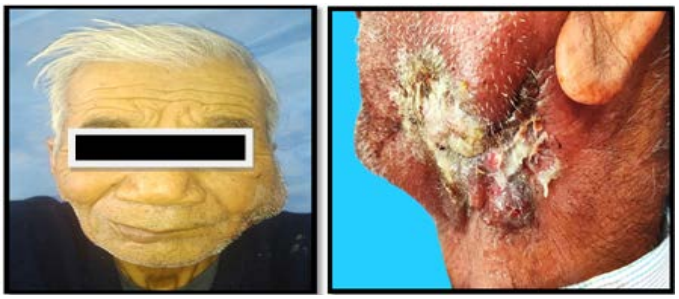

Figure 1

On general physical examination, the patient was found to be lean built, malnourished and underweight but was calm, conscious, coherent and well oriented to time place and person. A detailed clinical history was elicited and revealed multiple clinical interventions for various complain as mentioned in the Table-1.

Table 1

Time line	Diagnosis	Intervention	Inference
December 2017	Chronic cheek bite left buccal mucosa	Extraction w.r.t tooth no 38	Swelling 10-12 days post tooth extraction
January 2018	Left buccal space infection	Symptomatic treatment with antibiotics and antioxidants	Brief period of disease remission
	Underwent knife biopsy w.r.t Left buccal mucosa and diagnosed as Oral pemphigoid	No treatment was instituted	Patient did not follow up.
January 2019	Masseter hypertrophy	Underwent masseter debulking	Histopathological examination (HPE) revealed only chronic non-specific inflammatory tissue with giant cell reaction
June 2019	Pleural effusion and Pulmonary Tuberculosis	Anti-Tubercular Therapy was instituted for 09 months	Recovered completely
March 2020	Chronic suppurative osteomyelitis of mandible (Left side)	Sequestrectomy and debridement	Histopathological examination (HPE) revealed only chronic non-specific inflammatory tissue

The personal history revealed Khaini consumption for the last 45 years which was cessated only two years back. On specific maxillofacial examination, one could appreciate a gross facial asymmetry on the Left side of the face with the maximum inter-incisal mouth opening being only 5mm. The middle and lower one third of the face (left side) showed multiple draining sinuses with minimal erythema and pus discharge along with caseous material oozing out of multiple sites. Intraoral examination revealed no abnormality in the buccal mucosa. The NCCT face in axial, coronal, sagittal planes was done along with 3-Dimensional reconstruction of the images. Pus was sent for culture and sensitivity examination. The NCCT showed severe osteolysis of the left side mandible along with maxilla and swelling of the adjacent tissue as is appreciated in the soft tissue window (Figure-2 & Figure-3).

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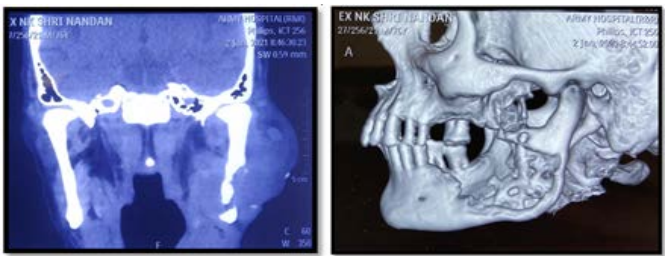

Figure 2 & 3

The patient was preliminarily diagnosed as a case of chronic suppurative osteomyelitis Left side mandible and was taken up for wide local excision (WLE) of the soft tissue, segmental resection of the mandible without disarticulation and reconstruction plate fixation along with final closure with PMMC Flap (Pectoralis Major my ocutaneous). (Figure 4,5,6 ,7&8).

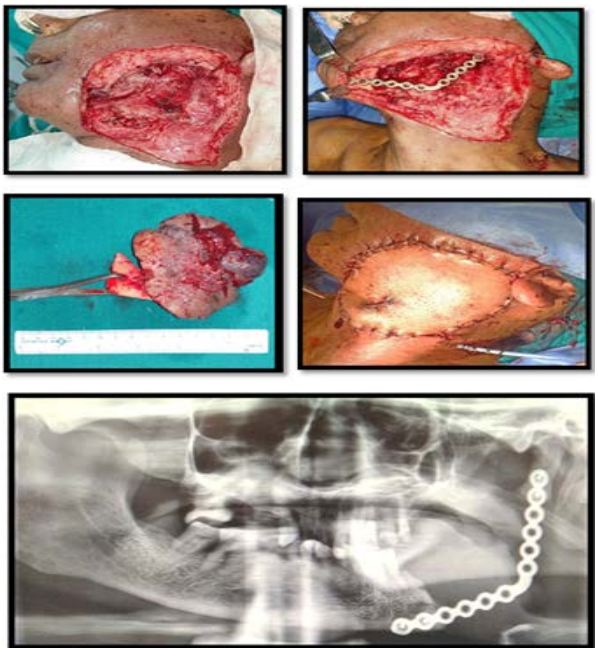

Figure 4,5,6,7&8

The patient was kept on periodic follow up and presented with cheesy discharge again from the superior aspect of the PMMC flap two months after the surgery along with reduction in mouth opening. MDR-TB (due to history of tuberculosis) test was done which was negative while Bone Scan findings corroborated with SPECT suggestive of ongoing inflammation wrt left mandible predominantly at the anterior aspect of reconstruction plate along with surrounding soft tissue inflammation. (Figure9 &10).

Figure 9&10

Re-exploration of the operated site and aggressive debridement was carried out till pterygoid plates thus removing all hard fibrous and rubbery tissue. Caseous material was found oozing from many planes which was all debrided, along with removal of the reconstruction plate and closure with existing PMMC flap. (Figure 11,12&13).

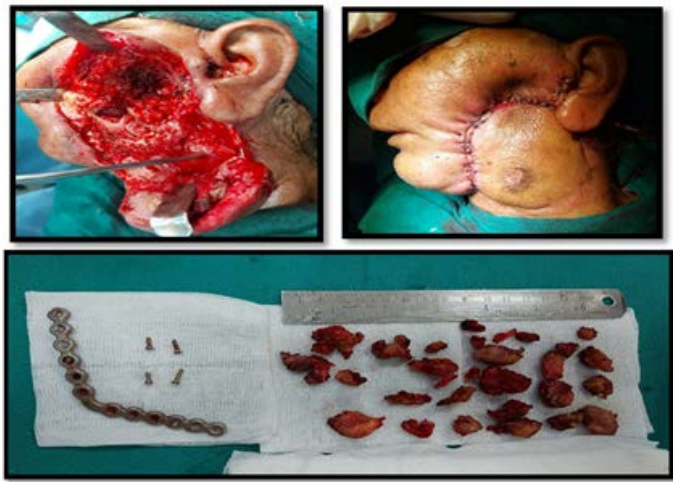

Figure 11, 12&13

The biopsy specimen including fibrous tissue and the left Parotid gland was sent for HPE which revealed keloid like storiform fibrosis and plasma cell rich infiltration indicating towards a rare IgG4-RD (Related disease) which was a rare entity in the maxillofacial region. Corticosteroids were started at the dose of 0.6mg/Kg/Day for two weeks followed by sequential tapering. The culture and sensitivity also revealed presence of Pseudomonas aeruginosa sensitive to

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Colistin antibiotic which was then started at the dose of 2.5mg/kg under constant monitoring of renal functions. The patient has been on regular follow up for a period of 1 year with no untoward complaints now. The surgical site has also completely healed uneventfully. (Figure-14,15,16&17).

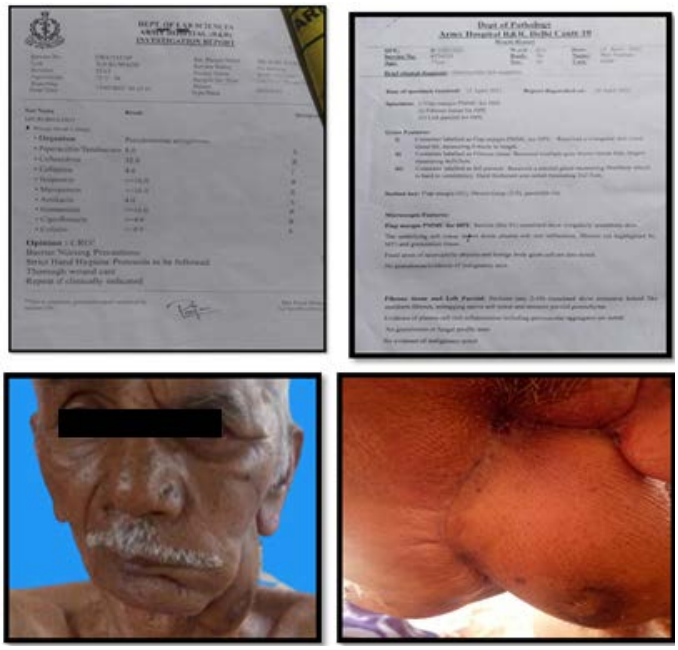

Figure-14,15,16&17

Discussion

Immunoglobulin IgG4-related disease is a multifocal systemic immune-mediated disorder characterized by a fibrosclerotic inflammatory pattern, which may affect multiple organs, most commonly presenting as a tumor or "masslike", and in smaller number, presenting infiltrative characteristic(1). The pathogenesis of the disease is still poorly understood, with autoimmune and infectious agents being considered as potential immunological triggers (2). The symptoms of IgG4-RD vary considerably depending on the organ(s) or tissues involved. The most common presentation is a mass lesion or organ enlargement. The most commonly affected organs are salivary and lacrimal glands, pancreas and biliary tract, and the kidneys; but any organ could be involved and multiorgan disease can also be seen (3). IgG4 is the least abundant of the IgG subclasses, accounting for less than 6% of the total IgG in the normal population. In bronchial asthma, Ige participates in the reaction to allergens, whereas IgG4 becomes prominent only late during chronic antigenic stimulation. Therefore, IgG4 producing plasma cells might behave as memory cells in allergic patients (4). The age-group of hyper IgG4 disease patients is typically between 50 and 70 years with men affected 2 to 3 times more commonly than women (5). Hamano et al (6) found only 20% of their IgG4-related AIP (Acute Intermittent Porphyria) patients had raised serum IgE levels; hence, it was unlikely for an exogenous antigen to be responsible for the rise in IgG4. They also found that the patients with sclerosing pancreatitis had elevated serum concentrations of immune complexes containing IgG4, and the concentrations decreased with steroid therapy, suggesting that the immune complexes are closely related to the pathogenesis of this disease. Kitagawa et al (7) in a review of CSS speculated that an increased IgG4 response to antigenic materials around ducts of salivary glands and pancreas may play a key step in the pathogenesis. It is important to remember that elevated plasma IgG4 levels are neither specific nor sensitive for IgG4 RD. Normal IgG4 can be seen in nearly one-fourth of the cases, and using a cut-off level for IgG4 of 135 mg/ dl has a positive predictive value for IGG4-RD of only 34% (8). The diagnosis of IgG4-RD can be challenging and the approach to diagnosis will depend on the site affected. A thorough process should always be followed to rule out the different diseases that can mimic IgG4-RD. These diseases include a broad spectrum of conditions including malignancy, lymphoproliferative disorders, Antineutrophil cytoplasmic antibodies (ANCA)-associated vasculitis, sarcoidosis, Sjogren's syndrome,

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Castleman's disease and others (9). The assessment should start with complete history and physical examination. Then when the disease is suspected, laboratory testing and appropriate radiology evaluation will depend on the site involved. Plasma IgG4 levels should be obtained in all patients (10). In the reported clinical case, the Patient presented with swelling and pus discharge. After debridement, the biopsy specimen including fibrous tissue and the left Parotid gland was sent for HPE which revealed keloid like storiform fibrosis and plasma cell rich infiltration indicating towards a rare IgG4-RD (Related disease). the patient was then placed on steroid for two weeks followed by sequential tapering. There are still no definitive strategies for the treatment of IgG4-RD. But there are some guidelines that were suggested in 2015. Of these, it is advised, in asymptomatic patients with limited disease, for example, with only involvement of the lymph nodes or salivary glands, "observe and wait", keeping the patients closely monitored. In symptomatic patients, interventions are indicated, either through glucocorticoids and B-cell depletion to induce remission of symptoms, or through surgical interventions as per Khosroshahi et al.2015 (10).

Conclusion

IgG-4 RD can present as a daunting challenge especially as it overlaps with clinical presentations with many other pathologies. Accurate patient diagnosis followed by a meticulously planned medical and surgical intervention may help to bring down patient morbidity and uneventful outcome. Close working of surgical and pathology team is very essential for such cases and should be upheld in all such clinical scenarios.

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