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Images in Rheumatology

Gingival Involvement in IgG4-Related Disease

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A 50-year-old male presented to the rheumatology clinic for a 4-year history of recurrent gingivitis associated with worsening burning sensation of the tongue and hoarseness of voice. Written informed consent was obtained from the patient who agreed to take part in the study. On physical examination, he had gingival erythema predominantly in the upper gum with no oral ulcers or swelling of the tongue or lips (Figure 1A-B). Laboratory investigations included significantly elevated total serum IgG level of 19.9 g/L (6.03-16.13 g/L) and IgG4 level of 1.28 (0.02-0.96 g/L). He had a negative IF-ANA, RF, CCP, dsDNA, anticentromere (ACA), Scl-70, Jo-1, SSA, SSB, antibodies. Inflammatory markers included a CRP of 1.5 mg/L (<5 mg/L) and ESR of 69 mm/h (2-28 mm/h). Other invetigations included a normal vitamin D, copper, vitamin B1, B6, B12 levels, HSV 1 and 2 PCR, and low ACE level. Computerized tomography (CT) of the chest and upper abdomen and an ultrasound of the kidneys that were unremarakble. He underwent an

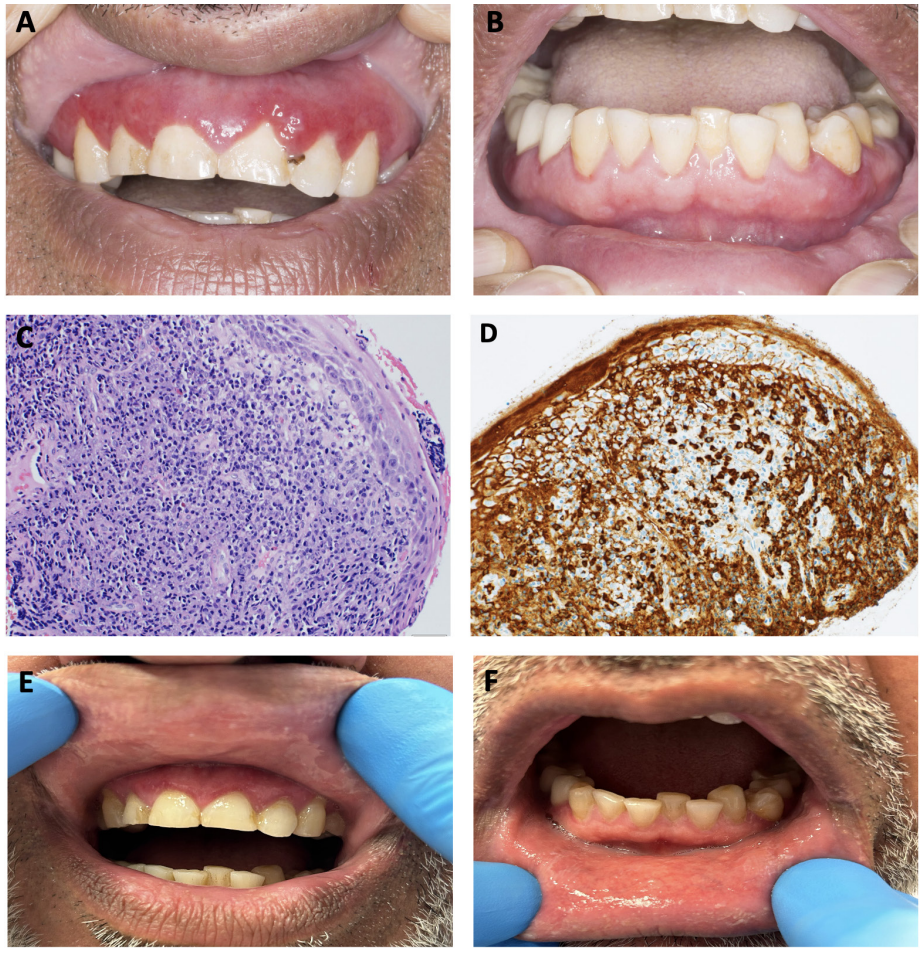

Figure 1. Figures A-B: Gingival erythema predominantly in the upper gum with no oral ulcers or swelling of the tongue or lips. Figure C: Hematoxylin and eosin (H&E) staining revealed oral squamous mucosal epithelium with a marked plasmacytic rich inflammatory infiltrate of the lamina propria and reactive hyperplasia of the overlying squamous epithelium with overall morphologic changes favor chronic gingivitis. Figure D: IgG4 immunohistochemical staining revealed increase in expression within plasma cells. Figure E-F: Improvement in gingival erythema after two years of immunosuppressive therapy.

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upper gingival biopsy that showed oral squamous mucosal epithelium with a marked plasmacytic rich inflammatory infiltrate of the lamina propria, reactive hyperplasia of the overlying squamous epithelium without signs of a subepithelial separation or eosinophils in the underlying infiltrate, or evidence of dysplasia and malignancy, particularly lymphoproliferative disease (Figure 1C). Therefore, pemphigus and pemphigoid seemed unlikely based on the biopsy findings. Additionally, sarcoidosis was felt to be unlikely due to the absence of non-caseating granulomas on biopsy, absence of hilar lymphadenopathy on CT imaging, and low angiotensin-converting enzyme (ACE) level. Additionally, immunohistochemical stain for IgG4 showed increase in expression within plasma cells (Figure 1D). With the impression of IgG4-related disease (IgG4-RD) involving the gingiva, he was commenced on azathioprine 100 mg daily. He noted progressive improvement in his physical examination and symptoms after 2 years of immunosuppressive therapy (Figure 1E-F).

Main Points

- Organ involvement in IgG4-related disease is heterogenous and should be considered as a differential for localized gingival involvement.
- Patients with localized involvement of the gingiva may benefit from corticosteroid-sparing therapy.

IgG4-related disease is a class of immune-mediated disorders with distinct pathological features, including dense lymphoplasmacytic infiltration, storiform fibrosis, obliterative phlebitis, and often elevated serum IgG4 levels.[1,2] In the craniomaxillofacial area, the occurrence of gingival involvement in IgG4-RDs is relatively rare, with only 5 cases reported in the literature with involvement of either the upper or lower gingiva.[3] Our case highlights a rare localized manifestation of IgG4-RD that received timely treatment that continues to have close follow up to observe for potential systemic involvement in the future.

Data Availability Statement: The data that support the findings of this study are available on request from the corresponding author.

Informed Consent: Written informed consent was obtained from the patient who agreed to take part in the study.

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