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IgG4-related Disease With Destructive Nasal Bone Involvement Leading to Saddle Nose Deformity

Giorgos Loizidis, MD, Department of Medicine, Division of Rheumatology; Mindy R. Rabinowitz, MD, Department of Otolaryngology – Head and Neck Surgery; Madalina Tuluc, MD, Department of Pathology, Anatomy and Cell Biology, Thomas Jefferson University, Philadelphia, Pennsylvania, USA. Address correspondence to Dr. G. Loizidis, Thomas Jefferson University Hospital, Division of Rheumatology, 211 S 9th Street, Philadelphia, PA 19107-5084, USA. Email: giorgos.loizidis@jefferson.edu. The authors declare no conflicts of interest relevant to this article. Ethics board approval is not required because this is a single case report and no intervention had been made for research. The patient gave written, informed consent to publish the material.

Sino-orbital disease from IgG4-related disease (IgG4-RD) has been described previously,[1,2] but nasal bridge collapse due to bone involvement has been rarely reported.[3,4]

A 34-year-old woman presented with right eye swelling. Computed tomography (CT) scan showed a posterior orbital mass extending to the nasal bones leading to saddle nose deformity (Figure 1). Sinus and nasal bone biopsy revealed concentric, storiform fibrosis (Figure 2A) and lymphoplasmacytic infiltrate. Immunohistochemistry for CD20, CD3, and in situ hybridization for kappa and lambda light chains showed a lymphoid infiltrate with positive immunostaining for IgG4, with an increased number of IgG4-positive cells, up to 25 per high-power field (HPF; Figure 2B). The biopsy had no evidence of malignancy, necrosis, giant cells, granulomas, or vasculitis. Tissue flow

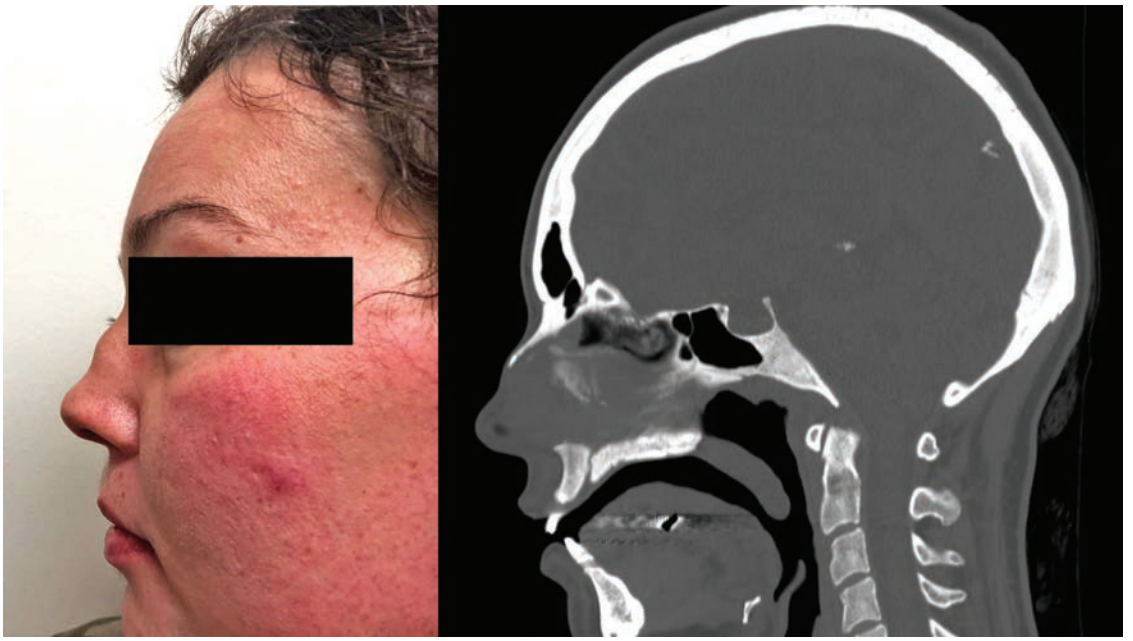

Figure 1. Saddle nose deformity with the corresponding sagittal CT scan view. CT: computed tomography.

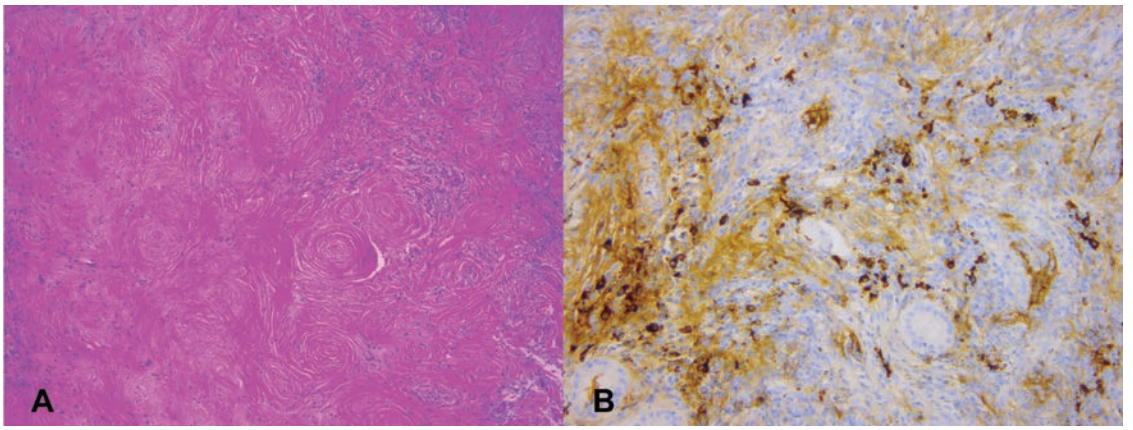

Figure 2. (A) Micrograph of the sinonasal mucosa biopsy showing concentric, storiform fibrosis, and (B) the immunostaining for IgG4 showing an increase in the number of IgG4-positive cells, up to 25 per high-power field.

cytometry and cerebrospinal fluid cytology were negative for malignancy. Peripheral IgG4 level was 271.3 mg/dL (normal range 4–86 mg/dL). Antineutrophil cytoplasmic antibody and myeloperoxidase and proteinase 3 were negative. Hemoglobin was 12.4 (normal range 12.5–15 g/dL), eosinophil count 0.18 (normal range 0.0–0.4 × 10^3/μL), and C-reactive protein was 0.4 (normal range < 0.8 mg/dL). CT scan of the abdomen and pelvis was negative for any major pathology. The ophthalmic disease reoccurred despite rituximab and glucocorticoids. Magnetic resonance imaging showed enlargement of the right orbital mass, requiring decompressive surgery. A repeat orbital and lacrimal bone biopsy showed extensive fibrosis and vascular sclerosis with lymphoplasmacytic infiltrate. Immunohistochemical stains showed lymphoid infiltrate (50% CD20+ PAX5+ B cells and 50% CD3+CD5+ T cells) with plasma cells (< 30/HPF), most of which (> 50%) expressed IgG4, consistent with partially treated IgG4-RD.

Clinical improvement was achieved with the addition of cyclophosphamide.

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