Immunoglobulin G4–Related Sclerosing Disease Mimicking Invasive Tumor in the Nasal Cavity and Paranasal Sinuses

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IgG4RSD involves multiple organs, particularly exocrine organs such as the salivary glands, pancreas, and biliary tracts.1 When it affects the head and neck, it commonly involves the salivary, lachrymal, and pituitary glands. There is insufficient radiologic data for IgG4RSD affecting the nasal cavity and paranasal sinuses.1–3 We present a case of IgG4RSD mimicking an invasive malignant tumor in the nasal cavity and maxillary sinuses on both sides.

Case Report
A 71-year-old man who had a brain contusion 25 years earlier complained of nasal occlusion and swelling of the right cheek. The patient had no further relevant medical history. A right maxillary tumor was suspected. His serum immunoglobulin level, including IgG4, was slightly elevated (114 mg/dL; normal range, 4.8–105 mg/dL; 7% of total serum immunoglobulin). The results of other biochemical tests were normal. MR imaging showed a mass in the nasal cavity and both maxillary sinuses, invading the right cheek subcutaneously along the right plexus dentalis superior, the left pterygopalatine fossa, and the left sphenoidal bone marrow (Fig 1). No osseous destruction was apparent. Masses of intermediate intensity and slight hyperintensity were observed on T1- and T2-weighted images, respectively (Fig 1A, -B), and an area of high T1-weighted signal intensity within the right maxillary sinus suggested hemorrhage (Fig 1A). Fat-suppressed T1-weighted imaging of the masses revealed homogeneous gadolinium contrast enhancement (Fig 1C).

We suspected malignant lymphoma, melanoma, or another malignant tumor with perineural and bone marrow invasion and biopsied the nasal mucosa. Hematoxylin-eosin staining revealed lymphoid follicles with a germinatal center among proliferative fibrosis, with lymphocyte and plasma cells without atypia (Fig 2A). Immunostaining revealed plasma cells expressing IgG4 (Fig 2B, -C). Other organ systems showed no abnormalities. Oral prednisolone therapy for 18 months (tapered to 10 mg/day from an initial dose of 40 mg/day) reduced the tumor volume to approximately one-third, and the tumor remained stable with time.

In conclusion, IgG4RSE may involve the nasal cavity and paranasal sinuses with perineural and bone marrow invasion, mimicking malignant lymphoma.

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