

IgG4-related disease with solitary nasopharynx involvement mimicking nasopharyngeal tumor

분당서울대병원 내과¹, 분당서울대병원 이비인후과²

유주리¹, 조성우², 강은하¹, 이윤종¹, 하유정¹

Background: Immunoglobulin G4-related disease (IgG4-RD) is an immune-mediated fibrotic disease that can affect multiple organs and form a tumor-like mass. It frequently involves exocrine glands, pancreas, and retroperitoneum and accompanies elevated serum IgG4 (sIgG4) levels. It is difficult to diagnose IgG4-RD of infrequently affected organs when sIgG4 level is within normal range. Here, we report a rare case of IgG4-RD with solitary nasopharyngeal involvement and normal sIgG4 level, mimicking nasopharyngeal tumor.

Case report: A 51-year-old man visited ENT department complaining of left otalgia and temporal area tingling sensation and soreness. MRI showed a 2.5cm-sized, mass-like infiltrative soft tissue lesion at the left nasopharynx and combined chronic otitis media in the left middle ear, suggesting nasopharyngeal cancer. The first nasopharyngeal biopsy demonstrated lymphoplasmacytic and neutrophilic infiltration with some focal IgG4 positive plasma cells (up to 20/HPF). The patient was transferred to rheumatology department for the evaluation of the systemic inflammatory diseases. Laboratory investigations showed a mild elevation of ESR (40 mm/hr) and CRP (0.6 mg/dL), negative anti-neutrophil cytoplasmic antibodies, and normal levels of serum IgG (1105 mg/dL [700-1700]) and IgG4 (81.9 mg/dL [3.9-86.4]). Chest and abdomen CT scans did not show systemic manifestation of IgG4-RD. For the differential diagnosis with other inflammatory conditions such as granulomatosis with polyangiitis, the second deep biopsy was performed, revealing fibrosis and dense lymphoplasmacytic and some eosinophilic infiltration with increased IgG4-positive cells (up to 80-90/HPF). We finally diagnosed him with IgG4-RD involving only the nasopharynx. After treatment with moderate-dose glucocorticoids (methylprednisolone 0.5mg/kg) and azathioprine, his related symptoms improved. Due to its rarity, nasal cavity involvement is not incorporated in 2019 ACR/EULAR classification criteria for IgG4-RD. When seeing patients with inflammatory mass-forming lesions in the nasal cavity, the possibility of IgG4-RD should not be overlooked and a deep biopsy might help make a diagnosis.

Figure. (A) PNS MRI showing infiltrative enhancing thickening of the left nasopharynx in the T1W-enhanced axial view. (B) Nasal endoscopic evaluation shows an irregular infiltrative lesion of the left nasopharynx. (C) Hematoxylin-eosin staining demonstrating fibrosis and dense lymphoplasmacytic and some eosinophilic infiltration (x 100). (D) Immunohistochemical staining for IgG4 showing abundant IgG4-positive plasma cells (80-90/HPF).

