A Case of IgG4-related Pharyngolaryngitis

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IgG4-related inflammatory diseases, including autoimmune pancreatitis and Mikulicz syndrome, have become known to physicians as new clinical entities, especially over the past few years. We would like to present a case of IgG4-related oropharyngolaryngitis. We encountered a 80-year-old male with a severe and intractable inflammation distributed exclusively in the hypopharynx and laryngeal surface of the epiglottis. The patient did not spontaneously recover and the disease was resistant to antibiotics and NSAIDs that were administered for 4 weeks. Frequent fiberscope inspections and punch biopsies failed to provide a diagnosis, and were followed by a detailed examination under laryngomicrosurgery, with multiple tissue samplings under general anesthesia. The samples revealed proliferative, non-malignant inflammatory lesions and the pathology showed IgG4-related symptoms; indicated by all of the criteria including blood and local IgG4 elevation being met. Symptoms were quickly suppressed by a week of steroid infusion drips, followed by a gradual tapering of oral steroids in accordance with a regimen for autoimmune pancreatitis. For many years, ENT physicians have often used corticosteroids empirically for atypical pharyngitis, but usage of steroids on conditions with uncertain criteria has always been criticized. This is, as far as we know, the first report of oropharyngolaryngitis to be associated with IgG4-related symptoms. The case is thought-provoking in that some of those recurrent atypical oropharyngolaryngitis cases might be explained in accordance with this information. Moreover, the results suggest that the serum IgG4 level might be a predictive marker for the requirement of corticosteroids administration.

Keywords : pharyngolaryngitis, IgG4-related diseases, autoimmune disease

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