IgG4 Associated Chronic Sclerosing Sialadenitis Masquerading as Lymphoma

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IgG4 associated chronic sclerosing sialadenitis (IgG4-CSS) is a rare subgroup of fibro-inflammatory diseases of the salivary glands that belongs to the recently described spectrum of IgG4 related systemic diseases, characterized by elevated serum level of IgG4 with histologic features of numerous IgG4-positive plasma cells in the affected organ. IgG4-CSS presents as salivary gland mass, typically affecting the submandibular gland. Here we describe a very unusual case of IgG4-CSS, which presented as cervical lymphadenopathy with classic features of lymphoma. The patient is a 58-year-old man who recently developed painless mass in the left neck. Physical examination revealed a nontender 4.5-cm left cervical lymphadenopathy, with supportive imaging studies. Excisional biopsy was done following a nondiagnostic attempt with fine-needle aspiration. Touch imprint revealed dense lymphoplasmacytic infiltrate. Flow cytometry showed no immunophenotypic evidence of non-Hodgkin lymphoma or clonal plasma cells. Histology revealed a salivary gland with dense nodular lymphoplasmacytic aggregates surrounded by thick sclerotic bands; and no evidence of any nodal structures. Immunohistochemical stain for CD138 highlighted the numerous plasma cells admixed with reactive B and T cells. The plasma cells were polyclonal for kappa and lambda light chains, but essentially express IgG4 while negative for other immunoglobulins. These findings with concurrently evaluated high serum IgG4 level confirmed the diagnosis of IgG4-CSS in this patient, who presented with clinical features of cervical lymphoma. A thorough clinical workup along with careful histomorphologic examination and appropriate ancillary studies are mandatory for accurate diagnosis of any atypical neck mass.