Lip Biopsy in Mikulicz’s Disease
Phenotype IgG4-related Disease

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A 73-year-old man has been suffering from severe exophthalmos since the age of 50 (Picture 1a). Magnetic resonance imaging (MRI) showed marked enlargement of both lacrimal glands (Picture 1c and d), and computed tomography (CT) disclosed nodular lesions in the lung and thickened band-like lesion around abdominal aorta (Picture 2a and c). Serum immunoglobulin G4 (IgG4) level was very high (1,490 mg/dL, normal ≤108 mg/dL). Histological examination of his lip biopsy revealed heavy infiltration of mononuclear cells (Picture 3a), adding that more than 50% of these cells with IgG immunoreactivity (Picture 3b) were positively immunolabeled with antibody for IgG4 (Picture 3c). After corticosteroid therapy exophthalmos dramatically relieved within 2 weeks (Picture 1b), and pulmonary and periaortal lesions also improved on CT 20th day later (Picture 2b and d). Mikulicz’s disease phenotype has been recently noted in IgG4-related lymphoplasmacytic proliferative diseases and lip biopsy is useful for the diagnosis of this disorder (1, 2).

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References