A 52-year-old woman was admitted with bilateral painless extensive periorbital mass (Figure 1A). In 1994, left submandibular swelling was found and biopsy was unremarkable. No pathology was ascertained in the 5 lymph node biopsies between 1994 and 1999. Except for hemoglobin 9.8 g/dl and serum globulin 4.3 g/dl, other laboratory tests were normal. Antinuclear antibodies, anti-dsDNA, SSA, and SSB were negative. Schirmer’s test and salivary gland scintigraphy were normal. Minor salivary gland biopsy was consistent with grade 4 lymphocytic infiltration according to Chisholm and Mason classification. The inflammatory mass in the extraocular muscles, lacrimal glands, and retromaxillary spaces visible in orbital magnetic resonance images was reported as lymphoma. Orbital mass biopsy revealed severe lymphoid infiltration and plasma cells with high degree IgG staining. The ratio of IgG4/IgG was 53% (1100/2079 mg/dl). With diagnosis of Mikulicz’s syndrome, therapy consisting of methylprednisolone 1 mg/kg and azathioprine 2 mg/kg was begun, and significant regression was found 2 months later (Figure 1B).

REFERENCES