Orbital Myositis and Primary Sjögren Syndrome

To the Editor:

In this report, we describe a new systemic manifestation associated with primary Sjögren syndrome (pSS): orbital myositis (OM). Considered a systemic disorder, pSS is primarily characterized by lymphocytic infiltration of exocrine glands, resulting in functional impairment of salivary and lacrymal glands. The inflammatory process, however, extends beyond the exocrine glands and can potentially affect any organ.

A 40-year-old white man with pSS was admitted with diplopia. Since 2002, his disease manifested with sicca syndrome, polyarthralgia, antinuclear antibodies, and anti-SSA and -SSB antibodies, and lymphocytic sialadenitis (focus score of 1) associated with Hashimoto thyroiditis. He previously developed 2 neurological complications: aseptic meningoradiculitis following treatment with infliximab in 2002 (in the Trial of Remicade in Primary Sjögren's Syndrome randomized clinical trial)[1], and bilateral idiopathic sudden sensorineural hearing loss in 2010.

In January 2014, he presented with vertical, binocular diplopia with conjunctival hyperemia, palpebral edema, and polyarthralgia. Examination revealed infracentimetrics axillary and inguinal lymphadenopathies without parotid gland swelling. Ophthalmologic examination did not reveal ophthalmoplegia, visual field defect, or accommodative trouble; slit lamp and retinal examination was normal. Pupil light reflexes were normal. Clinical examination was otherwise normal. Cerebral magnetic resonance imaging (MRI) showed a hypersignal of the superior rectus orbital muscle on the right side on T2-weighted sequences leading to diagnosis of OM (Figure 1).

Biological examination showed lymphopenia at 710/mm^3 with erythrocyte sedimentation rate at 50 mm/h and C-reactive protein at 16 mg/l. Thyroid function was normal and thyroid-stimulation hormone-receptor antibody was negative. Rheumatoid factor was positive at 79 UI/ml. Complement factor C3 was normal at 1.15 g/l (normal range 0.8–1.54) and C4 rate was low at 0.14 g/l (normal range 0.18–0.42). Ratio of kappa/lambda serum-free light chain of immunoglobulins was normal. Cryoglobulinemia tests were negative and serum IgG rate was increased (16.5 g/l, normal range 8–13.5 g/l), but immunoglobulin G4 (IgG4) serum level was low (< 0.04 g/l). Antineutrophil cytoplasmic, antiaquaporin 4, anti-DNA, anticardiolipin, and anti-β2 GP1 antibodies were negative. Serodiagnosis for human immunodeficiency virus 1 and 2, Lyme, and syphilitic diseases were negative. Cerebrospinal fluid showed mildly elevated protein (0.55 g/l) and normal glucose rate (2.9 mmol/l) without any pleocytosis (3 white blood cells/mm^3). Tomodensitometry revealed only infracentimetric axillary, mediastinal, and inguinal lymphadenopathies.

Figure 1. Inflammatory edema of the right superior rectus orbital muscle on cerebral MRI T2-weighted coronal sequence. MRI: magnetic resonance imaging.

A new labial minor salivary gland biopsy confirmed lymphocytic sialadenitis (focus score 1.12 with no germinal center), but did not reveal IgG4 plasma cells or B cell lymphoma. Inguinal lymphadenopathy biopsy demonstrated reactive lymphadenitis without lymphoma or granulomatosis. We did not perform orbital muscle biopsy because MRI features were not suggestive of malignant disease and clinical course was spontaneously favorable.

OM associated with a pSS was diagnosed. Hydroxychloroquine 400 mg/day was initiated to prevent relapse without prednisone because of spontaneous regression of symptoms. Six months later, clinical examination confirmed complete resolution.

OM is a rare idiopathic inflammatory disease, classified as part of orbital inflammatory disease[2,3] and defined by inflammation of the extraocular orbital muscles. Clinical characteristics include orbital pain, diplopia, proptosis, swollen eyelids, and conjunctival hyperemia. Recognized causes are autoimmune thyroid (Graves disease), antineutrophil cytoplasmic antibodies-associated vasculitis, sarcoidosis, and systemic lupus erythematosus[4,5,6], and recently, observations of OM have been described in IgG4-related disease[7]. Several inflammatory diseases have also been reported associated with OM: Crohn disease and Behçet disease. Differential diagnoses include orbital cellulitis, non-Hodgkin lymphomas, metastatic solid cancer, and melanoma. To our knowledge, this is the first case report of OM in the context of pSS.

Of the autoimmune diseases, pSS is most commonly associated with non-Hodgkin lymphoma[8]. We here described an atypical ocular manifestation, initially suggestive of lymphoma, but investigation and spontaneous regression revealed a new extraglandular association with pSS that is idiopathic myositis.

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J Rheumatol 2015;42:8; doi:10.3899/jrheum.141318

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