中文題目:IgG4 相關之肌炎病例報告

英文題目:IgG4 related myosititis-case report

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Introduction:

Immunoglobulin G4-related disease (IgG4-RD) is an uncommon systemic disorder characterized by mass-like sclerosing lesions or pseudo-tumor that can involve nearly all organs. Given the rarity of this disorder and its protean manifestations, the differential diagnosis is broad and includes malignancy, infection, and other inflammatory disorders. Reports of the manifestations of IgG4-RD in the muscle are scarce. Here we report a diabetic patient presented with painful swelling of left lower leg for two weeks and IgG4 related myositis was confirmed by elevated serum IgG4 level and IgG4+ plasma cells in the muscles.

Case report:

A 52-year-old female presented with progressive painful swelling of left lower leg for two weeks. She has the history of type 2 diabetes mellitus with poor glycemic control. On physical examination, her left lower leg exhibited redness with tenderness and warmth under the skin and the remainder of the physical examination was unremarkable. Pertinent laboratory studies showed leukocytosis (white blood cell count 17300/uL) and high C-reactive protein level (31.34 mg/dL). She was initially treated with amoxicillin under the impression of cellulitis, but in vain. Her leg redness seemed to settle but edema persisted. Elevated D-dimer test (3633 ng/mL) and technetium-99m venography confirmed the diagnosis of deep venous thrombosis(DVT). Low molecular weight heparin bridging therapy as interrupting the oral anticoagulation was prescribed. Edema of the left lower leg was improved but tenderness persisted. Computerized tomography venography demonstrated soft tissue

swelling with increased infiltration of left lower leg (Figure 1, red arrow). Muscle biopsy of left lower leg demonstrated infiltration of IgG4 (Figure 2, red arrow). High serum IgG4 level and the presence of IgG4 plasma cell in the muscle suggest the diagnosis of IgG4 related myositis. She was started on intravenous 40 mg of prednisone daily for one week. During this period, the patient showed a significant response, as evidenced by improvement in the painful swelling of the left lower leg. After two weeks, prednisone was tapered. After the taper, the painful swelling continued to improve, resolving completely within six months.

Discussion:

IgG4-RD is a newly classified and more and more commonly diagnosed condition involving inflammatory and fibrotic processes, and characterized by histological findings in various affected organs. IgG4-RD has been noted to affect the lacrimal glands, salivary glands, thyroid, liver, pancreas, kidney, lymph nodes, skin, and breast. Organ specific diagnostic criteria have been established for IgG4-related autoimmune pancreatitis, Mikulicz's disease, and kidney disease. Diagnostic criteria for other involved organs has been proposed in 2011, including two major characteristics: increased serum concentrations of IgG4 and infiltration of IgG4+ cells.[1] Our case showed elevation of serum IgG4, high ratio of IgG4 to total IgG (20%), IgG4 plasma cell infiltrated in the muscle, and good response to glucocorticoid had established the diagnosis of IgG4 related myositis. To our knowledge, this is the first case of IgG4 related myositis. When clinicians encounter infectious or inflammatory myositis, IgG4 related myositis should be considered as a diagnostic possibility.

Reference:

1. Umehara, H., K. Okazaki, Y. Masaki, et al. Comprehensive diagnostic criteria for IgG4-related disease (IgG4-RD), 2011. Mod Rheumatol 22: 21-30.

Figure 1

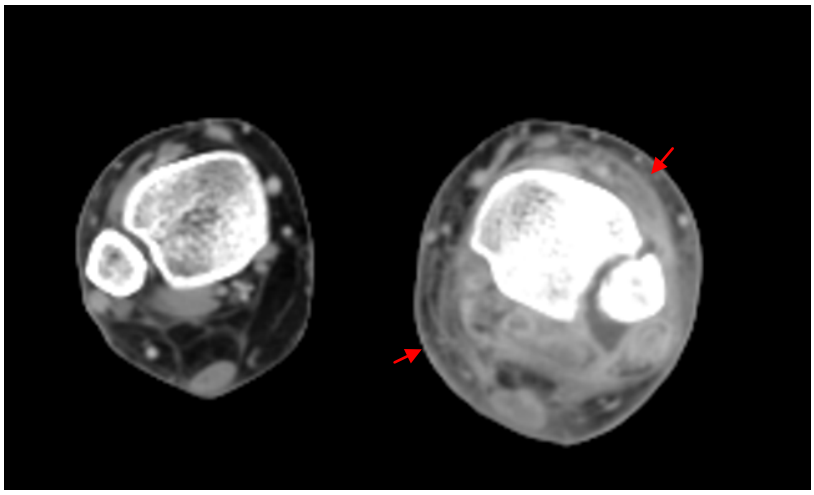

Figure 2