中文題目：一位同時具有肺、骨骼肌與脾臟侵犯之類肉瘤個案

英文題目：A sarcoidosis patient who presents with pulmonary, skeletal muscular, and splenic involvement.

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Introduction:

Sarcoidosis is a disease involving multiple organ systems, especially lung, skin, and lymph nodes. Non-caseating granulomas is characteristic pathologic presentation. Although musculoskeletal system involvement occurs in approximately one-fourth to one-third sarcoidosis patients, symptomatic myopathy is a rare extrapulmonary manifestation. Among muscular sarcoidosis, nodular myopathy is the most uncommon type. Here we present a young man who has sarcoidosis with multiple organ involvement.

Case Presentation:

A 29-year-old male presented to our department for bilateral lower leg swelling for 3 years, with rapidly progression within recent 3 weeks. Erythema nodosum over bilateral lower limbs were noted on physical examination. He also mentioned about acute polyarthritis of joints of lower extremities. He has tuberculosis (TB) lymphadenitis of mediastinum and has had complete anti-TB treatment about 3 years ago. Lower leg CT (computed tomography) scan with contrast enhancement revealed diffused, numerous subcutaneous and intramuscular micronodules in bilateral lower limbs. Chest X-ray revealed interstitial pattern of bilateral lung. Chest CT showed multiple lymphadenopathies in mediastinum and bilateral supraclavicular regions. Ground glass infiltration of bilateral upper lung and multiple splenic nodules were also found in the CT scan. We did excisional biopsy of subcutaneous nodular lesions of his lower leg. The pathologic study revealed multifocal aggregation of epithelioid histiocytes, few multinucleated giant cells and lymphoplasma cells within dermis. The CK and TB immunostains were negative. No fungal or mycobacterial microorganism can be demonstrated by GMS, PAS, and acid-fast stains. Sarcoid granuloma was highly suspected. Tissue culture of mycobacterium was negative. After discussing with the patient, we administrated oral corticosteroid as treatment for sarcoidosis. His lower leg swelling improved after treatment. Follow-up chest X-ray revealed improvement on bilateral interstitial pattern.

Discussion:

Sarcoidosis is a multi-organ involving disease with unknown etiology. The characteristic pathologic presentation is non-caseating granulomas. This disease typically involved lung and lymph nodes. Sarcoidosis affects young adults with peak incidence between 20 and 29 years of age. Up to one-third of the patients present with extrapulmonary manifestation. Skin, lymph nodes, eyes, and liver are the most involved organs. Skeletal muscle involvement has been estimated as many as half of all sarcoidosis

patients. However, symptomatic myopathy in sarcoidosis is rare, involving only 0.5-3% patients. There are three types of muscle involvement of sarcoidosis: chronic myopathy, nodular myopathy, and acute myopathy. Chronic myopathy is the most common form, usually involves female patients. In nodular sarcoid myopathy, the patient often mentioned about symmetric limb involvement and painful nodules. Acute sarcoid myopathy is the least common form of sarcoid myopathy, usually involving patient with age of 40 or younger. The clinical manifestation of acute sarcoid myopathy and other inflammatory myopathies were similar, with rapid onset of proximal weakness and myalgia. Serum creatine kinase level was usually elevated. Intraabdominal organ involvement is also presented in sarcoidosis patients. Liver is the mostly involved organ. Splenic lesion is approximal 40-60% in those with intraabdominal organ involvement. The presentation usually included splenomegaly or multi-focal splenic lesions. The aim of treatment is to reduce the burden of granulomatous inflammation and to prevent organ damage. Corticosteroid, such as oral prednisolone, is the mostly common treatment for symptomatic sarcoidosis patients. In corticosteroid-refractory disease, steroid-sparing agents were commonly administrated, despite there are limited data from controlled trials to guide the treatment. There are some studies that suggested methotrexate (MTX) as steroid-sparing regimen for sarcoidosis treatment. MTX is recommended to treat lung disease. However, there are only case reports that support MTX as treatment in musculoskeletal sarcoidosis. The duration of treatment remains uncertain. A treatment duration no less than 3 months is suggested to reduce the possibility of relapse. However, relapses are frequent following reduction or withdrawal of therapy.

Conclusion:

The diagnosis and treatment of sarcoidosis remain challenging. Histopathologic evidence of sarcoid granuloma did not exclude other granulomatous diseases. Clinicians should be aware of sarcoidosis when diagnosing patient with granulomatous diseases. Glucocorticoid therapy generally improves symptoms, but the optimal duration remains uncertain. Steroid-sparing treatment such as methotrexate could be considered but there is limited evidence on treating muscular sarcoidosis. Relapses are possible following reduction or withdrawal of therapy.