**2013 CARE Checklist**

**Title** – **Adult onset Still's disease with multiple lymphadenopathy: a case report and literature review.**

**Key Words** – Adult onset Still's disease,Lymphadenopathy,Paracortical areas,Autoinflammatory diseases,Case report.

**Abstract** –

Introduction – Adult onset Still's disease (AOSD) is a rare systemic autoinflammatory disorder that often presents with systemic multiple lymphadenopathy.In addition to the common paracortical and mixed patterns in AOSD lymph nodes histopathological features, the other morphological patterns include diffuse, necrotic, and follicular patterns.However, there have been very few reports to date on the histopathological description of AOSD lymph nodes.

Conclusion – The presentation of morphologic cases of AOSD lymphadenopathy will increase the awareness of AOSD among pathologists and clinicians, and aid in the diagnosis and differential diagnosis of AOSD lymphadenopathy from other reactive lymphadenopathies and lymphomas.

**Introduction** – Adult onset Still's disease (AOSD) is a rare group of systemic autoinflammatory diseases with complex, incompletely defined etiology and pathogenesis, mainly characterized by intermittent hyperthermia, transient skin rash, elevated blood leukocytes (neutrophils > 80%), polyarthritic pain, multiple lymphadenopathy, and a predilection for young adults [1].In 1971, Eric Bywaters first described 14 cases of Still's disease occurring from 17 to 35 years of age with clinical features very similar to those of childhood still's disease, mainly characterized by high fever, multiple skin rashes and polyarthritis, thus defining AOSD [2].Adult still's disease is often accompanied by liver and spleen enlargement and lymphadenopathy, and the clinical manifestations are complex and unspecific, sometimes similar and overlapping with lymphoma in clinical manifestations and histopathology [3], which may easily lead to misdiagnosis or missed diagnosis.In this review, we describe a case of adult onset Still's disease and review the relevant literature to explore the clinical features, pathomorphologic features of enlarged lymph nodes, and immunophenotype, with the aim of improving the level of pathologic diagnosis of the disease so as not to be misdiagnosed as lymphoma or other associated lymphadenopathy.

**Patient Information**

Patient, female, 18 years old.Two months earlier there was no obvious trigger for the development of pain in large joints of all extremities with pruritic and painless rash formation on the skin of the dorsum of the shoulders(Fig.a), both wrists and both sides of the thighs, which worsened with increasing symptoms with febrile and night sweats on admission at 6 days.

**Clinical Findings** – The patient received comprehensive medical treatment after operation, and the condition was stable without progression at the 11-month follow-up evaluation.

**Timeline** –

Table 1 Dagnosis and treatment course of patients

|  |  |
| --- | --- |
| Time | Dagnosis and treatment |
| July 23-August 7,  2020 | After admission, Ruyi Zhenbao pills and ibuprofen sustained-release capsules were taken orally, Tanreqing injection and loxef sodium were given intravenously, combined with local symptomatic treatment of traditional Chinese medicine.  On July 31, the left cervical lymph nodes were resected and sent to pathology department.  Pathological diagnosis: Adult onset Still's disease (AOSD) . |
| August 8-august 13, 2020 | Intravenous drip of methylprednisolone sodium succinate and magnesium isoglycyrrhizinate, oral calcium carbonate D3 tablets, calcitriol capsules, potassium chloride sustained-release tablets, omeprazole enteric coated tablets, soybean phospholipid powder, hepatocyte growth promoting factor enteric coated capsules, clopidogrel bisulfate tablets and cefoperazone sodium sulbactam sodium. |
| From August 14 | Methotrexate 10mg QW was given to suppress immunity, and folic acid tablets were given to reduce side effects. |
| August 15 | Discharge: no joint pain and discomfort, a little rash on both elbows, with pruritus, no exudation and ulceration, pigmentation on shoulder and back, no fever and chills. |
| August 15, 2020 to July 1, 2021 | Oral prednisone acetate tablets, 50 mg once a day, gradually reduced to 3 mg per day.  Oral methotrexate tablets 15 mg once a week, gradually reduced to 12.5 mg per week.The condition was stable with osteoporosis and poor sleep. |

**Diagnostic Assessment**

Multi joint pain of limbs for 2 months, aggravated with fever for 6 days, accompanied by pruritus.Admitted to the hospital for blood investigation: ferritin 466.4ng/ml, C-reactive protein 82.7mg/l, IL-6 (85.12pg/ml), white blood cells 15.55×109 / L (89.3% neutrophils), antinuclear antibody positive, but RF、ANCA, etc.were negative.Since onset, the patient lost 5kg of body weight.

Examination revealed bilateral posterior cervical lymph nodes, left supraclavicular lymph nodes and left posterior axillary lymph nodes without tenderness.Clinical suspicion of lymphoma, left neck lymph node resection was performed for pathological examination.

The differential diagnosis should be considered as follows: 1. Angioimmunoblastic T-cell lymphoma. 2.Dermatopathic lymphadenopathy.3.Infectious mononucleosis.4.Histiocytic necrotizing lymphadenitis (Kikuchi's disease).

Pathological diagnosis: Adult onset Still's disease (AOSD) .

**Therapeutic Intervention**

After admission, Ruyi Zhenbao pills and ibuprofen sustained-release capsules were taken orally, Tanreqing injection and loxef sodium were given intravenously, combined with local symptomatic treatment of traditional Chinese medicine.

Oral prednisone acetate tablets, 50 mg once a day, gradually reduced to 3 mg per day.

Oral methotrexate tablets 15 mg once a week, gradually reduced to 12.5 mg per week.The condition was stable with osteoporosis and poor sleep.

**Follow-up and Outcomes**

The condition was stable without progression at the 11-month follow-up evaluation.

Discharge: no joint pain and discomfort, a little rash on both elbows, with pruritus, no exudation and ulceration, pigmentation on shoulder and back, no fever and chills.The condition was stable with osteoporosis and poor sleep.

**Discussion**

AOSD often presents as a chronic passage, and patients may develop different complications within the course of the disease, which affect their clinical condition, treatment, and prognosis. Secondary hemophagocytic lymphohistiocytosis (HLH), aka macrophage activation syndrome (MAS), is the most severe complication and is associated with high mortality. Common complications are coagulopathy with multiorgan involvement including heart, lung, liver, spleen and other sites [4-5], and these patients often require more intensive treatment and have a worse prognosis. It has been shown that more than 20% AOSD patients experience recurrence and that patients with severe disease at the initial stage of the disease may be at an increased risk of recurrence, which requires intensive treatment and close follow-up [6].

**Patient Perspective** – Patients have been coming to the clinic for follow-up and treatment.

**Informed Consent** – The patient agreed to use her personal data to publish articles.

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