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A Case of Adult Onset Stills Disease

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Abstract: ***Adult-onset Still's disease (AOSD) is characterized by fever, rash, and joint pain and may lead to chronic arthritis. The cause of AOSD is unknown, and it is rare. We encountered a patient with adult-onset Still's disease following a sore throat and fever. The patient was a 40-year-old woman who consulted our hospital because of a sore throat, fever and arthritis. She was admitted and treated with antibiotics, but the fever persisted. Laboratory parameters of increased inflammatory activity, hyperferritinemia, a diagnosis of AOSD was made based on Yamuguchi criteria after the exclusion of other potential diagnoses. When acute pharyngitis is observed in association with significant changes in laboratory parameters despite mild local symptoms, or when pharyngitis is observed in association with joint pain, continuous fever, and a rash, it is important to consider AOSD.***

Keywords: Adult Onset Stills Disease , Rash , Arthritis , Sore Throat , Fever

1. Introduction

Adult onset Still's disease is a recently recognized systemic inflammatory disorder of unknown etiology and pathogenesis. It is likely to be the continuum of systemic onset juvenile arthritis. It was initially described in 1897 by George F. Still, a pathologist. The characteristic features of this illness have subsequently been reported in adults, as detailed by Eric Bywaters in 1971.

An infectious etiology has been inferred based upon the prodromal sore throat, reflecting non specific cytokines mostly IL-5, IL-6, IL-18, TNF-α. Although the disease is a sero-negative chronic polyarthritis, the initial presentation is almost always with fever and other non-specific manifestation. Most of the patients (about 75%) are between 16 -35 years age. Most striking manifestations are - quotidian fever,evanescent rash, prodromal sore throat, arthritis/arthralgia, malaise, weight loss. AOSD remains a clinical diagnosis of exclusion; with typical clinical features, laboratory abnormalities and absence of other explanations. Various diagnostic criteria have been proposed. Among them Yamaguchi criteria has the greatest sensitivity and specificity.

2. Case Report

An elderly 40 year old female patient non diabetic, normotensive, presented to JSS hospital in the third week of February with throat pain, fever, joint pains for two weeks and was admitted under internal medicine.

The patient problem appears to have started 20 days prior admission with continuous sore throat. This was followed by fever, high degree with non evanescent rashes over arms and thighs non pruritic with polyarthritis involving shoulder, elbows, wrists, knees and ankles symmetrically associated with morning stiffness lasting for more than one hour making it difficult to perform her daily routine activities. Swelling of both lower limbs up to upper 1/3rd of legs not preceded by peri orbital oedema. There was progressive loss of weight of 4kgs over 3 weeks.

There were no ocular symptoms, oro-genital ulcers or contact to infected person or major systemic symptoms. Examination revealed well built, middle aged female with macular rashes over lower back, thigh and arms, knuckle pigmentation. Bilateral pedal oedema. Posterior pharyngeal wall congested.

Patient was febrile 39.5 c with regular heart rate 110b/min, blood pressure – 130/70mmhg. She had tenderness over bilateral shoulder, wrist, knee and ankle joints with weak hand grip. Swelling of the wrist, knee, MCP joints. Limitation of finger and knee joint movements. No effusion / local raise of temperature.

Examination of abdomen-liver just palpable. Examination of the respiratory system , cardiovascular system , central and peripheral nervous system were unremarkable. The investigations revealed persistent neutrophilic leucocytosis with, left shift, increased band forms, toxic granules and vacuoles with total leukocytic count of 30,900cells/cumm , high Erythrocyte sedimentation rate of 90mm/1st hour. The electrolytes, renal parameters, sugar profile, thyroid function test were all normal. The thick and thin films for malarial parasites, serological tests for typhoid , rickettsia and brucella were all negative.

Investigations for her pyrexia revealed sterile throat swab, blood culture and urine culture. Sputum culture was negative for acid fast bacilli in three samples.

ECG, Echocardiogram and radiological assessment (including CXR, knee joint) all were normal. CRP was raised 96.6mg/dl, S.ferritin – 1650ng/ml, Negative Anti Nuclear Antibody (ANA), RA factor, ASLO titre.

Because of more than four weeks febrile illness, with continuous quotidian like fever of more than 39ºc, sore throat, arthritis and macular non evanescent rashes which was unusual to AOSD, marked acute phase reactants - high CRP, ESR and ferritin, negative ANA and RA latex and negative workup for pyrexia of unknown origin (PUO) , definite AOSD diagnosis was made according to Yamaguachi criteria. AOSD counts for 6% of all the PUO cases in the known series of cases (Data from R Aduan et al: Prolonged fever of unknown origin. Clin Res 26:558A, 1978). Unusual feature in this case was non evanescent nature of skin rash.

3. Discussion

AOSD is a rare disease affecting all races. The demographic study revealed a higher prevalence of female patients and a relatively younger onset of male patients compared with female. The reported prevalence is one per 100.000 adults aged between 16 and 35 years 1. The etiology of AOSD remains unknown, although some authors think that infective agents, especially viruses can be the trigger of the illness in susceptible patients. The viruses most commonly implicated in AOSD include rubella, parainfluza, Epstein Barr virus, Echovirus and parvovirus B19 2. A viral triggering mechanism is most often
identified by raised antiviral antibodies. There is strong association between cytokine and chronic articular disease in AOSD. Tumor necrosis factor (TNF), interleukin(IL)-18 were increased in both types of AOSD, even in remission. Soluble receptors, IL-4, IL-18 level and IL-8 were correlated with disease activity 2-3.

Stressful life events in the year preceding onset are significantly associated with increased risk for AOSD and there are no significant associations of AOSD with smoking, alcohol consumption, individual toxic substances, vaccination, blood transfusion, trauma or surgery, pregnancy or oral contraceptive use 2-5. Patient with AOSD typically present with high spiking fever, which is usually accompanied by an evanescent pink or salmon-colored macular rash on the trunk and proximal extremities. Arthralgia and polyarthritis appear later in the disease course and may be intermittent in early stages. Sore throat is common at onset, but cultures for group A streptococci are negative 4.

The major clinical manifestations consisted of fever, joints symptoms and rash, which were seen in almost all of the patients. Most had high temperature 39°c or higher, which lasted one week longer before treatment. Apparent arthritis which lasted two
weeks or longer, with three or more joints affected and also with normal findings on joint roentgenographs at the time of diagnosis, were seen in most of the patients. Typical rash was present in 87% of the definite cases 5, but another type of rash such as urticarial eruption and eczema was seen in a few cases.

Symmetrical or asymmetrical polyarthritis is found in more than 90% of patients during the first 6 months involving both large and small articulation (knees, wrists, ankles, elbows, shoulders, PIPs, DIPs, TMJ and cervical spine) 6. Initially synovitis may be fleeting or migratory, however the chronicity of synovitis is rare. These patients who had a chronic articular pattern or a polyarticular onset and course were at higher risk for develop disabling arthritis 7-8.

Other features may include pericarditis, pleuritis, splenomegaly and lymphadenopathy. Recent reviews of AOSD have emphasized the chronicity of this disease as well as highlighting involvement of major organ systems. Central and peripheral nervus system involvement is rare which include brain stem hemorrhage, seizures with fatal epilepticus, opthalmoplegia and encephalopathy were reported. Although thrombotic thrombocytopenic purpura (TTP) has been associated with autoimmune disease, usually with systemic lupus erythematosis or various form of vasculitis, it has rarely been observed in patients with AOSD. Patients with AOSD can develop multi-organ failure, which may be a manifestation of disease itself or secondary to gold therapy 8.

Laboratory studies show only non-specific abnormalities including anemia, leukocytosis with predominance of neutrophils, marked ESR elevation and thrombocytosis in most patients. Serum ferritin behaves as an acute phase reactant and is increased in many inflammatory and infectious illnesses, but for unexplained reasons it is disproportionately elevated in patient with AOSD. However an increased serum ferritin level is a nonspecific finding and should not be regarded as a diagnostic test. 80% or more of the patients present with high ESR, negative ANA, negative RF, leukocytosis, granulcytosis and liver dysfunction 8-10.

Making a diagnosis is difficult in the early stages of the disease, but is facilitated by ruling out infectious illnesses, recognizing the typical rash, and noting the development of chronic polyarthritis which resembles R.A. Diagnosis of AOSD should be considered in the course of evaluating patients with triad of fever, rash, arthritis, documentation of fever pattern and observation of patient over a minimum of at least 6-8 weeks prior to the possible diagnosis of AOSD. The Yamaguchi criteria 1992 (specificity 92% and sensitivity 96%) is the most widely used criteria to diagnose AOSD 10.

Therapy of AOSD is directed to control inflammatory symptoms and signs. About 25% of patients respond to NSAIDs and the remainder requires steroid therapy to suppress the acute systemic illness5. Systemic disease activity will require aggressive anti-inflammatory therapy with NSAIDs, steroid, methotrexate and will not respond to other conventional DMARD (gold and penicillin). Methotrexate and corticosteroids in combination may be indicated for the treatment of AOSD when polymyositis and erosive arthritis occur. Methotrexate can be used to control the acute symptoms and it is suggested that at least 6 months

of therapy should be considered to allow adequate time to assess theraputic effect with close monitoring of full blood count and liver function test. Infliximab may be effective in treatment of relapse of AOSD refractory to conventional therapy and requiring continuous high dose corticosteroid medication 10

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