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Still's disease in child and adult: A case each

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Abstract

Still's disease is very uncommon. We diagnosed 2 cases of Still's disease in a single month after being thoroughly investigated and treated for long. Both the patient presented very differently and after excluding all the infective, rheumatologic and malignant causes, they were diagnosed with Still's disease. One patient fully responded to a month treatment of steroids and Non-steroidal Anti-inflammatory drugs (NSAIDs) while the other patient was also put on Disease modifying drugs (DMARDS) and yet admitted with multiple relapses.

Keywords

still's disease; adult still's disease; yamaguchi criteria

Introduction

Still's disease is an inflammatory disorder with no known etiology and is a diagnosis of exclusion in patients with pyrexia of unknown origin. It is very uncommon with an annual incidence of 0.16 cases per 100,000 people, with an equal gender distribution and a bimodal age distribution (16-25 years and 36-46 years of age) [1]. When it starts before the age of 16 years it is called Still's Disease and after 16 years of age it is termed as Adult Still's Disease (ASD).

Yamaguchi criteria; Major: Fever>39C lasting at least 1 wk, Arthralgias/Arthritis lasting 2 wks or longer, Non pruritic papular or maculopapular salmon colored rash during fever, Leukocytosis (10,000/mm^3 or more) with atleast 80% granulocytes(PMNs). Minor: Sore throat, Lymphadenopathy, Hepatomegaly or Splenomegaly, Abnormal Liver Function Tests (particularly aspartate and alanine aminotransferase (AST, ALT) and lactate dehydrogenase (LDH)), Negative tests for antinuclear antibody (ANA) and rheumatoid factor (RF). Presence of 5 features with atleast 2 major is required for the diagnosis of Still's disease [1].

Yamaguchi criteria have the highest sensitivity (93.5%) in patients with a definite diagnosis of Still's disease [2, 3]. Serum Ferritin is markedly elevated in additional 70% of patients with Still's disease [4] and have been suggested as a serologic marker to monitor the response to treatment [5].

Case 1

16 years old Asian boy presented with 1 year history of spiking fever (100-103 F), anorexia, generalized body aches and weakness and weight loss of 7 kg. Past and Family history were insignificant. No significant findings on examination except for congested throat.

Persistent neutrophilic leukocytosis (Total Leukocyte Count, (TLC): 15000-29000/mm^3 with >80% Neutrophils(PMNs)), anemia (Hemoglobin (Hb): 7-10 mg/dl), thrombocytosis (Platelet, (PLT): 500000-600000/mm^3), Erythrocyte Sedimentation Rate (ESR): 30-120mm/1^{st} hr, with rest of the baseline investigations normal throughout the year. He was treated multiple times for pharyngitis with different antibiotics but no improvement.

2 months back, he developed generalized lymphadenopathy (LAD) and hepatosplenomegaly (HSM). Lymph node biopsy was suggestive of reactive changes. Although Tuberculosis (TB) workup (Montoux, Sputum for AFB and Gene Xpert) was negative but positive contact history for TB was and workup for TB, prolonged history and no alternative diagnosis, he was started on antituberculous therapy (ATT: Rifampicin, Isoniazid, Ethambutol, Pyrazinamide) for suspected TB adenitis. Within a month of ATT, he had no improvement in his symptoms and rather developed ATT induced hepatitis. ATT was stopped immediately and the diagnosis was reconsidered as he also developed wrist and knee joint arthritis.

Extensive workup was done to exclude infective, malignant and rheumatic diseases. Anti-Nuclear antibody (ANA)/Rheumatoid Factor (RF)/Anti-CCP/Brucella titers/Antii-Streptolysin O (ASO) titers/HBsAg/Anti HCV/Anti HIV/Blood Culture&Sensitivity/Urine Culture&Sensitivity was negative; serum ferritin was 2000 mcg/ml (Normal= 11-336 mcg/dl), Lactate Dehydrogenase(LDH) 394 mg/dl.

After excluding all other causes, diagnosis of Still's disease was made as he was fulfilling Yamaguchi criteria (Fever, neutrophilic Leukocytosis, Arthritis, LAD/HSM, Sore throat, -ve ANA/RF). He was started on steroids and non-steroidal anti-inflammatory drugs (NSAIDs) and responded within 24 hrs in the form of resolved fever, arthritis and LAD/HSM and ESR (18 mm/1^{st} hr). The patient was followed after a month and was found in good health with normal FBC, ESR (5 mm/1^{st} hr) and Ferritin (256ng/ml). NSAIDs were stopped and steroids tapered. The patient was followed next month after the treatment was stopped and was found perfectly fine. The patient is following up regularly and doing well off treatment.

Case 2

57 years old Asian Postmenopausal lady with no known comorbidities presented with 3 months history of high grade fever with rigors and chills. There was no focus of fever on history and examination. She used anti malarial and multiple antibiotics in past 3 months but no improvement.

Baseline investigations showed a significantly raised TLC of 23700/mm^3 with 93% neutrophils, low Hb of 6.7 and high platelet count, 484000/mm^3, on multiple occasions. ESR was 90 mm/1^{st}hr and C reactive protein (CRP) was negative. LFTs and RFTs were normal. All the blood and urine cultures were negative. During the admission, the patient developed wrist joint pain and a non blanch able maculapapular salmon colored rash over the body that was only during the fever. Autoimmune profile (ANA/RF/Anti-CCP/c-ANCA/p-ANCA) was negative. ASO titers and Brucella titers were also negative. Abdominal ultrasound showed mild renal parenchymal disease. There were no cast cells in urine but 24 hour urinary protein was 520 mg (Normal= up to 141 mg/24 hr) and Creatinine clearance was 56.7 ml/min (Normal= 70-150 ml/min). Serum Ferritin was 2000 mcg/dl.

Excluding all the possible causes, the patient was looked for ASD. The patient qualified the Yamaguchi criteria (Fever, Rash, Arthralgia/Arthritis, Neutrophilic leukocytosis, -ve ANA/RF) and hence was diagnosed with ASD. The patient was started on Steroids and NSAIDs. She responded to treatment. The patient was sent home and asked to follow up after a month. On next visit in outpatient setting (OPD), the patient was much improved in terms of her fever, rash and FBC/ESR but still had joint pains. She was asked to continue the treatment and come back for follow up. She was still not fully recovered on her next visit so the dose of steroids was increased.

She remained well with the treatment for some time but again presented with high grade fever. She was readmitted and still no focus was found. She was started on azathioprine along with NSAIDs and steroids and responded. The patient is doing well till date and is still having regular follow ups in medical OPD.

Discussion

Although very uncommon, yet 2 patients were diagnosed with Still's disease in a single month. Both of them presented with pyrexia of unknown origin for long. One was treated multiple times for sore throat. Case 1 fully responded to NSAIDs and steroids only and fully recovered within a month of treatment while case 2 was on Azathioprine as well and yet admitted multiple times with relapses. It is always tiresome and difficult to give a patient the diagnosis of exclusion and then stick to that diagnosis but once diagnosed, can save the patient from unnecessary investigations and treatments for lifelong.

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