A Case of an Adult-Onset Still's Disease

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INTRODUCTION

Adult-onset Still's disease (AOSD) is considered to be one of the rare systemic inflammatory disorders with unknown causes. Characterized by fever, arthritis, evanescent rash and other multi organ presentations. Initially, it was described in children by George F.still in 1896[1] Then the disorder was described in adults with similar symptoms by Bywaters in 1971[2]. The annual incidence of AOSD is 0.16 cases per 100000 indusial equally distributed between males and females, With bimodal age distribution between 15-25 years old and 36-45 years[3]. We present a case of 30 years old female who was diagnosed with Adult-onset Still's disease (AOSD)

THE CASE

A 30-year-old medically free female, having multiple food and drug allergy, Presented to Emergency department with history of fever, arthralgia and body rash for 10 days. The fever reaches 39 c, on and off associated with chills. Arthralgia mostly in the bilateral knees, elbows and wrists, lasts for 1 hour and worsens with activities during daytime. The rash is mainly over both legs and arms. Furthermore, she had similar type of on-off rash last year, non-pruritic in nature. Patient denies history of weight loss, night sweats, body lumps nor recent travel. Added to that there were no any previous admissions with similar symptoms in the past. There is positive Family history of colon malignancy of both non-consanguineous parents.

On clinical examination, patient was febrile (38 c), otherwise hemodynamically stable. Maculopapular, salmon colored rash noticed in both legs and arms mainly over the joints. No axillary lymphadenopathy appreciated. Other systems examination was unremarkable.

Lab investigations revealed a markedly increased level of White blood count, platelets, liver enzymes and ferritin. Inflammatory markers including CRP, ESR, and LDH were also elevated. patient was admitted in medical ward for further management. viral serology, TB screen, Rheumatoid factor (RF) and anti-nuclear anti body (ANA) reported negative. Prophylactic iv antibiotic was administered directly after taking blood and urine culture, which were also negative. Chest x-ray and Echo were normal. Abdomen ultrasound showed mild periportal fibrosis and chronic calcular cholecystitis with no other organomegaly.

Based on clinical and laboratory findings the patient was diagnosed with AOSD, fulfilling the yamaghuchi criteria. 60 mg of prednisolone was commenced with excellent response and showed clinical improvement of symptoms within couple of days. She was discharged on steroid tapering dose and Hydroxychloroquine. Currently, she is following up in outpatient clinic. Patient was following with dermatology clinic as well, skin biopsy done and confirmed the diagnosis of uraturia related to her medication allergy.

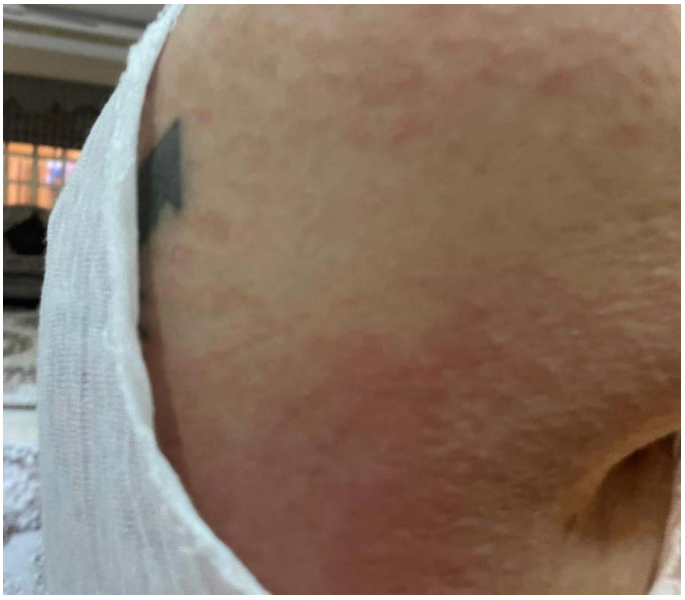

Figure 1: Salmon color rash over right back and deltoid region

DISCUSSION

Adult-onset Still's disease (AOSD) is a systemic, rare inflammatory arthritis that mimics many other illnesses. It was first described by Bywaters in 1971 with a similar picture of juvenile idiopathic arthritis in children[2]. The etiology of Adult-onset Still's disease (AOSD) remains unclear, some studies have concluded that both genetic predisposition and infections are contributors to the disease. Organisms such as Yersinia enterocolitis and mycoplasma pneumonia were identified as possible pathogens[4,5]. Moreover, it has been found that AOSD is associated with high levels of Interleukin 6, Interleukin 18 and tumor necrosis Factor-Alpha (TNF-A)[6].

Typical symptoms of AOSD are spiking fever, salmon rash and arthritis. Rash is described as maculopapular, evanescent, non-purpuric distributed mainly over trunk and extremities. Daily spiking fever is a feature of AOSD, it can reach up to 39 degrees[4]. Patients usually complain of arthralgia over wrist, knees, ankles, elbows, shoulder and proximal interphalangeal joints. Other possible presentations are myalgia, sore throat, pharyngitis and pericarditis. Clinical examination might reveal hepatosplenomegaly and lymphadenopathy[7].

Lab wise, AOSD is associated with increased level of serum and glycosylated ferritin, white blood count, liver enzymes, LDH and acute phase reactants (CRP, ESR, etc)[8].

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Since Adult-onset Still's disease is a diagnosis of exclusion, it is crucially important to rule out other pathologies such as infections, malignancy, drug reactions and other rheumatological disorders. The Yamaguchi criteria is the most commonly used to diagnose after excluding other potential causes. Yamaguchi criteria is subdivided to 4 major and 4 minor criteria. major criteria are: Fever of 39 c for at least 1 week, arthralgia for 2 weeks or more, salmon-colored non-purpuric rash over trunk and extremities, Leukocytosis (10,000 microL or more with 80% granulocytes. Minor criteria are: hepatomegaly or splenomegaly, sore throat, lymphadenopathy, deranged liver function test and negative test for both antinuclear antibody and rheumatoid factor. To diagnose AOSD, 5 of the criteria above must be present with at least 2 of the major criteria after exclusion of other pathologies such as: malignancies, other rheumatological disorders and infections[9].

NSAIDS are usually started as a supportive therapy during the process of diagnosing AOSD[7]. corticosteroids are considered the first line treatment for most of the moderate cases of AOSD combined with NSAIDS. Disease modifying anti rheumatic agents (DMARDs) are used in in cases where remission with corticosteroids was not achieved, as steroid sparring agent and in severe cases of AOSD[10]. Biologic therapies such as Anakinra, Canakinumab, TNF blockers and Tocilizumab are used in severe cases or steroid dependent AOSD not responding well to conventional therapy[11,12]. Most of the recent studies recommend the use biologic therapy, particularly Anakinra in severe cases of AOSD as it showed excellent response when compared with glucocorticoids and DMARDS[13].

CONCLUSION

Adult-onset still's disease is a rare disorder with unknown etiology, which remains a challenge to physicians with limited experience encountering such a disease. Thus, AOSD should be considered in patients presenting with un explained fever, rash and joints pain. A detailed history, proper physical examination and specific laboratory investigations are required to diagnose the disease and to rule other pathologies.

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