CASE REPORT

ATYPICAL CUTANEOUS PRESENTATION OF ADULT ONSET STILL DISEASE

Hammad Shah, Humaira Achakzai, Muhammad Zuhaid

Department of General Medicine, Rehman Medical Institute, Hayatabad, Peshawar-Pakistan

Adult onset still disease is a rare systemic inflammatory disease which presents with cardinal symptoms of spiking fever, arthralgia, and characteristic non pruritic evanescent salmon pink rash and neutrophilic leukocytosis. However it can have accompanied atypical cutaneous manifestations of pruritic plaques and papules. Clinicians need to be aware of this condition so that they can correctly diagnose it and prevent its serious complications.

Keywords: Still's disease, adult-onset, diagnosis, young, adult female, joint diseases

J Ayub Med Coll Abbottabad 2016;28(2):417–9

INTRODUCTION

Adult onset still disease (AOSD) is a rare systemic inflammatory disease which has four main cardinal symptoms of spiking fever > 39 °C, arthralgia or arthritis, skin rash and hyper leucocytosis (>10x10^9 /L) with neutrophil count of >80%.[1] The diagnosis of this disease is clinical and needs exclusion of infections, inflammatory and autoimmune diseases because it manifests very similarly to them.[2] Negative immunological markers like ANA , Anti CCP and RF are very characteristic in the diagnosis of AOSD.[3] There are raised levels of serum ferritin which is a marker of active phase of AOSD along with increase in inflammatory markers like ESR, CRP and LDH.[1,4–6] These patients also experience sore throat, unresponsive to antibiotics.[7] They have high grade fever which recurs daily.[7] In these patients 65% have lymphadenopathy, 42% have splenomegaly and 40% have hepatomegaly.[7,8] There are different clinical criteria for diagnosis of AOSD, however the most accepted criteria for preliminary diagnosis is yamaguchi criteria, it has highest sensitivity of 92% and specificity of 96.2%.[8,9] Although AOSD has characteristic salmon pink colored, non-pruritic skin rash but can manifest as atypical pruritic skin lesions.[10,11]

CASE REPORT

A 27 year old lady from Sakhakot, Malakand presented to us with 21 day history of high grade fever, sudden in onset, not associated with rigors and chills or sweating and recurring every day. She developed a pruritic rash two days later, which started over her shoulder and later extended to her upper chest and back. She was having generalized body aches with severe pain in both her knee joints. She also complained of sore throat with mild dry cough. The aches and pains were debilitating to the extent that they were interfering with her daily activities and she was bed bound. There was no history of photosensitivity, morning stiffness, burning micturition, nausea, vomiting, pain abdomen or change in bowel habits. Prior to this illness she was generally healthy although a similar though milder episode had occurred five years ago. No clear cut diagnosis was made and she made an unremarkable recovery. She is married with no family history of note. At presentation she was acutely unwell and had to be carried to ER.

On initial examination she was febrile with a temperature of 101°F, toxic looking with a flushed face. Her throat was clear and there was no palpable lymphadenopathy. There was an erythematous maculopapular rash over upper chest, shoulder and lower back. Abdomen was non tender with a palpable spleen. She had marked neck rigidity but no other neurological findings. Examination of joints was normal. Investigation showed Hb- 9.8 g/dl, WBC- 35.8x10^9/L, platelets- 361x10^9/L, Neutrophils- 89.5%. Peripheral Smear showed Neutrophilic Leucocytosis with toxic changes. All the inflammatory markers were markedly raised, having ESR of 87 mm at 1st hour, CRP of >32 mg/dl, Serum Ferritin- 3925.36 ng/ml, LDH-1026 U/L. An initial impression of meningitis was made and she was admitted to HDU (High dependency Unit). Blood and urine culture reports were sent and I/V antibiotics were started.

Immediate Lumbar puncture was done after ruling out papilledema on fundoscopy. CSF report returned as normal. By next morning her temperature settled and she improved. However she continued to have recurring high grade fever, persistence of rash along with return of body aches and joint pains. She was initially started on I/V ceftriazone and amikacin but had to be changed to more broad spectrum piperacillin/sulbactam with amikacin because of un-resolving fever Autoimmune screening was done including ANA, Anti CCP, anti ds-DNA, ASO titer and Hbs Ag

which were all negative. Her renal and liver functions were normal. Further infective workup like brucella, monospot, TB IgG/IgM, Blood C/S and Urine C/S were all negative. CT Chest showed hilar lymphadenopathy with minimal pleural effusion while CT abdomen showed hepatosplenomegaly. Once a negative blood and urine culture was reported and there were no other positive infective markers, the focus shifted to an autoimmune aetiology.

Given the negative autoimmune screening and her presentation diagnosis of Adult Onset Still Disease was considered. Yamaguchi criteria, was satisfied and patient was commenced on oral prednisolone. This lead to a dramatic improvement in her symptoms and was discharged on day 7th of admission when she improved subjectively, remained completely afebrile for 48 hours, her appetite increased, rash improved and joint pains resolved completely.

She came on day 7th of her discharge for follow up and was completely afebrile. All the inflammatory markers were reduced having CRP of <2 mg/dl, ESR of 27. Her Hb improved to 10.6 g/dl and TLC count reduced to 16.8x10^{9}/L.

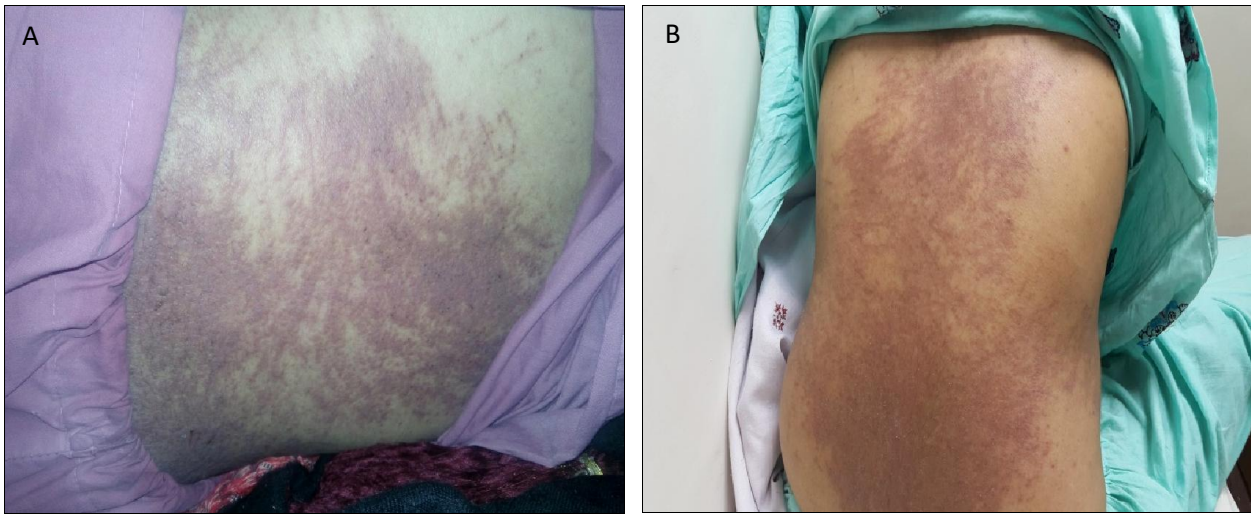

Figure-1: Erythrematous maculopapular pruritic rash in Adult onset still disease (A): Day 1st of admission. (B): Day 7th on day of discharge.

DISCUSSION

Fever, arthralgia, rash and splenomegaly is very common presentation and differential diagnosis include Rheumatic fever, Infectious mononucleosis, Systemic lupus erythematosis, Polyarthritis nodosa, Henosch schonlein purpura, Rheumatoid arthritis, Enteric Fever, Tuberculosis, brucellosis, lyme disease and Gonococal Meningitis.2 However neutrophilic leukocytosis,[1] spiky quotidian fever,[7] raised serum ferritin level,[4–6] lymphadenopathy,[7,8] hepato-splenomegaly,[7,8] negative immunological markers like ANA, Anti-CCP, ASO, Anti ds-DNA,[3,8] sore throat not responding to both broad spectrum and atypical cover antibiotics,[7] and a negative blood C/S and urine C/S favoured the diagnosis of AOSD[2]. Moreover it satisfies the Yamaguchi criteria which has a sensitivity of 92% and specificity of 96.2%.[8,9]

Although still disease has characteristic salmon pink, non-pruritic, evanescent rash during febrile episodes,[1,2,8] but it can present with atypical cutaneous manifestations as well[10,11]. This patient despite having peculiar erythematous rash over her cheeks with complete sparing of her nose during febrile episode also had a distinctive atypical pruritic papules over her left shoulder, upper chest and back. Therefore it is very essential that clinicians worldwide should be well aware of this for prompt diagnosis and prevention of complications.

REFERENCES

1. Fautrel B. Adult onset Still disease. Best Pract Res Clin Rheumatol 2008;22(5):773–92.
2. Bagnari V, Colina M, Ciancio G, Govoni M, Trotta F. Adult onset Still disease. Rheumatol Int 2010;30(7):855–62.
3. van de Putte LB, Wouters JM. Adult onset Still disease. Baillière's Clin Rheumatol 1991;5(2):263–75.
4. Fautrel B, Le Moël G, Saint-Marcoux B, Taupin P, Vignes S, Rozenberg S, *et al*. Diagnostic value of ferritin and glycosylated ferritin in adult onset Still's disease. J Rheumatol 2001;28(2):322–9.
5. Vignes S, Le Moël G, Fautrel B, Wechsler B, Godeau P, Piette JC. Percentage of glycosylated serum ferritin

remains low throughout the course of adult onset Still's disease. Ann Rheum Dis 2000;59(5):347–50.
6. Schwarz-Eywill M, Heilig B, Bauer H, Breitbart A, Pezzutto A. Evaluation of serum ferritin as a marker for adult Still's disease activity. An Rheum Dis 1992;51(5):683–5.
7. Cush JJ. Adult-Onset Still's Disease. Arthritis Foundation 49(6). [Internet]. [cited 2015 Feb 15]. Available from: http://radio-weblogs.com/0135175/gems/AOSD%20Arcticle%20by%20Dr%20Cush.pdf
8. Yamaguchi M, Ohta A, Tsunematsu T, Kasukawa R, Mizushima Y, Kashiwagi H, *et al*. Preliminary criteria for classification of adult Still's disease. J Rheumatol 1992;19(3):424–30.
9. Fautrel B, Zing E, Golmard JL, Le Moel G, Bissery A, Rioux C, *et al*. Proposal for a New Set of Classification Criteria for Adult Onset Still Disease. Medicine (Baltimore) 2002;81(3):194–200.
10. Affleck AG, Littlewood SM. Adult-onset Still's disease with atypical cutaneous features. J Eur Acad Dermatol Venereol 2005;19(3):360–3.
11. Yamamoto T. Cutaneous manifestations associated with adult-onset Still's disease: important diagnostic value. Rheumatol int 2012;32(8):2233–7.

Address for Correspondence

Dr. Hammad Shah, Department of General Medicine Rehman Medical Institute, Hayatabad Peshawar-Pakistan
Cell: +92 333 924 4644
Email: drhammadshah@gmail.com