Journal of the Turkish Academy of Dermatology
eISSN 1307-394X

Letter To The Editor
DOI: 10.6003/jtad.18122l1

Macular Rash on Upper Extremities: Adult Still's Disease

Dear Editor.- Adult Still's Disease (ASD) is a rare systemic inflammatory disease of unknown pathogenesis. ASD is more commonly seen in females and mostly affects individuals aged 15-45 years. Although the exact etiology of ASD remains unknown, numerous infectious agents and genetic factors have been implicated [**1**].

A 59-year-old male patient presented to our clinic with one-month history of fever, recurrent sore throat, myalgia, arthralgia, and skin rash. Physical examination revealed a temperature of 39.2 °C and salmon-colored maculopapular rash that appeared in the whole body, particularly in the upper extremities, during defervescence from high fever (**Figures 1 and 2**). In the submandibular area, a mobile and painless lymphadenopathy with a diameter of 1x1 cm was detected. Arthralgia was present in bilateral ankles. Laboratory parameters were as follows: leukocyte 20,810/mm^3 (90% polymorphonuclear leukocyte), hemoglobin 9.5 g/dl, thrombocyte 199,000/mm^3, C-reactive protein 110 mg/L, erythrocyte sedimentation speed 94 mm/h, and ferritin 2,504 ng/ml. Hepatosplenomegaly was detected on abdominal ultrasonography. Depending on these findings, the patient was diagnosed as ASD.

Typical clinical features of ASD include fever, skin rash, and arthritis. Fever is found approximately in 95% of the cases and it lasts at least one week [**2**]. ASD often presents as a salmon-pink colored macular rash that is found in 97% of the cases. ASD is predominantly seen in the proximal parts of the extremities and on the trunk and it can also occur on the neck and the face in 15% of the cases. The rash often becomes more noticeable during defervescence from high fever and spontaneously disappears as the fever decreases. The rash is generally not itchy. A biopsy investigation of the lesions often reveals mild perivascular infiltrate consisting of lymphocytes and neutrophils [**1,3**].

Arthritis and arthralgia have been reported in 90% of the patients with ASD. Lymphadenopathy is seen in 44% of the patients, particularly involving the submandibular lymph nodes. Splenomegaly is seen in 40% of the cases. Moderate hepatomegaly and increased transaminase are detected in most patients. Erythrocyte sedimentation speed is usually above 100 mm/h and C-reactive protein and ferritin levels are often high. Leukocyte rate is above 10,000/mm^3 in >90% of the cases and polymorphonuclear leukocytes appear in most cases [**2,3,4**].

There is no specific diagnostic test for ASD. However, the diagnosis of ASD is often made based on the presence of suspicious clinical and laboratory findings and by ruling out infectious diseases, malignancies, other collagen tissue diseases, and fever of unknown causes [**5**]. In our patient, the diagnosis was made by excluding infectious diseases, malignancies, and collagen tissue diseases.

The initial step in the treatment of ASD includes nonsteroidal anti-inflammatory drugs and corticosteroids. Subsequently, low-dose methotrexate (7.5-25 mg/week), hydroxychloroquine, azathioprine, and cyclosporine can be used [**1,3**]. Our patient was initiated on methylprednisolone 1 mg/kg/day and methotrexate 15 mg/week.

The diagnosis of ASD is often missed or delayed since ASD does not have specific laboratory findings and it is not prioritized in clinical practice. ASD should be kept in mind in patients presenting with high fever and salmon-pink colored macular lesions in the proximal parts of extremities.

Published: *J Turk Acad Dermatol* 2018; **12 (2)**: 18122l1.This article is available from: http://www.jtad.org/2018/2/jtad18122l1.pdf
Key Words: Adult, Still's Disease, Rash

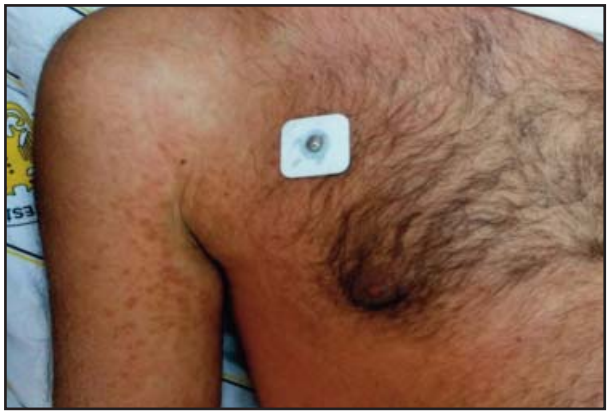

Figure 1. Salmon-colored maculopapular rash scattered in the whole body

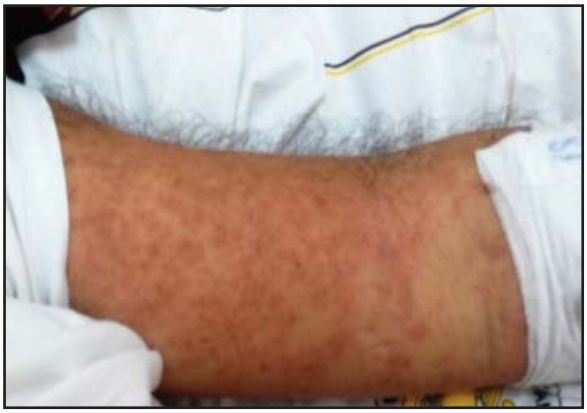

Figure 2. Salmon-colored maculopapular rash seen in the upper extremities

Isa An,* MD
Derya Ucmak, MD

*Department of Dermatology, Dicle University Medical Faculty, Diyarbakır, Turkey

E-mail: is_an89@hotmail.com

References

1. Kontzias A, Efthimiou P. Adult-onset Still's disease: pathogenesis, clinical manifestations and therapeutic advances. Drugs 2008; 68: 319-407. PMID:18257609
2. Mehrpoor G, Owlia MB, Soleimani H, Ayatollahi J. Adult-onset Still's disease: a report of 28 cases and review of the literature. Mod Rheumatol 2008; 18: 480-485. PMID:18754076
3. Pouchot J, Sampalis JS, Beaudet F, et al. Adult Still's disease: manifestations, disease course and outcome in 62 patients. Medicine 1991; 70: 118-136. PMID: 2005777
4. Yamamoto T. Cutaneous manifestations associated with adult-onset Still's disease: Important diagnostic values. Rheumatol Int 2012; 32: 2233-2237. PMID: 22198666
5. Nagai Y, Hasegawa M, Okada E, Hattori T, Tago O, Ishikawa O. Clinical follow-up study of adult-onset Still's disease. J Dermatol 2012; 39: 898-901. PMID: 22734944