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CASE BASED REVIEW

Cutaneous manifestations of adult-onset Still's disease: a case report and review of literature

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Abstract Adult onset Still's disease (AOSD) is a systemic inflammatory disorder of unknown etiology and pathogenesis characterized by high spiking fever, arthralgia or arthritis, sore throat, lymphadenopathy, hepatosplenomegaly, serositis, and transient cutaneous manifestations. Although more common in children, cases are seen also in adults. Cutaneous involvement is common and may be suggestive for the diagnosis. A case of AOSD in a 35-year-old man is reported here, presenting with urticarial maculopapular rash of trunk, high spiking fever, acute respiratory distress syndrome, and myopericarditis. Skin biopsy showed interstitial and perivascular mature $CD15^{+}$ neutrophils. A comprehensive review of literature showed that cutaneous involvement occurs in about 80 % of patients, with various clinical presentations. The most common skin manifestation is an evanescent salmon pink or erythematous maculopapular exanthema, predominantly on the trunk and proximal limbs, with rare involvement of face and distal limbs. Less common manifestations include persistent erythematous plaques and pustular lesions. A constant histopathologic finding is the presence of interstitial dermal neutrophils aligned between the collagen bundles. This pattern may provide an easy accessible clue for the definitive diagnosis of AOSD and exclude other diagnosis such as drug eruptions or infectious diseases.

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Introduction

Adult onset Still's disease (AOSD) is a rare systemic inflammatory disorder of unknown etiology, characterized by spiking fever, usually exceeding 39 °C, an evanescent cutaneous manifestations, arthralgia or arthritis, and multiorgan involvement. AOSD occurs worldwide, and women seem to be affected more often than men. AOSD usually affects young people, with 75 % of the cases reporting disease onset between 16 and 35 years of age [1]. AOSD may affect several organs and the full constellation of symptoms may not be present at onset, so the evolution of case may take several weeks or months [2]. Fever is a constant feature, associated with cutaneous lesions and arthralgia or arthritis of big joints. Other organs involved are the liver, the cardiopulmonary, and the hematologic systems. There are several sets of classification criteria for AOSD, all developed from retrospective studies, but Yamaguchi's criteria [3] were shown to be by far the most sensitive (93.5 %) [4]. Cutaneous manifestations are well accepted as a major diagnostic criterion, and, more important, the skin is the most easily accessible site for obtaining histological samples. Despite these well-established diagnostic criteria, delayed diagnosis is still common, and the skin lesions are often misdiagnosed as infectious exanthemas or allergic reactions to drugs. Herein, a case of AOSD is reported that presented with skin lesions which allowed an easy diagnosis. Moreover, the literature has been reviewed for cases of AOSD in whom cutaneous lesions and the corresponding histology had been described in detail.

Clinical case

A 35-year-old man presented with 15-day history of high fever, with evening spike of 40 °C, asthenia, cough, dyspnea, and a sense of tightness in the chest, not responsive to antibiotics (cephalosporin) and anti-inflammatory (ibuprofen) therapy. Laboratory investigations revealed elevated C reactive protein (262 mg/L; n.v. <5 mg/L), alanine amminotransferase (ALT) (167 U/L; n.v. 5–45 U/L), and ferritin (258 μg/L; n.v. 20–200 μg/L) and increased alpha-1 protein (9.6 g/L; n.v. 2.4–5.6 g/L), alpha-2 protein (8 g/L; n.v. 4.8–9.6 g/L), and gamma globulin (17.6 g/L; n.v. 8–16 g/L). Moreover, there was increased creatinine (2.3 mg/dL; n.v. 0.59–1.29 mg/dL) with diuresis reduction (600 mL/day) and normal blood cells count and triglycerides. Echocardiography showed minimal reduction of global function of left and right ventricles (ejection fraction of 33 %), with pericardial effusion referable to acute myopericarditis. Thoracic computed tomography (CT) scan revealed bilateral parenchymal densifications and interstitial thickening, more evident in the right superior lobe. Continuous positive airway pressure (CPAP) was started for respiratory failure. Screening for autoimmune diseases was negative, such as antinuclear antibodies (ANA), extractable nuclear antigens (ENA), anti-neutrophil cytoplasmic antibodies (ANCA), anti-liver and anti-kidney microsomes antibodies (LKM), and rheumatoid factor. All microbiologic tests performed were negatives (research on urine of *Streptococcus pneumoniae* and *Legionella pneumophila* antigens, malaria test, research on blood of *Aspergillus* antigen, Epstein-Barr virus (EBV) DNA, hepatitis C (HCV) RNA, hepatitis B (HBV) DNA, *Toxoplasma gondii* DNA, serologies for *Salmonella typhi*, *Brucella abortus*, *Rickettsia conorii*, *Legionella pneumophila*, *Coxiella burnetii*, *Herpes simplex* viruses 1–2, adenovirus, coxsackie virus, cytomegalovirus, parvovirus B19, enteroviruses, leptospira, respiratory syncitial virus (RSV), influenza viruses A–B, parainfluenzae viruses 1–3, EBV, West Nile virus, chikungunya virus, dengue

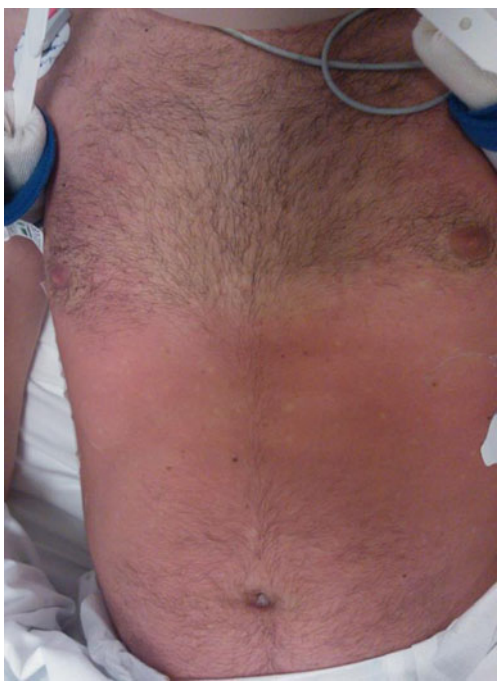

Fig. 1 Erythematous-edematous plaques on the trunk and upper limbs (**a**), confluent in wide areas of erythema (**b**)

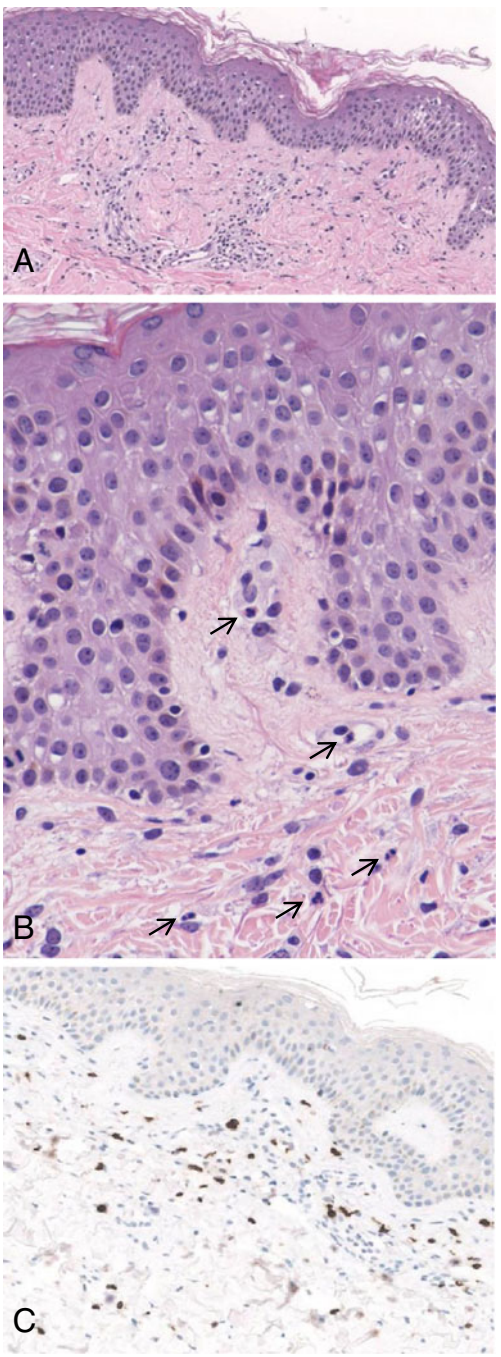

Fig. 2 Mild epidermal spongiosis and dermal superficial inflammatory infiltrate, consisting of perivascular lymphocytes and histiocytes (hematoxylin and eosin, original magnification×20) (**a**); interstitial and intravascular neutrophils among the inflammatory cells (hematoxylin and eosin, original magnification×40, *arrows*) (**b**); confirmed by immunohistochemistry positive for CD-15 antibody (original magnification×20) (**c**)

Table 1 Characteristics of patients with cutaneous involvement in adult onset Still's disease

Date/authors	No. of patients/ sex/age/age range	Systemic involvement	Type of cutaneous findings	Skin histology
2014/Kikuchi N et al. [14]	6/F/33–83	Arthralgia, hepatopathy, pericardial and pleural effusion	Persistent papules, plaques with linear distribution on trunk and limbs	Dyskeratosis in upper epidermis with focal hyperkeratosis
2014/Rathindra et al. [12]	1/M/50	Fever, arthritis, hemophagocytic lymphohistiocytosis	Persistent plaques, linear pigmentation	Necrotic keratinocytes, lymphocytes and neutrophils in papillary and mid-dermis
2001/Suzuki et al. [11]	1/M/25	Fever, arthralgia	Persistent plaques, linear pigmentation	–
1999/Lubbe J et al. [15]	1/F/16	Fever, sore throat, arthritis, pericardial effusion, hepatosplenomegaly	Persistent brownish papules and plaques	Acanthosis, parakeratosis, and intraepidermal necrotic keratinocytes; perivascular infiltrate of lymphocytes, neutrophilic granulocytes, and histiocytes without frank vasculitis
2005/Thien Huong NT et al. [16]	1/F/23	Fever, polyarthralgia, polyadenopathy	Pigmented plaques	–
1994/Kaur S et al. [17]	1/F/19	Arthralgia, lymphadenopathy, hepatosplenomegaly, sore throat	Erythematous papules and plaques on trunk and legs	Epidermal hyperkeratosis and perivascular lymphomononuclear infiltrate
2006/Yang CC et al. [18]	1/F/47	Fever, dyspnea, sore throat, polyartralgia, pleural effusion, splenomegaly	Erythematous and violaceous maculopapules	Necrotic keratinocytes in the upper epidermis and a superficial perivascular infiltrate of lymphocytes and neutrophils
2005/Affleck AG et al [19]	1/F/29	Fever, sore throat, arthritis	Erythematous, fixed plaques slightly raised	Dermal perivascular edema with a perivascular infiltrate of lymphocytes and neutrophils
2013/Said NH et al. [20]	1/M/23	Fever, sore throat, myopericarditis, arthralgia, hepatosplenomegaly	Macules, or maculopapules of salmon-pink color	Necrotic keratinocytes in the upper half of epidermis with a neutrophilic infiltrate in the papillary dermis
2014/Cho YT et al. [13]	1/F/38	Fever, sore throat, arthralgia, lymphadenopathy hepatosplenomegaly	Erythematous and brownish persistent papules and plaques on anterior chest wall, abdomen and back	Parakeratosis, acanthosis, spongiosis, scattered dyskeratotic cells in the upper epidermis, and mixed dermal perivascular infiltrate of lymphocytes and neutrophils
1998/Setterfield JF et al. [21]	1/F/32	Arthralgia	Urticarial annular lesions on trunk and limbs	Dermal edema with perivascular neutrophilic infiltrate
2000/Salaffi S et al. [22]	1/F/55	Fever, arthralgia,	Urticarial lesions on trunk and limbs	Papillary dermal edema with perivascular and interstitial neutrophilic infiltrate, without vasculitis
2006/Criado RF et al. [23]	1/F/52	–	Urticarial eruption	–
2011/Criado PR et al. [24]	2/F/24,34 1/M/26	Arthritis, sore throat, lymphadenopathy, pleural effusion, splenomegaly	Erythematous edematous plaques on trunk and linear erythematous lesions	Perivascular and interstitial inflammatory infiltrate of lymphocytes and neutrophils with dermal edema
2004/Soy M et al. [26]	1/M/38	Sore throat, arthritis	Urticarial eruption and angioedema of lips, soles and palms	–
1990/Otha A et al. [10]	30/M/35 60/F/46	Arthritis, sore throat, lymphadenopathy, hepatosplenomegaly, pericarditis, pneumonitis	Maculopapular, urticarial, and eczematous lesions, acne-like lesions	–
2002/Lee JB et al. [27]	1/F/46	Polyarthralgia, splenomegaly	Vesiculopustules on hands and feet	Fibrin thrombi in small vessels with inflammation in the upper and mid-dermis, subepidermal bulla, and ischemic necrosis of the overlying epidermis
2006/Bachmeyer C et al. [28]	1/F/24	Sore throat, arthritis/arthralgia	Erythematous papules and plaques with overlying	Dermal edema with perivascular infiltration of neutrophils; subcorneal

Table 1 (continued)

Date/authors	No. of patients/ sex/age/age range	Systemic involvement	Type of cutaneous findings	Skin histology
			vesicles and nonfollicular pustules	neutrophilic pustule with epidermal spongiosis, dermal edema without vasculitis
2001/Perez C et al. [29]	1/M/39	Sore throat, polyarthritis hepatosplenomegaly	Generalized erythema with papules and plaques	Perivascular infiltrate of dermal vessels with eosinophils, histiocytes, lymphocytes, and neutrophils within the vascular lumen
2003/Fujii K et al. [30]	1/F/64	Lymphadenopathy, polyarthralgia	Generalized erythema with edematous plaques on trunk	Mononuclear cell infiltration with a few neutrophils around dilated blood vessels in the edematous upper-mid-dermis; epidermis intact
2013/Ciliberto H et al. [31]	1/F/40	Sore throat, polyarthralgia	Erythematous, scaly plaques in a shawl distribution and hyperpigmented, erythematous, excoriated papules in a linear arrangement on back and lower extremities	Orthokeratosis and numerous clustered dyskeratotic keratinocytes in epidermis. Mixed perivascular and interstitial inflammatory infiltrate in the papillary dermis with neutrophils and lymphocytes

virus, *Leishmania donovani*, *Mycoplasman pneumoniae*, *Chlamydia pneumonie*, *Borrelia burgdoferi*, Quantiferon TB gold test, coproculture). During the fifth day of hospitalization, the patient developed erythematous-edematous plaques on the trunk and upper limbs, confluent in wide areas of erythema, moderately itching (Fig. 1). A skin biopsy taken from the left side of the trunk demonstrated mild epidermal spongiosis and dermal superficial inflammatory infiltrate, consisting of perivascular lymphocytes and histiocytes (Fig. 2a, b). Interstitial and intravascular neutrophils were also evident among the inflammatory cells, confirmed by their positive for anti-CD-15 antibody by immunohistochemistry (Fig. 2c). No hyphae and/or mycotic spore were detected with Alcian-PAS stain. A therapy with intravenous steroids (metilprednisolone 500 mg for 3 days and then 75 mg for 7 days), intravenous immunoglobulins (300 mg for 5 days), and oral antibiotics (ciprofloxacin 500 mg twice a day for 7 days) was started, achieving remission of symptoms (autonomous breathing and stable hemodynamic values) within 2 weeks. Steroids were gradually decreased, and an immunosuppressive therapy with methotrexate 15 mg/week was started. At a 1-month follow-up, the skin was still completely clear from lesions and cardiopulmonary symptoms were resolved.

Discussion

An accurate review of literature through the PubMed search (1987–2013; terms: adult onset Still's disease, adult onset Still's disease and skin, neutrophilic urticarial dermatosis) was performed to retrieve data regarding the cutaneous findings of patients with AOSD. The mean age at diagnosis is 35 years, and the disease affects a slightly larger number of women as compared to men. The fever is usually more than 39 °C, typically quotidian or double quotidian in pattern, with the highest temperatures seen in the late afternoon or early evening; it is transient and its incidence is estimated at about 96 % according to the largest retrospective studies of literature [5–7]. Arthralgia and arthritis of big joints (oligoarticular and transient but sometimes evolving into a more severe, destructive, symmetrical, and polyarticular form) occur in the majority of patients with AOSD, with incidences ranging from 64 to 100 %, although it is not mandatory for diagnosis in adults [4, 8]. Other manifestations are hepatomegaly and elevation in liver enzymes (50–75 %) [4], sore throat (69 %) [4], cardiopulmonary involvement like pleuritis (26.4 %) or pericarditis (23.8 %) [6, 10], tamponade, myocarditis, pulmonary fibrosis, pleural effusions, and adult respiratory distress syndrome [4]. Laboratory test largely reflects the systemic inflammatory nature of the disease process, and none of the findings are specific for AOSD. An increased sedimentation rate is universal, and there is no association with rheumatoid factor or ANA. Patients frequently have a marked blood neutrophilia, probably secondary to bone marrow granulocyte hyperplasia [4]. According to the largest retrospective studies, the rash occurs in about 80 % of patients [1, 5–7, 9], being present from the initial onset of the disease in 28 % of patient [9]. The most common cutaneous manifestation is an evanescent salmon-pink or erythematous maculopapular eruption which frequently appears during febrile attacks and is predominantly found on the proximal limbs and trunk with rare involvement

of the face and distal limbs [1, 5–7, 9]. The rash can be mildly itchy or may be associated with burning sensation. In general, it lasts for hours and may change daily, enhancing with the fever spike and fading with the fever down; however in some patients, the duration of skin lesions correlates with the degree of systemic activity and may last for days or weeks without change [11–17]. However, other types of cutaneous manifestations have been described. The most frequent are persistent plaques and linear pigmentation [11–14], fixed plaques [15–20], and urticarial lesions [21–24]. The latter are characterized by "atypical wheals," persistent for more than 24–36 h, with symmetrical distribution [25]. Less common manifestations include angioedema [26], eczematous lesions [10], acne-like lesions [10], pustules on hands and feet [27, 28], persistent generalized erythema [29, 30], and flagellate erythema [31] (Table 1). Histological findings of classic skin lesions show normal epidermis overlying a mixed mild perivascular inflammation of the superficial dermis composed of lymphocytes and neutrophils [32, 33]. Persistent papules and plaques are histologically characterized by necrotic keratinocytes in the upper half of the epidermis with a neutrophilic infiltrate in the papillary dermis [33, 34]. In patients with urticarial lesions, the histopathologic findings show an intense neutrophilic infiltrate around the blood vessels but also with significant interstitial involvement. These elements are mature $CD15^{+}$ neutrophils and often have a peculiar disposition between the dermal collagen bundles, in single file or "en file indienne." This type of dermatosis has recently been described as a clinicopathological entity called neutrophilic urticarial dermatosis (NUD) [35]. Leukocytoclasia is present, but blood vessel walls are not damaged and therefore vasculitis can be excluded [35]. NUD has been found also in other diseases such as monogenic autoinflammatory syndromes, Schnitzler syndrome, and neutrophilic diseases like Behçet syndrome and Crohn's disease [35]. NUD is then considered a cutaneous marker of autoinflammation, which can unify more clinical entities under a common pathogenetic mechanism. Typical Still's disease rash needs to be differentiated from various other types of maculopapular eruptions, which are often drug- or infection-related. The histopathologic finding of a superficial dermal infiltrate containing neutrophils supports the diagnosis of Still's disease rash [33]. Eosinophils, which are commonly seen in drug eruption, are consistently absent in AOSD-associated skin lesions. The atypical persistent lesions, on the other hand, need to be differentiated from a variety of disorders that may manifest urticarial papules, erythematous to violaceous or pigmented lichenoid papules, or plaques clinically and necrotic keratinocytes histologically. The unique pattern of dyskeratotic cells, singly or in aggregates, mainly located in the upper epidermis, and the presence of a neutrophilic and lymphocytic infiltrate in the upper dermis appears very distinct and allows relatively easy differentiation from the disorders in which infiltrate is in band at dermo-epidermal junction [33]. Schnitzler's syndrome is a paradigm of an acquired/late onset autoinflammatory disease which needs to be differentiated from AOSD. The diagnostic criteria include a chronic-relapsing urticarial rash frequently leaving a brown hyperpigmentation and usually not pruritic. Histologically, it manifests with a neutrophilic urticarial dermatosis [8, 25], a monoclonal IgM component, and at least two of the following signs: fever (recurrent and above 40 °C), joint and/or bone pain, lymphadenopathy, spleen and/or liver enlargement, increased ESR, neutrophilia, and abnormal bone imaging findings [36]. The monoclonal IgM component is a defining feature of the syndrome and is present in all patients with Schnitzler's syndrome. The clinical presentation in our patient fulfilled the Yamaguchi's criteria for diagnosis of AOSD. The skin lesions were not the first sign of disease, but they prove to be essential for diagnosis. Moreover, exanthematous/urticarial manifestations reflected one of the most common skin patterns of AOSD and clinicopathological correlation allowed the correct diagnosis. Early recognition of AOSD can be particularly helpful during the initial workup when the clinical manifestations do not yet meet the Yamaguchi's criteria. A careful evaluation of skin lesions and a proper lesion selection for histological examination may be very important for the early diagnosis of AOSD. A clinicopathological correlation is then essential as both clinical and histological features are not pathognomonic.

Disclosure None.

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