Flagellate dermatoses associated with myocarditis in Adult-onset Still's disease

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Abstract

A 23-year-old woman followed for adult-onset Still's disease (AOSD) presented fever and chest pain. Clinical examination showed erythematous papules suggestive of flagellate dermatoses. Laboratory findings showed increased Cardiac troponin. Myocarditis due to AOSD was therefore suspected. The patient was treated with prednisone and methotrexate with significant clinical improvement.

Introduction

Flagellate dermatoses are nonspecific inflammatory skin manifestations characterized by linear or curvilinear streaks and plaques, occurring mainly on the trunk. They are rare and most often caused by Bleomycine or the intake of shiitake mushrooms. The association with adult onset Still's disease is rare. The patients with atypical skin manifestations of Adult-onset Still's disease (AOSD) like flagellate dermatoses had more severe and persistent disease with systemic complications. We report in this regard a case of flagellate dermatoses associated with myocarditis in AOSD.

Case report

A 23-year-old woman was reffered to internal medicine with a 3-week history of fever, asthenia and joint pain affecting her elbows, wrists, hands, knees and ankles. She also had history of odynophagia, loss of appetite and asthenia. However, there was no photosensitivity, malar rash or muscle weakness.

On physical examination, she had fever (39.4 °C), tenderness and mild swelling of wrists and interphalangeal joints of hands. There was diffuse macular erythematous eruption that appears with the spikes of fever mainly on the neck, trunk and upper limbs. Laboratory studies showed leukocytosis (30350/uL, 95% neutrophils), anemia (9.3 g/dl), increased levels of C-reactive protein (286 mg/L), high erythrocyte sedimentation rate (150 mm) and a very high serum ferritin (18000 ng/mL). The results of liver function tests were within normal limits. Tests for antinuclear antibody, rheumatoid factor, and anti-cyclic citrullinated peptide antibody were negative. Parvovirus B19 immunoglobulin M (IgM) titers were normal. Findings for cytomegalovirus, human immunodeficiency virus, and hepatitis were negative. Urine cytobacteriological examination was normal as well as Chest radiograph, abdominal ultrasound and echocardiography.

The diagnosis of AOSD was considered in view of clinical presentation : fever and polyarthritis without evidence of infection, autoimmune disease or malignancy. Her clinical and laboratory parameters fulfilled the Yamaguchi's diagnostic criteria (fulfilled four major and two minor criteria) for the disease. The patient was treated with oral prednisone 60 mg once daily with resolution of all symptoms.

Two months later, the patient experienced the recurrence of fever, arthralgia associated with odynophagia and chest pain while she was off treatment. On physical examination, she had spiking fever (39.4 °C), tenderness of wrists, hands, knees and ankles. Cardiopulmonary auscultation was without abnormalitis.

There was no organomegaly or lymphadenopathy. Skin examination showed macular erythematous eruption appearing with the spikes of fever associated with fixed linear erythematous streaks on the trunk, lower back and stomach with a flagellated arrangement compatible with flagellate erythema (Figure 1,2).

The laboratory findings showed leukocytosis : 20260/uL, neutrophils 19900 e/mm^3, increased levels of C-reactive protein (480 mg/L), thrombocytosis (Plq=1328 000 e/mm^3) and increased Cardiac troponin 849 nmol/mL. The electrocardiogram showed bundle branch block. Electrocardiography and cardiac MRI were normal. Chest computed angio-tomography demostrated only minimal bilateral pleural effusion, without pulmonary embolism. An infectious disease was ruled out. Myocarditis due to AOSD was suspected. The patient was treated with Bisoprolol, prednisone 1 mg per kg daily and methotrexate 10 mg per week. The patient became afebrile with significant clinical improvement in rash and flagellate dermatoses, fever and chest pain. There was also normalisation of all biological parameters including cardiac troponin. After four years of follow-up, the progression was favorable despite tapering steroids dosage.

Discussion

AOSD is a systemic inflammatory disorder of unknown etiology and pathogenesis that usually affects young adults with an estimated prevalence of 1.5 cases per 100,000–1000,000 people [1].

Clinical manifestations include spiking fever, arthralgia or arthritis, serositis, transient cutaneous manifestations, lymphadenopathy and hepatosplenomegaly. The main differential diagnoses are infections, neoplasms and autoimmune disorders [2].

The typical skin findings are an evanescent, non pruritic salmon-pink maculopapular eruption that appears concomitantly with fever spikes and subsides when fever resolves. It occurs in 60–80% of patients and is predominantly found on the proximal limbs and trunk with rare involvement of the face and distal limbs [3].

In recent years, atypical cutaneous manifestations of AOSD have been reported often in addition to the typical evanescent rash in 14% of cases but may be the only skin manifestation [4]. Flagellate dermatoses are uncommon figurate dermatoses characterized by parallel linear or curvilinear arrangement simulating the marks of whiplashes. Persistent pruritic papules and plaques some with a flagellate appearance resembling our patient are a well-described feature of chikungunya, bleomycin-induced, dermatomyositis, shiitake mushroom and poisoning [5]. It is a rare cutaneous feature in AOSD.

Other commonly reported cutaneous manifestations are non pruritic persistant erythema, urticarial or lichenoid papules, pigmented plaques and prurigo pigmentosa–like [6-7]. Atypical skin findings may appear at any time over the course of the disease but In the great majority of cases at the time of disease onset concurrently with systemic symptoms, or shortly afterward [4].

It was reported that majority of the patients with atypical skin manifestations of AOSD had more severe and persistent disease. These patients develop complications like pericarditis, myocarditis, serositis, lung involvement, neurological involvement and reactive hemophagocytic syndrome [8]. The same findings were observed in our case since flagelate dermatoses was associated with probable myocarditis. Cardiac involvement in adult-onset Still's disease (AOSD) usually manifests as a pericardial disease (up to 37% of cases) while myocarditis is uncommon with a prevalence of 7% [9].

Patients with atypical skin manifestations have also a worse prognosis with a mortality rate of around 8% [10]. Majority of the patients with atypical manifestations of ASOD require moderate to higher doses of glucocorticoids. Around 40% of these patients require immunosuppressant therapy like methotrexate, azathioprine, cyclosporine and hydroxychloroquine [11]. Our patient was treated with prednisone and methotrexate because of suspected myocarditis with significant clinical improvement of chest pain and flagellate dermatosis.

Conclusion :

Our case illustrate a rare presentation of AOSD with flagellate dermatosis and probable myocarditis. Atypical skin findings such as flagellate dermatosis have been reported in AOSD and may be associated like other

atypical skin manifestations to systemic complications and worse prognosis. Awareness of nonclassic skin findings in AoSD may lead to earlier diagnosis of systemic complications.

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