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A 7 Years Old Girl With Maculopapular and Erythematous Rash, Fever and Hypocomplementemia

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ABSTRACT

Cutaneous eruptions and arthralgia in children can occur after infections, drugs and immunologic processes via different mechanisms.
This is a report of a 7 years old girl with a history of maculopapular rashes that changed to target-shaped lesions and pruritus with non-pitting edema of ankles, strawberry tongue and periungual scaling of extremities, with no history of any drug usage. She had elevated liver enzymes and positive anti-viral capsid antigen (VCA) (IgM and IgG), and depressed C3, C4 and CH50, that returned to normal after 3 months.
Here we explain the states that could cause similar clinical scenario and discuss them briefly.

Implication for health policy/practice/research/medical education:
Skin eruptions and fever are common manifestations in pediatric group , complete physical exam and logical laboratory investigations helps us to correct diagnosis and prevent us from unnecessary drug administration.

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1. Introduction

Fever and rash are common symptoms in childhood, most of them have self-limited behavior and in most situations common viral infections are responsible. Recurrence of cutaneous rash and appearance of other musculoskeletal or systemic manifestation may raise the clinical suspicion to rheumalogic disorders (1-3). Recognition and clinical follow up patterns are essential for diagnosis, in some pediatric practical cases single diagnostic and positive tests in absence of disease would not make diagnostic difficulties however variety of clinical and cutaneous manifestations in viral disease should be considered in order to avoid unnecessary diagnostic tests (4). In the present study a seven years old girl with clinical and laboratory signs were subjected to our study without any attribution to a specific diagnosis.

2. Case Presentation

A 7 years old girl was admitted in Mofid Children's Hospital, with a history of 11 days of maculopapular rashes that progressed to target shaped and erythematous lesions. They started from hands and feet with trunk extension. Fever and severe itching also developed from 8 days

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before admission. Pain and non-pitting edema of ankles appeared a few days before admission. She had intermittent abdominal pain without nausea, vomiting, diarrhea or bloody stool.

She was the second twin of a 30 years old mother with 2050 g birth weight, normal growth and development and a complete vaccination history with no history of admission.

At physical examination, she had erythema and fissuring of lips, strawberry tongue, patchy scattered maculopapular and erythematous rashes throughout body with target shape lesions in a few of them, and fine scaling at extremities (*Figure 1*). She had not any significant lymphadenopathy. Ear and throat examinations were normal but mild epigastric tenderness was present. Ophthalmologic examination was normal, there was a mild pericardial effusion in echocardiography. ECG was normal. Sonography revealed mild ascites and splenomegaly. Chest X-ray showed left costophrenic angle hazziness and paracardiac lung infiltration in right side. Laboratory investigation showed: (normal ranges are in parenthesis)

Hgb, 14.2 mg/dL (11.5-15.5); WBC, 11000/mm^3 (5500-15500); Polymorphonuclear, 38%; Lymphocyte, 60%; Eosinophile, 2%; PLT, 87000 /mm^3; ESR, 2; PT, PTT normal; CRP, 3+; LDH, 390 IU/L (Up to 430); SGOT, 67 IU/mL (5-45); SGPT, 60 IU/mL (5-45); Alk Phosphatase, 284 IU/mL (NL) and blood culture was negative. Complement level was low in admission and returned to normal 3 months later, Anti CMV and Anti-Herpes antibody was negative. Anti-viral capsid antigen (VCA) antibody was positive and she had an acute Epstein Bar virus infection (*Table 1*). All physical examination and radiographic findings were normal after three months.

3. Discussion

Fever and rash are common symptoms in childhood and may occur due to drug hypersensitivity syndromes, henoch schoenline purpra, collagen vascular disease, urticarial vasculities, lymphoproliferative disorders, serum sickness and serum sickness like and some infectious diseases (1-3).

In every patient with such complaints, seeking for special physical findings or laboratory results can guide to the etiologic cause, our patients with straw berry tonque and scaling in extremities, the first diagnosis could be Kawasaki disease but no history of prolonged fever, with normal ESR and thrombocytopenia is unusual in this syndrome, so the patient's complaints did not fulfill the classic criteria of Kawasaki (4, 5). Cutaneous lesions are erythema multiform like and target lesions so other causes such as scarlet fever (rough skin with goose-pimple appearance and pallor around the mouth) or toxic shock syndrome (rough erythematous skin) is less possible because of their specific rash (4).

Serum sickness and a very similar state; serum sickness like reaction (SSLR); is a possibility in this girl with rash, fever and joint swelling accompanied with low complement level. Serum sickness occurs after injection of antigens like anti-thymocyte globulin, tetanus vaccination, insect venom and antibiotics (6-8) and SSLR occurs 7-15 days after consumption of drugs like sulfonamides, macrolids, ciprofluxacin, bupropion, antidepressants (Fluoxetine), anti-inflammatory drugs, beta-lactams (cefaclor) (9) and rifampin and anti epileptic drugs (10-12) and oral penicillin (13, 14) and also after viral and streptococcal infections and a variety of vaccines. The pathogenesis of serum sickness is immune complex mediated but In SSLR, the pathogenesis is not well understood, although it likely does not depend upon high titers of antibodies and circulating immune complexes, as in classical serum sickness, but in cefaclor, the metabolites production that are genetically influenced are toxic for lymphocytes so this disorder has familial distribution (15).

3.1. Another Condition

Urticarial vasculitis is an interesting disorder first brought to light by McDuffie *et al*., (16) is a systemic disease with a longer-lasting (3-7 days) urticaria. They are often painful or 'burning' and leave residual bruising or hyper pigmentation.

40% of patients with urticarial vasculitis will have associated angioedema, pain and non-pitting edema of both ankles in this girl can be angioedema but the median age of incidence is 43 years. In most cases urticarial vasculitis

Table. Laboratory Test in Admission and Three Months Later

Test Results	Admission	3 Months Later
C3 (89-187)	43 mg/dL	95 mg/dL
C4 (16-38)	7 ng/dL	20 ng/dL
CH50 (90-100)	< 80	95
Anti VCA [a] IgG Pos [a] > 20	148 IU/mL	
Anti VCA [a] IgM Pos [a] > 40	100 IU/mL	

[a] Abbreviations: Pos, positive; VCA, viral capsid antigen antibody

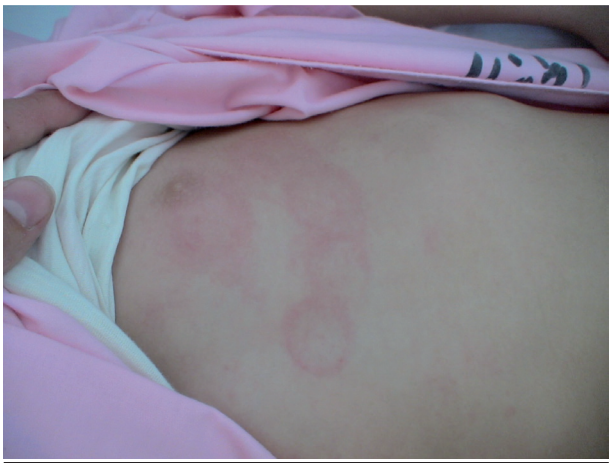

Figure. Cutaneous Rash in Seven Years Old Girl With Fever and Hypocomplementemia

is idiopathic, but it may be associated with connective tissue diseases such as SLE or Sjogren's syndrome; infections such as hepatitis B and C, Lyme disease and infectious mononucleosis; treatment with drugs, including angiotensin converting enzyme inhibitor (ACEI), cimetidine, diltiazem, penicillins, sulphonamides and thiazides; and lymphoproliferative diseases such as mixed cryoglobulinaemia and IgM gammopathy (16, 17). Two categories of urticarial vasculitis are hypo-complementaemic and normocomplementaemic (18).

Patients with hypo-complementaemic urticarial vasculitis syndrome (HUVS) are more likely to have an associated connective tissue disease and systemic symptoms such as fever, arthralgia, gastrointestinal involvement, pulmonary disease, and glomerulonephritis, progressive renal disease is associated with SLE. Other rare manifestations include eye involvement, lymphadenopathy, splenomegaly and pericardial effusions (19) and may have IgG antibodies to the collagen-like domain of C1q (20).

In this patient low complement level and positive Anti VCA antibody shows a systemic reaction which can be compatible with serum sickness or urticarial vasculitis, but normal complement level after 3 months and no residual systemic manifestations indicates the self-limited nature of problem. She had no history of drug usage, so we can find individual; patients which cannot be categorized precisely in one diagnostic dilemma, EBV is the responsible pathogen in this patient and patients manifestations subsided after disease process, then we can think of SSLR in this specific case. The patient's complaints eliminated in about two weeks and the antibodies titers decreased during following months.

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Author's Contribution

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