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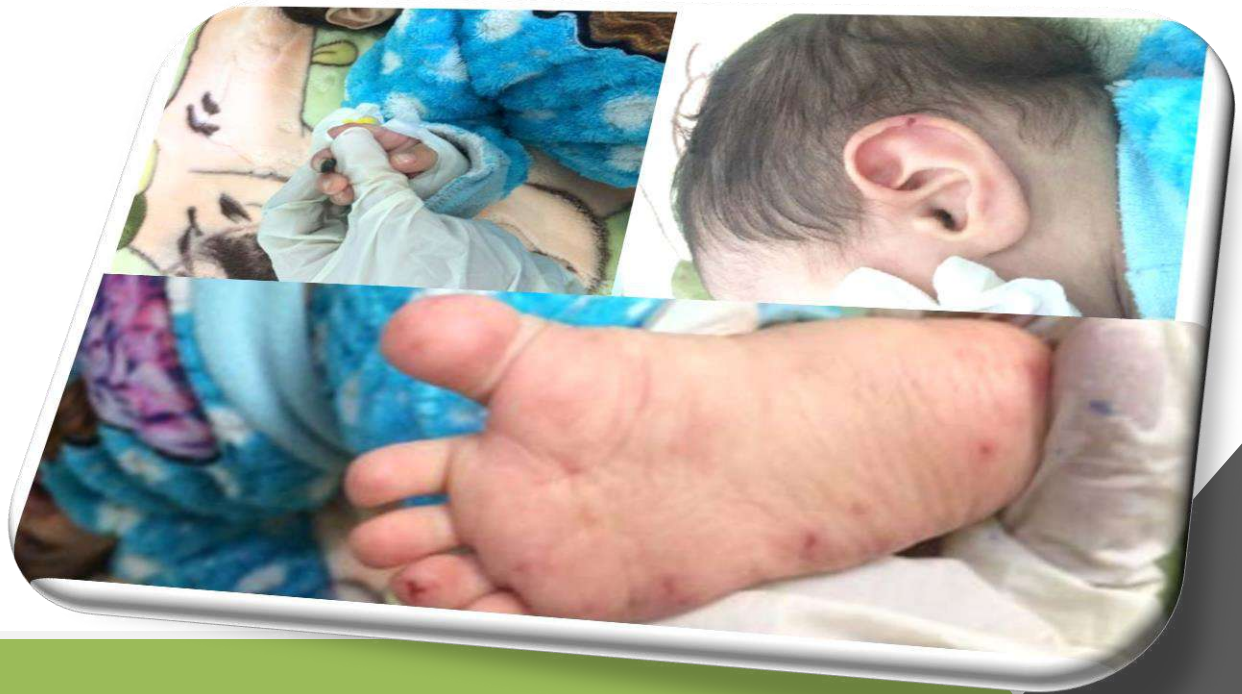

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ContusM Publications

A Rare Presentation of Systemic Lupus Erythematosus in an Infant- A Case Report

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Abstract

Background: SLE is a very common autoimmune disease in children with varied presentations. It is extremely rare in infancy. Digital ischemia and gangrene can be an initial presentation in this age group.

Aim: To represent uncommon clinical manifestations of a common autoimmune disease.

Objectives: In this we report a case of infantile onset SLE who presented initially with rare presentations and unusual clinical course.

Case Report: An 8-month-old girl presented with digital discoloration of the middle finger of her right hand and vasculitic skin manifestations. She had positive results of both anti-nuclear antibodies and antibodies to double stranded DNA with low C3 and C4 complements. The infant was treated with prednisolone for about six months. She was followed up for one year at our nephrology clinic until she was free of symptoms, with negative anti- nuclear antibodies and antibodies to double stranded DNA.

Conclusion: This case emphasized the need to think about rare symptoms of common diseases like SLE that can present in infancy and the importance of prompt treatment of such critical condition like digital ischemia.

Keywords: SLE in infant; Digital gangrene; Skin manifestations; Vaculities

Abbreviations

SLE: Systemic Lupus Erythematosus

cSLE: childhood-onset Systemic Lupus Erythematosus

ANA: Anti-Nuclear Antibodies

Anti-dsDNA: Antibodies to Double Stranded DNA

SLICC: Systemic Lupus International Collaborating Clinics

Introduction

Systemic Lupus Erythematosus (SLE) is a multisystemic autoimmune disease in which the immune system attacks itself by the production of autoantibodies and immune complexes leading to inflammation and may be damage to many organs [1]. The hallmark of SLE is the variability in clinical phenotypes and the production of multiple autoantibodies that can unpredictably affect any organ and system in the body [2].

SLE prevalence in children and adolescents (1-6 / 100,000) while in adults (20-70/100,000) [3]. It occurs between the ages 3 and 15 in pediatric age group, with the girls to boys ratio 4:1. The presentations of childhood-onset SLE (cSLE) are often rare, strange and sever compared to adults [4].

Skin symptoms in children with SLE may be peculiar, rare and unpredictable as well [5]. Some of these symptoms have been described in studies as Raynaud phenomenon, gangrene, periungual erythema, nail problems, and subacute discoid lupus erythematosus [5]. Often, they are the primary symptoms in adults but they are rarely the first and only symptoms in children [6].

Digital gangrene was mentioned in numerous reports in children and adults with SLE but it is extremely rare in infancy [7].

Case Presentation

Patient Complaint

An 8 months old girl was brought to our nephrology clinic with the compliant of blackish discoloration, ulceration and

sloughing of skin of the tip of the middle finger of her right hand with small red papules on her ear lobes.

Sudden bluish (cyanotic) discoloration was noticed on the finger's tips of both hands one week before attending nephrology clinic, followed by sloughing, ulcerating. The condition associated with purpuric skin rash on both hands and feet. The finger changes healed spontaneously in one week duration without traces, except for the middle finger of her right hand which persisted.

Past Medical History

Two months earlier, she was admitted into the communicable disease ward, with complaint of high-grade intermittent fever relieved by antipyretic, decreased activity and poor feeding with noticeable weight loss, considered as a case of sepsis and was treated with empirical antibiotics for two weeks.

Asking the mother about her child's past medical history, she recalled the appearance of red papules on the right and left side of the scalp (at age of 3 months), that have been faded spontaneously then returned one month later but bigger than the previous one which also been faded without treatment. (No available photos for the lesion at that time unfortunately).

She has no history of photosensitivity, no history of joint pain, no oral ulceration. Her pre-natal, natal history was uneventful. No history of consanguinity. She lived in a rural area.

Past family history was free from any chronic diseases. No history of trauma.

Examination

Upon admission and on examination: She had ill looking and febrile, her heart rate was 100/min, respiratory rate was 20/min, and her blood pressure was 85/60 mm of Hg. Her Weight was 6.5 kg which is below 5th percentile. Her length was 63 cm below 5th percentile. OFC was 42.5 cm which is on 25th percentile. Her general and systemic physical examination was normal except for a small blackish discoloration and sloughing of skin of the tip of the distal phalanx of the middle finger of her right hand without a line of demarcation with normal arterial pulsation (Figure 1). A small palpable non-itchy (1/2 cm in diameter) red papules on her ear lobes (Figure 2). Multiple small, non-scaly, palpable erythematosus purpuric rash all over both hands and feet (Figure 3). All peripheral pulses were palpable.

Figure 1: Blacked, ulcerated and sloughed tip of middle finger of right hand.

Figure 2: A small red papules over the ear lobe.

Figure 3: Erythematosus purpuric and petechial rash over feet.

Investigation

Hematological investigations were as the following: Her complete blood count showed haemoglobin-10.7g/dl total leucocytes count - 7.7*10^9/L (neutrophil-78%, lymphocyte-16%) and platelets were 500*10^3/L. Blood film slide showed microcytic hypochromic anemia with reticulocyte count was 1.5%. ESR was 80 mm in 1st hour (elevated). PT 11 (n.13), INR 1 (n.1.2), PTT-23 (n.31). Negative coomb's test. Prothrombotic workup including antithrombin C, protein C and S were not done.

Renal indices and electrolytes were as the following: Urea -9 mg /dl creatinine-0.5 mg/dl, sodium - 145 meq/L, potassium -4.2 meq/L. Her 24-hour urine collection for protein was 125 mg. liver function test shows: SGPT-45 IU, SGOT- 40 IU.

Septic screen: C - reactive protein -55 mg/dl (high), General urine examination was normal with no growth of any bacteria in urine and blood culture. Lumber puncture for cerebrospinal fluid analyses was normal.

Immunological tests were as the following: Her Antinuclear antibody level was 1.6 IU (>1.2+ve) (homogenous pattern), Anti ds DNA IgM positive 41.3 IU (>30 +ve), Anti ds DNA IgG positive 98.4 IU (>30 +ve), and C3 84 mg/dl (90-180), C4 2 mg/dl (10-40) [1]. ANCA and other lupus serology markers as antiphospholipid antibody, anticardiolipin antibody, lupus anticoagulant was negative also. Viral screen for HIV and hepatitis were negative.

Imaging

Chest x-ray, abdominal ultrasonography, and Doppler ultrasound of both upper and lower limbs were normal. Echo study was normal.

Diagnosis

After discussion with a multidisciplinary team that included: a general pediatrician, nephrologist and pediatric surgeon, a diagnosis of SLE was made with the view of her critical digital pregangrenous condition in addition to general ill health along with her investigations. The patient was diagnosed as SLE with Systemic Lupus International Collaborating Clinics (SLICC) criteria. The patient had fulfilled four criteria with one clinical criterion as she had cutaneous lesions along with digital ischemia, ulceration and pregangrenous changes in her digits with skin lesion in her earlobe with the presence of three immunological criteria as she had ANA above laboratory reference range, Anti-dsDNA was also above laboratory reference range with low complements (low C3 and C4).

Line of Treatment

The child was started on steroid, she was treated with prednisolone (2 mg/kg/day) with hydroxychloroquin (5-7 mg /kg /day) [3]. During the hospital stay, pain subsided and the pregangrenous skin discoloration in the right hand resolved and there were no other vasculitic changes. During weekly follow up visits she was well.

On her follow up visit after 1 month, she was asymptomatic without any skin manifestations and she was a healthy infant. Prednisolone was gradually tapered to 1 mg/kg/day with the subsequent follow up visits during the next 3 months [3].

Within the treatment plan, there were consultations for parents and constant recommendations to avoid exposure to sun light or cold weather. Vitamin D drops as 400 IU was added to her supplements.

Six months later, anti- nuclear antibodies and antibodies to double stranded DNA were negative, so prednisolone was stopped after gradual tapering.

Over the next follow up visits, the patient had normal clinical examination and investigations. The patient is still being followed up.

Discussion

We report an infant girl with digital and skin manifestations as the only presenting features and her investigations showed positive anti- nuclear antibodies and antibodies to double stranded DNA with low complements. These rare presentations were diagnosed as a case of infantile SLE.

The etiology beyond skin and digital involvement as peripheral gangrene in SLE is multifactorial and may include many causes as antiphospholipid syndrome, vasculitis, infectious disorder, early atherosclerosis, vasospasm and thromboembolic phenomena due to the presence of antiphospholipid antibodies or hypercoagulability. Digital and skin manifestations without these etiologies are very rare in SLE [8-10]. Antiphospholipid syndrome and thromboembolic phenomena were excluded by clinical and laboratory workup [11].

Gangrene of the digits especially of the upper extremities is extremely rare, occurring in about 1% of SLE patients [7]. Gangrene in children with lupus has been described by several authors, but in the present case, the age of onset was very early at 8 months [7].

Although digital gangrene has been seen in many adult cases as an initial presentation of SLE, it is rarely reported in

pediatric age group [5,7].

One of the challenges in the management of such critical case was the diagnosis of the exact cause of ischemia or gangrene whether it was due to vasculitis as it is the main cornerstone in SLE usually of small vessels or the cause was antiphospholipid syndrome or hypercoagulability state and thrombosis that were excluded by history, clinical examination, investigations and imaging [11].

The diagnosis went mostly with vasculitis depending on SLICC criteria that taking in consideration clinical and serological findings [10]. Although histopathology can help us in diagnosis, it wasn't available at time of the case presentation and admission in our hospital [11].

Starting steroid with hydroxychloroquin earl was very helpful in controlling disease activity and prevents progression of gangrene and save the infant from the risk of digital autoamputation [3].

Conclusion

SLE is a rare disease in infancy and it is considered as a challenge because it is difficult to diagnose and to manage.

In our case, after reaching an accurate diagnosis as a case of SLE, prednisolone was started along with Hydroxychloroquin. Obviously, she responded to the dual treatment by the disappearance of the digital discoloration, other skin changes and general good health condition.

This case emphasized the need to think about rare symptoms of common diseases like SLE that can present in infancy and the importance of prompt treatment of such critical condition like digital ischemia.

Digital ischemia and gangrene in children with SLE have been described in many literatures but very few reported cases in infancy. Shetty et al. [7] reported the first case of SLE presenting with foot and hand gangrene in a neonate and an infant of 11 months old [5,7]. Ziae et al. [5]. Reported a 12 years old female with peripheral gangrene as rare presentation of systemic lupus erythematosus in a child [7]. While Tareq Z. Alzughayyar et al. [10] reported a 10 years old girl with SLE and multiple autoimmune diseases and had extensive peripheral gangrene [10].

Recommendations

We recommended SLE evaluation in all children with vasculitic symptoms. We also emphasized on proper treatment of critical digital and skin manifestations which can prevent the ischemic harm to the digit and prevent progression of gangrene. The recommended treatment for digit and skin involvement is steroids.

Ethical Clearance

Permission was taken from the patients' parents for taking photos and publishing the article.

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