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Case Report *Volume 4 Issue 1*

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

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Abstract

An 8 month old girl presented with digital discoloration of the middle finger of her right hand and vasculitic skin manefestations. She was found to have positive nuclear antibodies and antibodies to double stranded DNA, and low C3 and C4 complements.. The infant was started on oral prednisolone, which was discontinued after six months. At one year of follow up she was asymptomatic, with negative nuclear antibodies and antibodies to double stranded DNA.

Conclusion: We are presenting this case to highlight the importance of knowledge of very rare manifestations of a common disease like SLE at presentation in infancy and treatment of critical digital ischemia.

Keywords: SLE in Infant; Digital Gangrene; Skin Manifestations; Vasculitis

Background

Systemic lupus erythematosus (SLE) is the prototype of systemic autoimmune diseases characterized by the production of autoantibodies and immune complexes leading to protean systemic manifestations [1].

The reported prevalence of SLE in children and adolescents (1-6 / 100,000) is lower than that in adults (20-70/100,000) [2]. The onset of childhood SLE occurs between the ages 3 and 15, with the girls outnumbering boys in the ratio 4:1.The clinical manifestations of childhood-onset SLE (cSLE) are diverse, severe and often atypical as compared to the adults [3].

SLE in children has many manifestations .Cutaneous manifestations were shown to be a rare manifestations of SLE in children [4].

In several studies on SLE in children, Raynaud phenomenon, gangrene, periungual erythema, nail problems, and subacute discoid lupus erythematous, have been described as a rare manifestation of SLE.4 In adult patients with SLE, these rare symptoms have been described as the presenting symptoms, but they have not been described as the first and only presenting symptoms of systemic lupus erythematous in children [5].

Although there have been reports of digital gangrene in children and adults with SLE, peripheral gangrene in an infant with SLE is extremely rare [6].

Case Report

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. Two months earlier, she was admitted into the communicable disease ward, with complaint of high grade intermittent fever

relieved by antipyretic, decrease activity and poor feeding with noticeable weight loss, considered as a case of sepsis and was treated with empirical antibiotics for two weeks. Sudden bluish (cyanotic) discoloration was noticed on the fingers tips of both hands followed by sloughing, ulcerating and an associated purpuric-petichial skin rash on both hands and feet. All the pregangrenous changes resolved without any intervention and with no sequels, except for the middle finger of her right hand.

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.

Apart from these complaints there was no history of photosensitivity, joint pain, oral ulcer and seizure. The patient was a product of normal pregnancy and delivery of non-consanguineous marriage, lived in a rural area. Past history and family history was noncontributory. No history of trauma.

At admission child was febrile with temperature-38 C, heart rate was 100/min, respiratory rate was 20/min, blood pressure was 85/60 mm of Hg and capillary refill time was <3 second. 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. General physical examination was normal except for a blackish discoloration and sloughing of skin of the tip of the middle finger of her right hand without a line of demarcation with normal arterial pulsation (Figure 1). A small (1/2 cm) red papules on her ear lobes (Figure 2). Purpuric and petechial rash all over both hands and feet (Figure 3). Systemic examination was apparently normal. All peripheral pulses were palpable.

Figure 1: Blacked, Ulcerated and Sloughed Tip of Middle Finger of Right Hand.

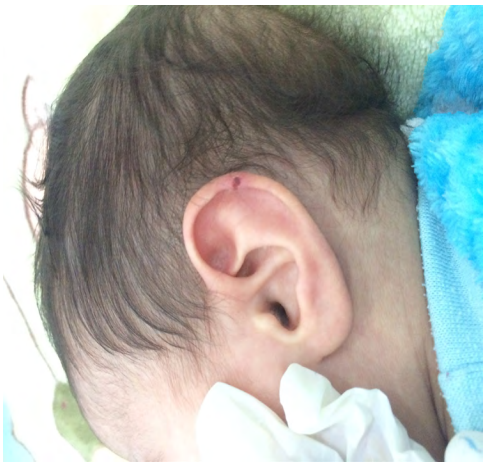

Figure 2: Small Red Papules over the Ear Lobe.

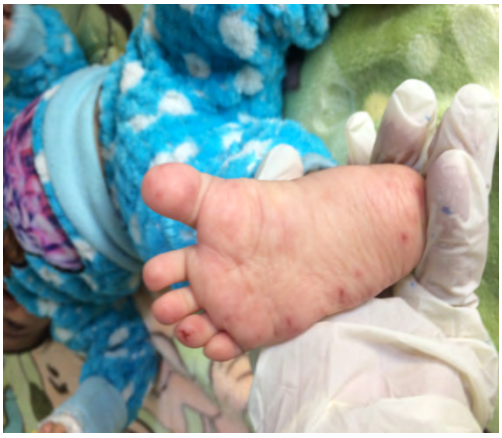

Figure 3: Purpuric and petechial rash over feet.

Her investigation showed haemoglobin-10.7g/dl total leucocyte count – 7.7*10^9/L (neutrophil-78%, lymphocyte-16%) and platelets were 500*10^3/L. Peripheral smear showed microcytic hypochromic anemia with reticulocyte count was 1.5%. PT 11(n.13) INR 1 (n.1.2), PTT-23(n.31). Urea-9mg /dl creatinine-0.5mg/dl, sodium-145meq/L, potassium -4.2 meq/L. SGPT-45 IU, SGOT- 40 IU. C - reactive protein -55 mg/dl (high), ESR was 80 mm in 1st hour (elevated). Urine routine and microscopic examination was normal and urine and blood culture was sterile. 24 hour urine protein was 125 mg. 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) l. Viral markers (HIV, HbsAg, and HCV) were negative. ANCA, Rheumatoid factor was negative. Work up for ant phospholipid antibody syndrome (anticardiolipin antibody, lupus anticoagulant) was negative. Negative comb's test.

Prothrombotic workup including ant thrombin C, protein C and S were not done.

Chest radiographs, electrocardiographs, renal and abdominal ultrasonography, and cerebrospinal fluid analyses were normal. Doppler ultrasound of both upper and lower limbs was reported as normal.

A diagnosis of SLE was made and with the view of critical digital pregangreanous condition in addition to general ill health and her investigations, the child was started on steroid, She was treated with prednisolone (2 mg/kg/day) along with hydroxychloroquin (5-7 mg /kg /day) [2]. During the hospital stay pain subsided and the pregangrenous changes in the right hand disappeared and there was no further progress of the vasculitic changes. During weekly follow up visits she was well.

After 1 month she was symptoms free in the extremities with very good general condition .The dosage of prednisolone was reduced to 1 mg/kg/day with gradual tapering and her condition has been under control for 3 months [2].

In treatment plan, proper counseling was done to the child's parents with strict avoidance of sun exposure and cold exposure. Vitamin D supplements were further added to the patient.

Prednisolone was discontinued after six months as nuclear antibodies and antibodies to double stranded DNA were negative.

Over the next year, the patient showed improvement in her clinical parameters and laboratory indices. The patient is still being followed up.

Discussion

We described an infant girl with digital and skin manifestations as the only presenting features and later on investigations found to have positive for SLE but negative for ant phospholipid antibody.

The etiology of digital involvement in SLE is complex and may involve several factors including Ant phospholipid Syndrome, vacuities, premature atherosclerosis, vasospasm and thromboembolism. Digital and skin manifestations without these etiologies are very rare in SLE [7].

Gangrene of the extremities is very rare, occurring in about 1% of SLE patients, and most often affects the upper extremities [8]. 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 [8].

In adults there are many cases of digital gangrene as an initial manifestation in SLE at presentation but in pediatric age group only few cases report [8,9].

We also emphasized on aggressive treatment of critical digital ischemia which can save the ischemic digit and prevent progression of gangrene.

Conclusion

SLE in pediatric age group is a challenging disease both difficult to diagnose and to treat.

In our case, after confirmation of diagnosis of SLE, we began treatment with prednisolone and Hydroxychloroquine. She appeared to respond to this combination, judging by the disappearance of the digital discoloration, other skin changes and general good health condition.

This case highlights the importance of precise management and awareness of very rare manifestations of a common disease like SLE. Gangrene can be initial symptom of SLE in children.

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 vacuities is steroids.

Conflict Interest: None

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Ethical Clearance: Permission was taken from the Patients' Parents for Taking Photos and Publishing the Article.

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