An 11-year-old Girl with Burning Sensation in Her Hands

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An 11-year-old girl presented with burning sensation, high temperature, and erythema involving both hands, especially the right hand, along with the right arm (see Figure). These complaints had been present for the past 4 months, getting more severe in hot weather and with exercise, while showing some relief with cold application and with rest.

Systemic questioning did not reveal morning stiffness, rash, alopecia, dryness in eyes or mouth, or bloody diarrhea. Only occasional pain in joints of hands and feet were described. Family history was uneventful.

In physical examination, vital signs were within normal ranges. Widespread erythema was present on both hands, with a more reticular pattern on the dorsal part of the right forearm. Fingers were edematous, and periungual erythema was present on the fingers of right hand. Persistent symptoms of burning sensation and temperature eleva-

Figure. An 11-year-old girl presented with burning sensation, high temperature, and erythema involving both hands, especially the right hand, along with the right arm.

Editor’s note: Each month, this department features a discussion of an unusual diagnosis in genetics, radiology, or dermatology. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via e-mail at pedann@slackinc.com.
tion, along with local vasodilation and hyperemia, which are activated by environmental heat and exercise and are relieved by cold application and rest, suggested erythromelalgia. Treatment with misoprostol was begun, considering the low rate of adverse effect of the drug and to avoid masking any probable underlying collagen disorder.

Laboratory evaluation for differential diagnosis and classification of erythromelalgia yielded the following results:

- Hb 13.3 g/dL;
- leukocyte 5000/µL;
- platelet 134,000/µL;
- urine specific gravity 1010;
- pH 6.0;
- protein (1+);
- blood (-);
- 2-3 WBC/hpf;
- glucose 82 mg/dL;
- BUN 21 mg/dL;
- creatinine 0.55 mg/dL;
- albumin 4.6 g/dL;
- LDL 160 mg/dL;
- creatinine 0.55 mg/dL;
- albumin 4.6 g/dL;
- LDL 160 mg/dL;
- HDL 77 mg/dL;
- triglycerides 68 mg/dL.

Electromyography and upper extremity and renal Doppler ultrasonography were normal. Serologic tests revealed hypocomplementemia as C3 74 mg/dL (82-173 mg/dL), C4 9.6 mg/dL (13-46 mg/dL), and positivity in ANA (1/160); anti-dsDNA (1/20), pANCA and anticardiolipin IgM (40 MPL/mL), and IgG (24 GPL/mL) antibodies. Other laboratory results were erythrocyte sedimentation rate (ESR): 42 mm/h, CRP: 1.2 mg/dL (0.1-8.2 mg/dL), rheumatoid factor: negative, extractable nuclear antigen panel: negative.

Steroid treatment with 2 mg/kg/day prednisolone was started, and misoprostol was stopped. On the seventh day of prednisolone treatment, erythema, pain, and edema in the right forearm decreased significantly, while ESR regressed to normal values. Prednisolone was decreased to alternate day doses at the end of 4-week treatment course, and acetyl salicylic acid (100 mg/d) was added.

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DISCUSSION
A patient with complaints of burning sensation, erythema, and an increase in temperature of the extremities, relieved by cold application and rest, should raise the suspicion of erythromelalgia. A careful evaluation in such a case is needed for differential diagnosis of some clinical conditions, such as Raynaud's syndrome, burning feet syndrome, reflex sympathetic dystrophy, angiodypskinesis, acrocyanosis, lipodermatosclerosis, and erysipelas.1,2

Five criteria suggested by Thompson et al. are used in the diagnosis of erythromelalgia:3 burning pain in the extremities; increase in pain in exposure to high environmental temperature; decrease in pain by cold application; erythema in the affected body area; and increase in temperature in the affected skin region. All of these were observed in this child, and she was diagnosed with erythromelalgia.

Erythromelalgia can be primary but also may appear secondary to another disease. Therefore, other underlying clinical conditions that might be the cause of erythromelalgia should be properly excluded, such as drugs (verapamil, nicardipine, bromocriptine, pergolide, mercury); vasculitis syndromes; cardiovascular diseases (hypertension, venous insufficiency); hematologic disorders (pernicious anemia, thrombocytopenia, thrombotic thrombocytopenic purpura, polycythemia, hereditary spherocytosis, leukemias); neuropathies (polyneuropathy, hereditary sensory neuropathy, central nervous system lesions); infections (syphilis, AIDS); autoimmune diseases; musculoskeletal disorders; cholesterol crystal embolism syndrome; diabetes mellitus; hypercholesterolemia; gout and familial nephritis; conversion disorder, and malignancies.1-6

In this patient with erythromelalgia, history of arthralgia, hypocomplementemia, high ESR, along with low CRP, ANA and anti dsDNA positivity, and complaints that affect the life quality, there were suggestions of erythromelalgia secondary to SLE, although the lupus criteria were not completely fulfilled. However, the diagnosis of SLE is a dynamic process. Thus, a patient may fulfill the SLE criteria in time.

The dramatic improvement of the patient in response to steroid treatment provided further support for SLE. It is probable that, when the dose of steroid is decreased or stopped completely, the patient will develop other SLE manifestations. Thus, the patient should be in close follow-up for other clinical findings and complications of SLE.

Primary erythromelalgia can be quite resistant to treatment, although the symptoms in the secondary type usually decrease with the treatment of the underlying condition.2 In primary erythromelalgia, various treatment modalities have been tried, such as:

- non-steroidal anti-inflammatory drugs (NSAIDs; aspirin, indomethacin);
- beta-blockers;
- antihistamines; physical methods;
- vasodilators (nitroprusside);
- anticonvulsants (gabapentin, carbamazepine);
- antidepressants (amitriptyline);
- antimitotics (carmustine);
- alpha-blockers (prazosine, doxazosine);
- immunosuppressants;
- migraine medications;
- dapsone;
- prostaglandin E1 analogues;
- topical nitroglycerine;
- pentoxyphylline;
- dipyridamole;
- mexiletine HCl;
- quinine;
- muscle relaxants;
- surgical treatments, such as sympathectomy.

The effectiveness of these medications is variable.1,4,7,8 However, improvement in secondary erythromelalgia is usually observed with the treatment of the underlying cause.2

In this patient, prostaglandin E1 analogue, misoprostol, was administered initially. However, because secondary erythromelalgia in SLE has been reported to be a cutaneous vasculitis responding to oral prednisolone,9 the treatment was changed to prednisolone when SLE was considered...
as the primary clinical condition. Although steroids also have been mentioned to be effective in idiopathic erythromelalgia, the rapid response of the patient to steroid treatment favored the diagnosis of SLE associated erythromelalgia.

CONCLUSION

The presentation of this case demonstrates that when erythromelalgia is considered in a pediatric patient, underlying clinical conditions should be carefully sought before it is diagnosed as idiopathic, and proper treatment should be instituted accordingly.

REFERENCES