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PHOTOQUIZ

Palmar telangiectasias in a 4-year-old girl

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CASE PRESENTATION

A 4-year-old girl presented with a 10-month history of asymptomatic red lesions on her palms. Since appearing, the lesions had not changed. Her parents did not recall any specific incident of trauma to her palms. In addition, they denied any preceding illness, swelling, hives, trouble swallowing, epistaxis, ataxia, cold sensitivity, or Raynaud's phenomenon. She had a remote history of neonatal lupus erythematosus (NLE), having been born to an anti-Ro antibody-positive mother. Complete cardiologic evaluation at birth was normal with no signs of heart block. In infancy, cutaneous NLE involving the lateral scalp and neck was treated with triamcinolone cream, resulting in resolution of those lesions. Family history was negative for sclerodactyly.

On examination, there were blanchable red macules on her bilateral hypothenar eminences that appeared vascular under dermoscopy (Figures 1 and 2). There was no involvement of the oral mucosa, no similar lesions were noted elsewhere on her skin, and no other cutaneous findings were noted. Antinuclear antibody (ANA) and extractable nuclear antigen (ENA) profiles were negative.

WHAT IS THE DIAGNOSIS?

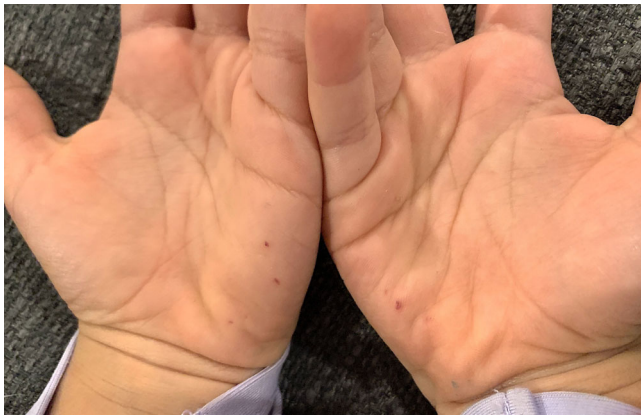

FIGURE 1

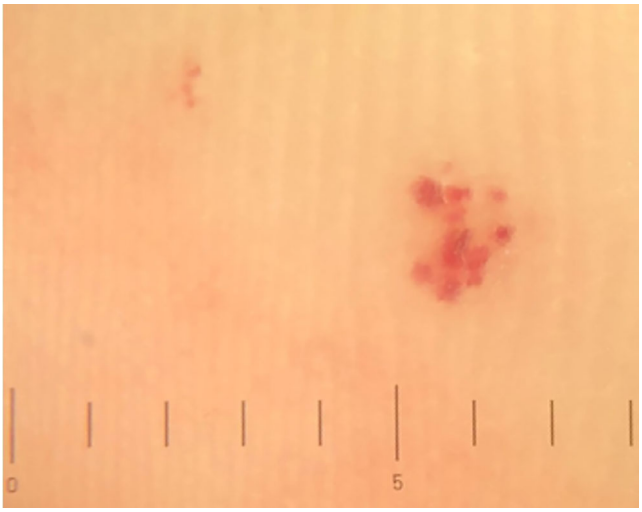

FIGURE 2

Diagnosis: Cutaneous telangiectasias as a sequela of neonatal lupus erythematosus

DISCUSSION

The initial differential diagnosis included viral or trauma-associated telangiectasias, capillary malformation, capillary malformation-arteriovenous malformation (CM-AVM) syndrome, hereditary hemorrhagic telangiectasia (HHT), hereditary benign telangiectasia (HBT), ataxia telangiectasia, angiokeratomas, petechial eruption as a sequela of dermatitis herpetiformis, and CREST syndrome.

CM-AVM syndrome was less likely as our patient lacked the pathognomonic thumbprint-like capillary malformations and halos surrounding pinpoint telangiectasias.[1] It would be unusual for an individual with HHT to develop cutaneous lesions at 4 years old; lesions usually develop later in adolescence with epistaxis starting around age 12.[1] Our patient's few localized telangiectasias were not consistent with HBT, which is characterized by a widespread distribution of telangiectasias on the face, arms, and upper trunk.[1] Ataxia-telangiectasia was also unlikely as ataxia typically precedes cutaneous findings.[2] While solitary angiokeratomas were considered, they usually appear as blue or black papules with marked vascular dilation, although it is possible early angiokeratomas could have a bright red appearance similar to the lesions found in our patient.[3] Reports exist of petechial eruptions on the palms due to dermatitis herpetiformis, with resolution of the petechial lesions after 2 weeks of a gluten-free diet, but our patient had no personal or family history of celiac disease.[4] In addition, our patient did not have any symptoms of Raynaud's disease, nor did she have a personal or family history of sclerodactyly, esophageal dysmotility, or calcinosis cutis to suggest CREST syndrome.

Given her medical history and clinical findings, the patient was ultimately determined to have a cutaneous sequela following infantile NLE. Annual follow-up for a full review of systems, as well as ANA, SS-A, and SS-B testing every 1–2 years, were recommended to screen for collagen-vascular disorders such as CREST syndrome and juvenile idiopathic arthritis. Periodic follow-up is recommended in NLE patients given an increased risk for late-onset collagen-vascular disease.[5] Early intervention may improve patient outcomes.[6]

Cutaneous lesions occur in 50% of patients with NLE typically by 6 weeks of age.[5,7] These lesions tend to appear as erythematous, annular, and scaly plaques in photosensitive locations, though variations may occur. In particular, a subset of patients may present initially with multiple telangiectasias.[5] The rash may also appear in non-sun exposed areas or be present at birth.[7] The majority of these lesions resolve after 6 months as maternal antibodies wane, but a minority may subsequently develop residual dyspigmentation, atrophic scars, or persistent telangiectasias.[5,8] Our patient did not develop telangiectasias until 3 years of age. Previous studies have shown that biopsy of telangiectasias in cutaneous NLE shows superficial vascular dilation without significant inflammatory infiltrate.[5,7] Mechanisms thought to mediate the relationship between persistent late onset telangiectasias and NLE include prior inflammation, secretion of angiogenic factors from affected epidermal or inflammatory cells, immune complex deposition, hormones, and photosensitivity.[5,7] It may take time for these pathogenic mechanisms to accumulate. Cutaneous sequelae following infantile NLE are primarily located in areas of prior involvement but can less commonly be seen on previously unaffected skin. Less than 15% of patients develop telangiectasias; however, these telangiectasias may remain for years.[9] In our patient, it is notable that she had no known prior NLE involvement at the site of the telangiectasias. Only one case of palmar telangiectasias with no prior NLE rash at that site has been described.[7]

It is important to recognize and diagnose cutaneous NLE as affected children are at higher risk of developing hematologic, hepatic, and cardiac disease.[7] Rarely, patients may develop hydrocephalus.[7] Information regarding the long-term prognosis for patients with cutaneous NLE is limited by short interval follow-up as the majority of lesions resolve within 6 months. Cardiac manifestations, most commonly AV block or cardiomyopathy, and rheumatologic/autoimmune diseases are noted to be the most common long-term sequelae of this relatively rare disease. In a study of 49 children with NLE, 12.2% of children with NLE were subsequently diagnosed with a rheumatologic/autoimmune disease, such as juvenile rheumatoid arthritis, more than 8 years after initial diagnosis. This finding highlights the importance of continued follow-up in this patient population.[10] As for the cutaneous manifestations of NLE, patients with telangiectasias should be counseled that the telangiectasias can persist for years but may be effectively treated with pulsed dye laser therapy.[7]

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