Mysterious Vasculitis-Like Rash In A Young Female

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INTRODUCTION

- Vasculitis disorders can present with a variety of cutaneous findings. Livedo reticularis rashes can be physiologic or associated with vessel wall pathology. This can be seen in diseases such as polyarteritis nodosa, autoimmune connective tissue diseases, hypercoaguable states, and cryoglobulinemia. A newly described lymphocytic thrombophilic arteritis phenomenon has been associated with livedo reticularis. It was observed mainly over the lower extremities in a case series of young females.

- We report the case of a 12 year old female with a chronic livedo-reticularis rash located on her lower extremities. Her clinical, laboratory, and histological findings are not conclusive for a specific diagnoses. However, her presentation meets some criteria for lymphocytic thrombophilic arteritis and cutaneous polyarteritis nodosa.

CASE REPORT

- A 12 year old female presented with a one year history of livedo reticularis-like changes on her bilateral lower extremities (Figure 1). The rash initially looked like bruises and was pruritic intermittently. Hydroxyzine gave her some relief from the itching. The initial lesions were noted several months after she started menstruation. No correlation was noted between her menstrual cycles and the cutaneous signs. She denied any pain. The rash was not temperature or elevation dependent. She once noticed aggravation of the rash with hiking on inclined hills/mountains. She complained of occasional joint pains in her shoulders and knees which responded to massages and have resolved. She would get vague abdominal pain that responded to intervention.

- Laboratory analysis included a complete blood count with differential, comprehensive metabolic panel, coagulation studies, thyroid function studies, iron studies, EBV cuttles, ASO titers, ESR, CRP, ANA, RF, p-ANCA, c-ANCA, and cryoglobulins. All studies were negative/unremarkable.

- Punch biopsies were performed on three separate clinic visits. The most recent biopsy was taken from a palpable area of the livedo reticularis on the shin. All three biopsies had non-specific findings. They showed a superficial and mid-dermal perivascular dermatitis without evidence of vasculitis or panniculitis (Figure 2).

- The patient was referred to pediatric hematology and rheumatology for further evaluation. There was no evidence of a hematological disorder. Furthermore, rheumatology concluded there was no definitive support for a systemic autoimmune diagnosis.

DISCUSSION

- The current working diagnosis for this case is lymphocytic thrombophilic arteritis (LTA) versus cutaneous polyarteritis nodosa (CPA).

- LTA presents with slowly progressive patchy hyperpigmentation, and livedo reticularis primarily located on the lower extremities. The cases described in the literature appear to predominantly affect younger women. Four out of five patients had antiphospholipid antibodies. Three out of five patients had elevated erythrocyte sedimentation rates. Histologically, LTA presents with lymphocytes and histiocytes infiltrating the muscular walls of small arteries, located at the dermosubcutaneous junction. Furthermore, a characteristic fibrin ring is present with nuclear dust in the lumen. The condition may respond to prednisone.

- It is currently unknown if there is any significant role for antiphospholipid antibodies in the pathogenesis of LTA. Several factors mitigate a prominent role for this finding. These include: low serum levels in the patients studied, no systemic involvement in the patients presented, no histological evidence of macrovascular thrombosis, and the presence of a dense lymphocytic infiltrate.

- CPA can present with livedo reticularis, palpable purpura, painful nodules, ulceration, and severe digital ischemia. Histologically, a neutrophilic infiltration with fibrinoid necrosis of medium and small-sized arteries is characteristic.

- Our patient seems to clinically and demographically match better with the diagnosis of LTA. However, she lacks the characteristic deeper vessel involvement and intraluminal fibrin ring development histologically. She lacks the nodular and ulcerative lesions more classic for CPA. However, she experienced some temporary bouts of joint pains, abdominal pain, and fatigue which could fit more within the diagnosis of CPA or a mild systemic form of polyarteritis nodosa. Her overall histological interpretation is non-specific. Currently, she is asymptomatic, and has elected to abstain from any pharmacological intervention.

REFERENCES