CASE STUDIES

Cutis marmorata persisting into adulthood

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Abstract

The differential diagnosis of livedoid rashes in an adult includes a broad range of conditions. Persistence of physiologic cutis marmorata into adulthood is noted in certain cases. Fine retiform rash, appearing on cold exposure, is one of the characteristic features. Fixed asymmetric livedo with secondary skin changes or associated systemic symptoms could be a harbinger of underlying anti-phospholipid syndrome or vasculitis.

Keywords: livedoid rashes, livedo reticularis, cutis marmorata

Introduction

The differential diagnosis of livedo reticularis is highly challenging for physicians, dermatologists, rheumatologists and other specialists. The differentials vary widely. The present study discusses one such case encountered by the obstetrician during the course of a normal pregnancy.

Case report

A 24-year-old woman, in her 7th month of first pregnancy, presented to the clinic. She had oligohydramnios and rheumatology consult was sought for a lace-like erythematous rash (livedo) on the chest, abdomen and limbs (Fig.1 & 2). She had had this rash since childhood, with episodic worsening on exposure to cold and disappearance on warming. She had no other features suggestive of any connective tissue disorder. There was no known family history of similar skin rashes. Her hemoglobin was 9.3 gm/dL, total count 13,300/mm^3 with absolute lymphocyte count of 4000 and platelets 1.9/1000 mm^3. Renal and liver function tests were normal and urine examination was unremarkable. Anti-phospholipid syndrome was ruled out with IgM anti-cardiolipin 6.2 mpl/mL, IgG anti -cardiolipin 4.7 gpl/mL, and negative lupus anticoagulant, anti-β2 microglobulin and anti-nuclear antibodies. Cryoglobulin

Fig. 1: Fine retiform erythematous lacy rash on the skin of the anterior abdominal wall in the primiparous patient

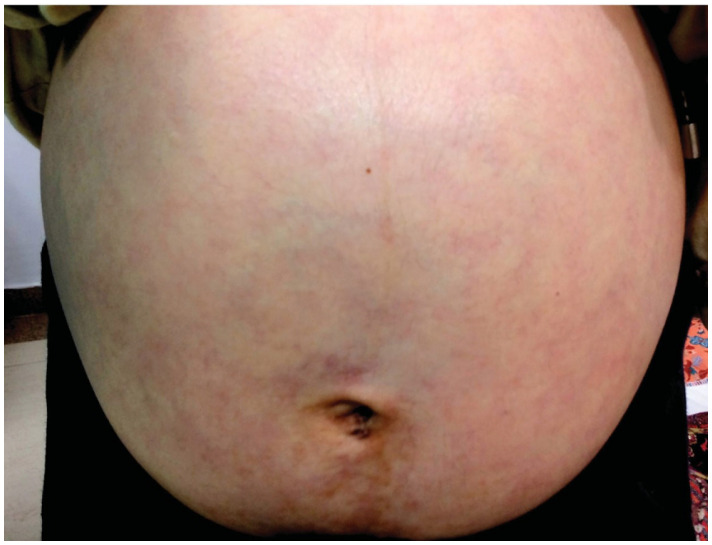
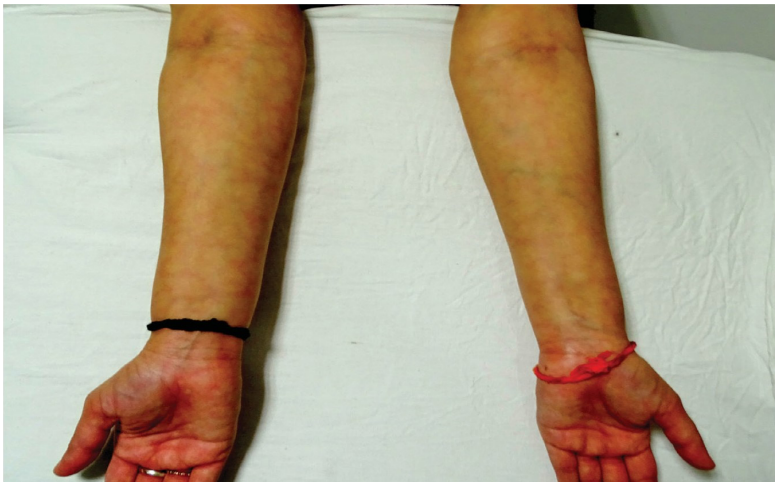


Fig. 2: Prominent livedoid racemose rashes on the unexposed areas due to cold temperature



testing was not carried out, since the duration of symptoms obviated the possibility of a chronic illness. Based on the findings and clinical investigations, she was diagnosed with physiological cutis marmorata.

Discussion

The first description of livedoid rashes was made by Von Lohuizen in 1922.[1] Sneddon, in 1965, described the association of such rashes with recurrent strokes and hypertensive kidney disease.[2] Livedoid rashes have been classically divided into two types, livedo racemosa and livedo reticularis.[3] Livedo racemosa consists of broken lattice pattern forming incomplete circles with branching faint vessels, which vanishes on warming and reappears with cold exposure. Alternatively, livedo reticularis, appears as fixed red to purple branching lines that form complete circles. Clinical differentiation may be difficult. However, more importantly, asymmetry and fixed rash should prompt search for other underlying diseases.

The rash appears due to altered blood flow. The appearance mirrors the cutaneous microvascular anatomy, with perpendicularly oriented arterioles dividing into capillary beds and then draining into the venous plexus. Livedo reticularis can manifest by any process that reduces arteriole blood flow (vasospasm, hyperviscosity, inflammation, thromboemboli) or venous outflow, leading to accumulation of deoxygenated venous blood.[4]

Cutis marmorata is a rare congenital condition with livedoid rash having less than 300 cases reported worldwide.[5] Most cases have underlying morphological abnormalities requiring medical attention at birth. These children present with various internal organ anomalies or limb atrophy in the distribution of the lesions. Some may be cases of neonatal lupus. The condition needs to be differentiated from physiologic cutis marmorata that usually disappears in infancy.

In adults, livedo can be a salient sign of underlying anti-phospholipid syndrome (APS). In a rheumatology clinic, livedo is the most common rash associated with APS, seen in 25%-40% cases, and up to 70% in APS associated with lupus.[6] It results from bland vasculopathy with endothelial dysfunction and vessel occlusion culminating in stellate scars and pigmentary changes, known as ‘atrophie blanche’. In lupus and other vasculitides, the presence of livedo is the cutaneous hallmark of ongoing small vessel vasculitis that needs urgent immunosuppression as these lesions have a tendency to form nodules and bullae and then ulcerate. Rarely, it could be the first presentation of an underlying cold agglutinin disease in the elderly. Amantadine, a drug used for Parkinsonism, can induce livedo as well.[3]

Livedo that resolves with warming is likely to be physiologic. Nevertheless, this needs to be looked at in context, as sudden onset of blanching rash in an elderly male could herald the onset of cold agglutinin disease. The role of skin biopsy is limited to differentiation of vasculitis from bland vasculopathy. Physiologic livedo reticularis is a clinical diagnosis. Physiologic marmorata may persist into adulthood in rare cases, causing anxiety to the unaware physician and patient alike.

Conclusion

The key feature to be noted for physiologic marmorata is the disappearance of the rash on warming and reappearance on cold exposure. Reassuring the patient would suffice. The symptoms may improve spontaneously with age. In the present case, a healthy male baby was delivered at term.

Competing interests

The author declares that she has no competing interests.

Citation

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