Cutis marmorata telangiectasia congenita with painful ulcerations

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Abstract

Cutis marmorata telangiectatica congenita (CMTC) is a rare, congenital, vascular disorder that may sometimes be associated with ulcerations of the involved skin. We present a case of CMTC, asymptomatic since birth, that began developing painful ulcerations during adolescence. Although laser therapy may benefit the superficial aspect of this vascular anomaly, the presence of deeper involvement in lesions with ulcerations may not respond favorably to laser therapy and the best approach needs to be further evaluated.

Keywords: vascular anomaly, cutis marmorata, ulceration, laser therapy

Introduction

Cutis marmorata telangiectatica congenita (CMTC) is an infrequent, congenital, sporadic vascular anomaly. It is characterized by erythematous-to-violaceuos patches in a reticulated pattern and may be associated with ulcerations [1]. We describe CMTC on the back of an adolescent male who began to develop ulcerations and associated back pain.

Case Synopsis

A 15-year-old boy presented for evaluation of a vascular lesion of the left lower back. The lesion has been present since birth and was diagnosed as CMTC. The condition had been asymptomatic for most of the patient's life and had grown in proportion to the patient's growth. Two years prior to presentation, the patient began developing painful ulcerations in the medial aspect of the lesion. Around the same time, he started having back pain and there was concern for erector spinae muscle weakness and hypoplasia. The pain was described as occasional discomfort and as quick and sharp in nature; however, it did not affect his physical activity. The lesion tends to look red at times while blue at others, but no known correlation of color change has been observed with activity or temperature changes. Family history is pertinent for a maternal uncle with vascular anomaly associated with increased size of the right hand. There was no relevant medical history in our patient.

Physical examination of the lower right back revealed a reticular, deep purple patch with prominent vasculature, measuring 19cm in width × 13cm in length. The medial aspect of the lesion revealed ulcerations with overlying crusting and scaling (**Figure 1**).

The patient was referred for evaluation of pulsed dye laser (PDL) therapy, which should be of benefit for the superficial portion. An MRI was considered prior to any treatment to assess for the degree of hypoplasia associated with the CMTC given the onset of back pain with concern for muscle hypoplasia. In addition, we were interested in assessing for any concurrent underlying vascular malformation given the presence of continued ulcerations in our patient. Unfortunately, our patient declined further evaluation.

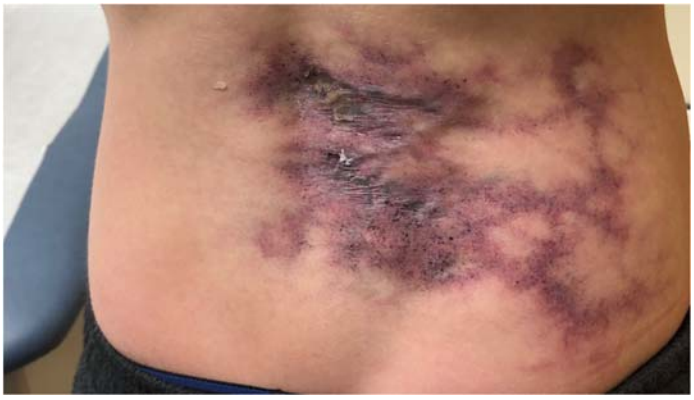

Figure 1. *Cutis marmorata telangiectasia congenita involving the lower back, including reticular, deep purple patch with prominent vasculature, measuring 19cm in width × 13cm in length. Medial aspect of the lesion reveals ulcerations with overlying crusting and scaling.*

Case Discussion

Cutis marmorata telangiectatica congenita is a rare, benign congenital cutaneous vascular anomaly characterized by persistent reticular vascular skin pattern with a marbled bluish to deep purple appearance. It is often present at birth but can appear later in infancy. Involvement can be generalized or most commonly localized to limbs, followed by the trunk and face [2, 3]. Localized erythema is often sharply demarcated and unilateral without crossing the midline [4]. Ulceration and atrophy of the involved skin can be seen in the neonatal period and may continue during infancy or childhood [5]. The pathogenesis of CMTC is unclear and is likely a multifactorial sporadic condition [4]. A wide variety of systemic anomalies are reported in patients with CMTC with limb asymmetry being the most consistent [4]. Glaucoma has been reported in cases of facial CMTC and performing tonometry is advisable [6].

The diagnosis of CMTC is clinical and histologic findings are nonspecific [5]. These usually reveal dilated capillaries and veins within the dermis [6]. Cutis marmorata telangiectatica congenita may resemble physiological cutis marmorata, which is normal skin mottling in response to cold temperatures in the first few weeks of life that disappears on warming [3]. Diagnostic criteria for CMTC have been suggested but the validity has not been established [4]. These include the presence of all three major criteria (congenital presence of reticulate erythema, absence of venectasia, and lack of response to local warming) and two of the five minor criteria (fading erythema within two years, telangiectasia, port-wine stain outside of CMTC affected area, ulceration, and atrophy), [6].

Skin lesions and dermal changes generally improve during the first two years of life and treatment is not required. Laser therapy in patients with persistent CMTC has demonstrated variable outcomes with generally poorer response compared to capillary malformations treated with pulsed dye laser and increased risk of scarring [5].

Conclusion

Although laser therapy may benefit the superficial aspect of this vascular anomaly, the presence of deeper involvement in lesions with ulcerations may not respond favorably to laser therapy and the best approach needs to be further evaluated.

Potential conflicts of interest

The authors declare no conflicts of interests.

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