A 13-year-old girl with a linear dark patch on her forehead: A case of scleroderma en coup de sabre in a child with skin of color presenting with a bruise-like appearance

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INTRODUCTION

Localized scleroderma represents a group of connective tissue disorders that primarily affect the skin and subcutaneous tissue. Linear scleroderma is the most common type of localized scleroderma in children; it is thought that 67% of cases of linear scleroderma are diagnosed before the age of 18. According to some reports, scleroderma (both localized and systemic) represents the third most common rheumatologic condition in children after juvenile idiopathic arthritis and systemic lupus erythematosus, although localized scleroderma is more likely to affect the pediatric population than systemic scleroderma. Linear scleroderma of the forehead is referred to as scleroderma en coup de sabre. Here we describe an early case of scleroderma en coup de sabre that appeared as a bruise-like patch in our patient with skin of color, a relatively rare presentation. We also discuss a relatively recent classification scheme for localized scleroderma and potential treatment guidelines for pediatric patients.

CASE REPORT

A 13-year-old Hispanic girl presented to the pediatric dermatology clinic for the evaluation of discoloration on the left side of her forehead that appeared approximately 5 months earlier. It had not changed in size, shape, or color since appearing. The patient’s mother reported that the discoloration started suddenly, and the patient denied trauma to the area. The patient denied pruritus, pain, or bleeding in the area of the discoloration. She denied headaches, loss of sensation in the area, or weakness in any part of her body. The patient’s primary care doctor had prescribed triamcinolone 0.1% cream but with no improvement. The primary care provider, believing that this may be a vein, also tried to drain the area with no improvement seen. The patient had no significant medical history, denied a history of recent illness, and was not currently using any other prescription medication.

On the left side of the forehead of this patient there was an approximately 7- × 1-cm linear, hyperpigmented patch extending from the medial end of the left eyebrow to the hairline (Fig 1). No atrophy was associated with the lesion, nor was there any pain on palpation. A punch biopsy was performed on the superior aspect of the patch.

Histopathology found moderate fibrosis of the deep reticular dermis with focal, mild lymphohistiocytic infiltrate (Fig 2). In the superficial dermis, there were dermal melanophages with scattered necrotic keratinocytes. A diagnosis of linear scleroderma/morphea was given based on these findings.

DISCUSSION

Our patient’s scleroderma presented as en coup de sabre, the term given for linear scleroderma of the forehead. Our patient’s presentation was unique in that she did not have any induration or atrophy on the forehead. She did not have any induration or atrophy on the area.
physical examination, but rather, appeared as a bruise. This bruise-like presentation is a relatively uncommon presentation of scleroderma en coup de sabre in a pediatric patient with skin of color that we believe has only been documented 1 other time in the literature. Weibel et al estimated 4% of linear scleroderma cases in pediatric patients were initially misdiagnosed as a bruise. The flattened appearance of the lesion usually correlates with an early stage of linear scleroderma. Because of the lack of atrophy or induration, discoid lupus erythematosus (DLE) was also considered in the differential diagnosis. DLE can present as patches and be localized to the scalp and face in up to 80% of the patients; however, DLE is found to be responsive to topical calcineurin inhibitors. Our patient was put on 0.1% tacrolimus cream by her primary care doctor and had not shown a response to the treatment.

Although there has been extensive work done on classifying systemic scleroderma and developing its criteria, reciprocal measures for localized scleroderma have yet to be taken. A consensus conference in Padua, Italy in 2004 attempted a new classification scheme for localized scleroderma (Table I). Although scleroderma en coup de sabre is a form of localized scleroderma, it is not uncommon to find systemic manifestations in patients with this condition, and it is estimated that roughly one quarter of patients with localized scleroderma will have extracutaneous manifestations. A study done in 2007 found that the scleroderma en coup de sabre subtype in children had significantly more ocular abnormalities than any other scleroderma subtype. The most common ocular manifestations were adnexal abnormalities involving the eyelashes, eyelids, and lacrimal glands.

Adding to that, there is a growing body of literature that suggests that children with scleroderma en coup de sabre have neuroimaging performed, even when the patient is asymptomatic. Several studies have found that neurologic symptoms, or lack thereof, are not an accurate predictor of abnormal neuroimaging studies. The most common findings on imaging tend to be T2 hyperintensities, calcifications, and cerebral atrophy. The most common neurologic symptoms reported in children with scleroderma en coup de sabre are headaches and seizures. Our patient denied any neurologic symptoms including headaches, loss of sensation, or weakness. Despite a negative neurologic review of systems, our patient had magnetic resonance imaging performed. Her magnetic resonance image showed evidence of en plaque dermal thickening of the right paramedian frontal extracranial soft tissues.

In 2012, a work group of 12 pediatric rheumatologists along with 2 dermatology consultants, all of whom had a special interest in localized scleroderma, came together to begin the development of a standardized treatment of localized scleroderma. Three treatment regimens were proposed: (1) methotrexate alone (subcutaneous administration was preferred), (2) methotrexate plus intravenous (IV) corticosteroids, and (3) methotrexate plus oral corticosteroids. Each of these treatment regimens was designed to last 12 months. The recommended dose for methotrexate is 1.0 mg/kg/wk, with a maximum dose of 25 mg/kg. For IV corticosteroids, the recommended dosing was 30 mg/kg/IV dose with a maximum dose of 1,000 mg. For oral
corticosteroids, the recommended dosing was 2 mg/kg/day orally with a maximum dose of 60 mg. Dosing recommendations for mycophenolate mofetil were also given should a patient be unresponsive or intolerant of one of the methotrexate-based approaches. Our patient is currently on a 2-year course of oral methotrexate and receiving weekly, pulsed solu-medrol infusions.

### Table I. Classification of localized scleroderma subtypes

| Group                  | Subtype          | Description                                                                                                                                 |
|------------------------|------------------|--------------------------------------------------------------------------------------------------------------------------------------------|
| Circumscribed morphea  | Superficial      | Oval or round circumscribed areas of induration limited to epidermis and dermis often with altered pigmentation and an erythematous, violaceous halo (lilac ring). They can be single or multiple. |
|                        | Deep             | Oval or round circumscribed induration involving the subcutaneous tissue that extends to the fascia and may involve underlying muscle. The primary site may involve the subcutaneous tissue without and involvement of the overlying skin. They can be single or multiple. |
| Linear scleroderma     | Trunk/limbs      | Linear induration involving the skin, subcutaneous tissue, and, sometimes, the underlying muscle and bone. |
|                        | Head             | En coup de sabre: Linear induration sometimes affecting the underlying muscle and bone. On the face and scalp. Perry-Romberg (progressive hemifacial atrophy): Loss of tissue on one side of the face that may affect muscle and bone. The overlying skin is mobile. |
| Generalized morphea    |                  | Induration of the skin starting as individual plaques (4 or more and larger than 3 cm) that become confluent and involve at least 2 of the 7 anatomic sites: Head-neck, Right upper extremity, Left upper extremity, Right lower extremity, Left lower extremity, Anterior trunk, Posterior trunk |
| Pansclerotic morphea   |                  | Circumferential involvement of the limb(s) affecting the skin, subcutaneous tissue, muscle and bone. There are typically severe limitations on the range of motion in affected joints. The lesion may involve other anatomic sites without any internal organ involvement. |
| Mixed morphea          |                  | Combination of 2 or more of the previous subtypes. The order of the concomitant subtype, specified in brackets, will follow the predominant representation in the individual patient (ie, mixed morphea, [linear-circumscribed]). |

Adapted from Laxer et al, 2006. Original source: Consensus Conference, Padua, Italy, 2004.

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