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Dermatologic ultrasound in the management of childhood linear morphea

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Abstract

Linear morphea is the most common subtype of localized scleroderma in the pediatric population. This condition can be quite disabling, with complications such as growth defects and painful flexion contractures. Assessment of disease progression and early intervention are key to minimize morbidity. We report linear morphea in a previously healthy 12-year-old girl. The patient presented with a one-year history of a linear plaque crossing her left antecubital fossa, measuring 7×3cm. The diagnosis was confirmed by biopsy, in which deep tissue involvement was noted. Subsequent management and evaluation of the disease activity was done by ultrasound, which allowed precise guidance of pharmacotherapy. The patient improved both clinically and sonographically with a methotrexate course. Sonographic changes accurately described the disease activity on follow up assessments. Features suggestive of an active phase include a thickened and hypoechoic dermis contrasting hyperechoic subcutaneous tissue. The atrophic stage is characterized by a thinned-out dermis and subcutaneous area. Typical vascular traits of each disease phase can also contribute to the assessment. Ultrasound is a grossly underused tool in the field of dermatology. It can provide accurate and sensitive information about disease activity in linear morphea, allowing for more timely intervention and optimal patient management.

Keywords: dermatology, linear morphea, localized scleroderma, pediatrics, ultrasound

Introduction

Linear morphea, the most common subtype of localized scleroderma in the pediatric population, mainly affects the skin but can extend to the subcutaneous tissues with occasional involvement of muscle and bone [1]. This condition can be quite disabling, with complications such as growth defects and painful flexion contractures when lesions extend beyond joint lines [2]. Assessment of disease progression and early intervention are key to minimize morbidity. We present a case of childhood linear morphea for which management was guided by high frequency ultrasound (HFUS) assessments.

Case Synopsis

A previously healthy 12-year-old girl first presented to the department of orthopedic surgery for functionally impairing pain of the left wrist. Magnetic resonance imaging (MRI) was performed and findings were consistent with a possible post-traumatic synovitis. Laboratory investigations were unremarkable with the exception of positive anti-nuclear antibodies. A diagnosis of juvenile arthritis was made at this time, for which an oral corticosteroid taper and a six-month course of methotrexate was initiated.

One month after discontinuing the methotrexate treatment, the patient noted the appearance of subtle cutaneous changes on her left arm. This progressed and one year after its onset, the patient was unable to fully extend her arm, holding it in a 50-

degree flexed position. The patient was then referred to our dermatology department. On examination, an indurated linear plaque of slightly atrophic hypopigmented skin was noted. The plaque measured 7×3cm and extended from the distal bicep to the proximal forearm, crossing the joint line over the radial aspect of the left antecubital fossa (**Figure 1A**). There was reduction in the range of motion at the left elbow, as well as mild limitation of the left wrist extensors, associated with tenosynovitis of the left wrist.

Ultrasound of the cutaneous plaque, performed with two linear variable frequency 6-18MHz and 10-22MHz probes on Esaote class C equipment (Geneva, Italy), exhibited dermal thickening and hyperechoic subcutaneous tissue when compared to the normal skin of the contralateral limb (**Figure 1B**). This picture was highly suggestive of active morphea. A biopsy was taken at the time of this assessment, demonstrating lymphoplasmocytic septal panniculitis compatible with deep morphea, thus confirming the diagnosis. At this point, the patient was restarted on methotrexate at the same dose of the prior treatment, with an anticipated extended taper over two years.

At three-month follow up, the plaque had substantially improved clinically. On ultrasound, there was some recovery in dermal echogenicity, whereas the subcutaneous tissue remained hyperechoic. The dermal-subdermal interface was gaining definition but was not completely recovered, a picture compatible with inactive morphea (**Figure 2**).

We saw the patient again one year into this treatment, at which point she had recovered full range of motion at her left elbow joint. Clinically, the linear morphea appeared to show post inflammatory hyperpigmentation (**Figure 3A**). Sonographically, it displayed a complete recovery of the dermal-subdermal interface (**Figure 3B**).

Case Discussion

Several non-invasive tools for characterization of disease activity and severity in scleroderma have

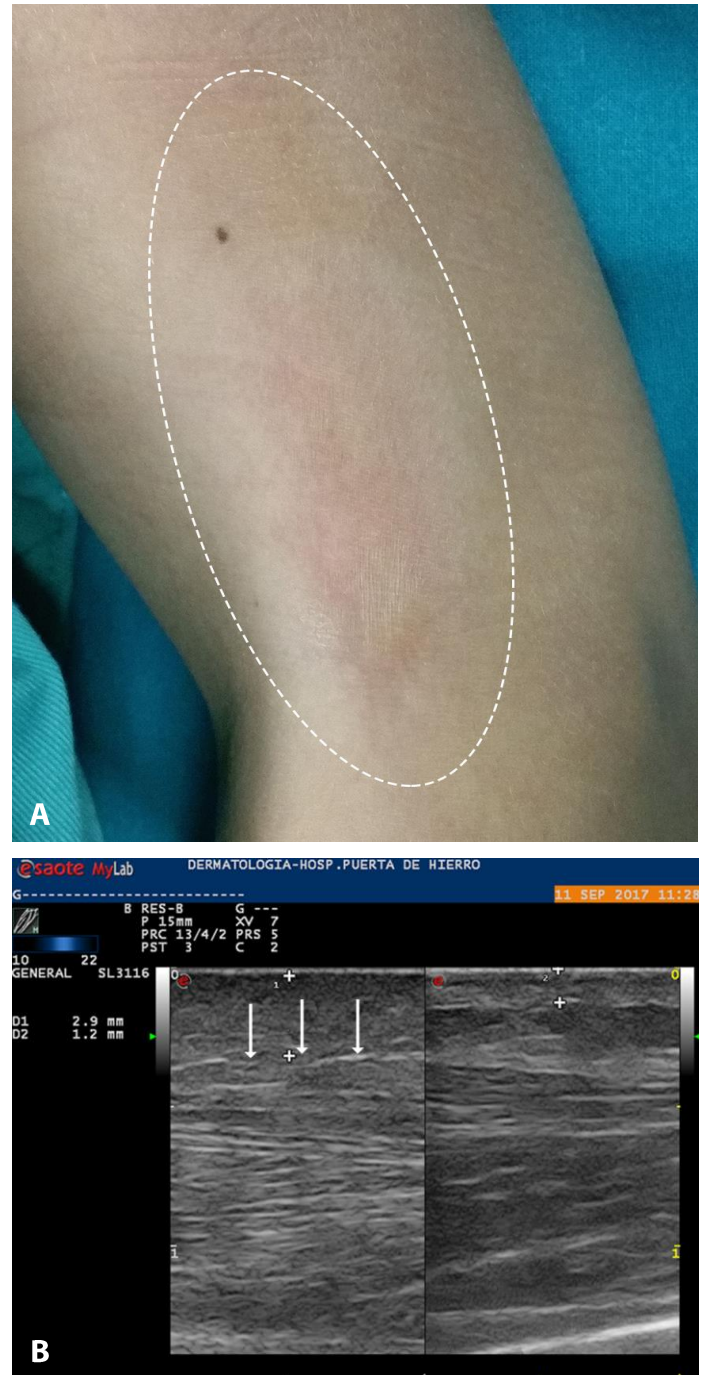


Figure 1. Assessment at presentation. **A)** Clinical picture of the linear morphea at presentation. An indurated linear plaque of slightly atrophic hypopigmented tissue was noted (dotted circle). The lesion measured 7×3cm and extended from the distal bicep to the proximal forearm, crossing the joint line over the radial aspect of the left antecubital fossa. **B)** Sonographic assessment at presentation. Left of picture, morphea plaque (left arm) showing hypoechoic dermal thickening of 2.9mm with hyperechoic subcutaneous tissue (between the two cross markers). Dermal-subdermal interface is obliterated (arrows), highly suggestive of active morphea. Right of picture, contralateral arm (right arm) normal skin for control, dermal thickness of 1.2mm

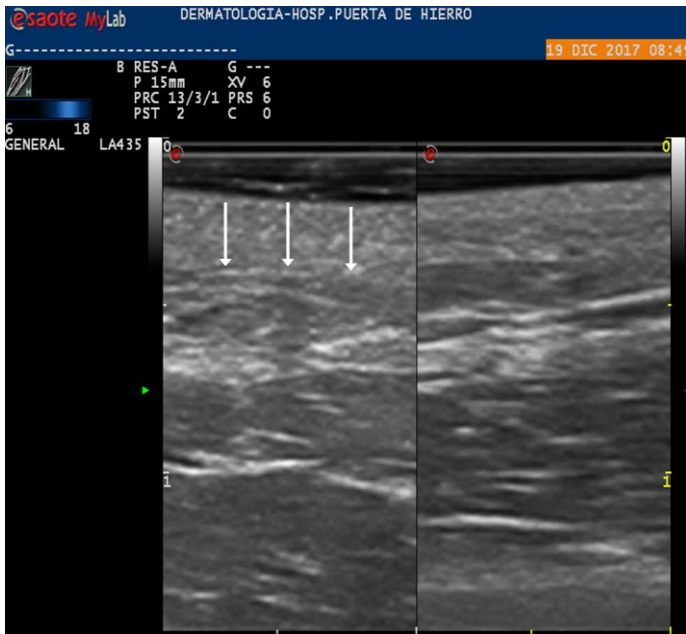


Figure 2. Sonographic assessment at 3-month follow-up. Left picture, Morphea (left arm) showing some recovery in dermal echogenicity whereas subcutaneous tissue remains hyperechoic. Dermal-subdermal interface is improving but not yet clearly defined (arrows), highly suggestive of inactive morphea. Right of picture, contralateral arm (right arm) normal skin for control.

been developed, including infrared thermography [3], laser doppler evaluation [4] and MRI [5]. Unfortunately, these techniques have proven to lack resolution and to be time consuming, resource intensive, as well as prohibitively expensive. Further, clinical tools including the modified Rodnan Skin Score (mRSS) evaluate thickness by palpation as a surrogate for disease evolution and severity in scleroderma [6,7]. Various studies have compared the mRSS to U.S. findings and some showed a high correlation between the two [8,9], whereas others found a poor correlation [10,11]. Poor agreement is speculated to be at least in part related to changes in skin texture being perceived clinically as changes in thickness, whereas HFUS can assess thickness specifically. However, it appears consistent that HFUS improves the previously documented poor intra and interobserver variability of mRSS and reflects more subtle changes in thickness [10-14].

A study conducted on all varieties of primary cutaneous morphea lesions in both adult and pediatric patients validated sonographic criteria for assessment of morphea activity. Active, inactive, and

atrophic lesions were distinguished on the basis of dermal thickness, dermal echogenicity, subcutaneous echogenicity, and blood flow to dermal or subcutaneous tissue. Ultrasound was found to have a sensitivity of 100% and specificity of 98.5% in distinguishing between disease activity phases when compared to histological analysis [15]. The criteria for active morphea lesions include increased cutaneous blood flow to the dermis or subcutaneous tissue or two or more of the following criteria: increased dermal thickness, decreased dermal echogenicity (focal or diffuse), and increased subcutaneous tissue echogenicity. Although blood

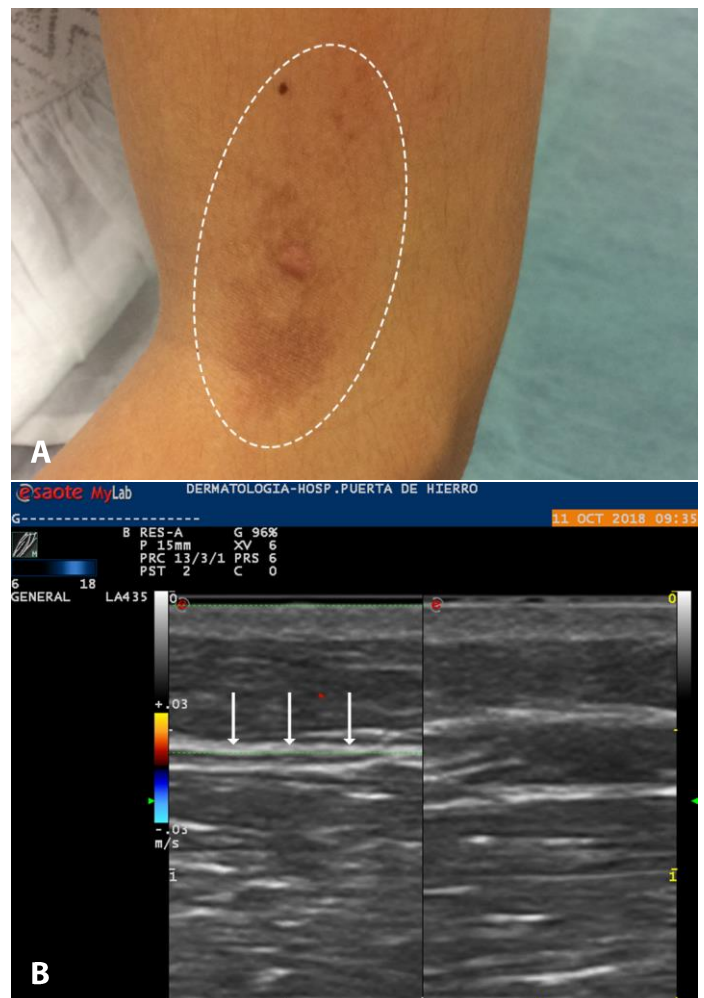


Figure 3. Assessment at 1-year follow up. **A)** Clinical picture of the lesion at 1-year follow-up (dotted circle). Lesion is now hyperpigmented related to post-inflammatory changes, with a small circular well circumscribed atrophic plaque. **B)** Left of picture, morphea (left arm) showing thinning of the now hyperechoic dermis. Full recovery of the dermal-subdermal interface (arrows), highly suggestive of atrophic morphea. Right of picture, contralateral arm (right arm) normal skin for control.

flow was not assessed in our patient, the evaluation met all three additional criteria of an active morphea plaque on her first assessment. Our patient improved both clinically and sonographically on methotrexate treatment. Sonographic changes accurately described the disease activity on follow up assessments. No optimal technical parameters have been thus far described for the specific evaluation of morphea skin changes by U.S., other than general guidelines established by the DERMUS (Dermatologic Ultrasound) group for standardized dermatologic ultrasound assessments [16], which were diligently followed by our team.

There has been evidence of important delays and misdiagnosis of children with localized linear morphea [17]. Knowing that morphea may cause significant morbidity, such a validated technique for diagnosis and monitoring is of great value and may improve the time to diagnosis and patient management. Furthermore, this technique is safe, as

it does not involve ionizing radiation or require anesthesia. Lastly, resolution, cost effectiveness, and availability are considerable advantages of sonography as alternative assessment techniques.

Conclusion

Ultrasound is a grossly underused tool in the field of dermatology. It can provide accurate and sensitive information about disease activity in linear morphea, allowing for more timely intervention and optimal patient management, as demonstrated in the case presented. Although the diagnosis of linear morphea remains clinical, the use of cutaneous sonography has been validated for monitoring of progression/resolution [15].

Potential conflicts of interest

The authors declare no conflicts of interests.

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