

Linear scleroderma and foot deformity in children: Case report

Zahi Siham*; Mahir Laila; Belhaj Karima; Lmidmani Fatima; El Fatimi Abdellatif

*Zahi Siham

Physic medicine and rehabilitation department -CHU IBN ROCHD-CASABLANCA, MOROCCO.

Email: zasi1986@hotmail.com

Abstract

Localized scleroderma or morphea is confined to the skin and/or underlying tissues. It is a rare disease of unknown etiology. Linear scleroderma is a subtype of localized scleroderma generally observed in children, and may produce secondary bone and joint deformities. Its localization at the foot or ankle is rarely reported. A complete review of the literature reveals 22 cases of foot or ankle deformity due to linear scleroderma to which we added one case. Treatment should be started very early, before complications occur due to the high morbidity rate of individuals with localized scleroderma.

Keywords

linear scleroderma; localized scleroderma; morphea; foot deformity

Introduction

Localized scleroderma or morphea is a chronic connective tissue disease of unknown etiology [1]. The localized forms of scleroderma are more common in children [2,3]. Localized forms are not associated with systemic manifestations but may cause significant atrophy of the underlying bones and muscles, thereby limiting joint mobility and causing growth deformations [4,5]. Linear scleroderma is the most common subtype of localized scleroderma [6]. It is a rare disease with an incidence of around 0.3 to 3 cases per 100,000 inhabitants/year [7].

Linear scleroderma is characterized by circumscribed fibrotic bands confined to a limited segment of skin and subcutaneous tissue on the extremities or face [2,8]. It can extend to the dermis, subcutaneous tissue, muscle, and bone.

We report a case of a child suffering from a localized scleroderma with foot and ankle deformity and discuss and compare therapeutic modalities with literature.

Observation

An 8 years old boy consulted for oval-shaped plaques of skin in the right thigh and foot. The clinical diagnosis of linear scleroderma was made and a simple monitoring was proposed without active treatment. Two years later, he came to us for an evolutionary and painful deformation of the right foot, interfering with footing.

The physical examination found:

- Skin lesions at the right thigh and ankle (figure 1, 2)

- A short amyotrophic right lower limb (figure 3)
- Monoparesia grade 4/5
- Stiffness of the right ankle: passive dorsiflexion of the ankle limited to 10 degrees and plantar flexion to 15 degrees
- The podoscopic examination found hollow feet grade 1 with calcaneal valgus more accentuated at right (figure 4)
- Walking is done with lameness and attack by the forefoot.

Radiographies of the foot and ankle in charge confirmed the clinical valgus (figure 5). Treatment consisted of corticosteroid administered orally (1mg/kg/j = 20mg/j). A physical therapy was established in association with prescription of orthopedic soles for hollow feet grade 1 and intern corner; In addition to a correction of the inequality of lower limbs of 3 cm at left.

With an 8-month follow-up, walking was done without lameness or pain. Ankle mobility was 30 degrees in dorsal flexion and 40 degrees in plantar flexion.

Discussion

There are two categories of scleroderma: systemic sclerosis characterized by cutaneous sclerosis and visceral involvement; and localized scleroderma, which classically presents benign and self-limiting and is confined to the skin and/or underlying tissues. Localized scleroderma is a chronic connective tissue disease of unknown etiology [1,9].

Linear scleroderma is characterized by one or more linear streaks of cutaneous induration that may involve dermis, subcutaneous tissue, muscle and underlying bone. Linear scleroderma is often observed in children and adolescents, and is the most frequent form of scleroderma in childhood, affecting 40-70% of the children studied [10-13]. Approximately 67% of patients with linear scleroderma are diagnosed before age 18 years [7]. It is usually a single, unilateral lesion of linear distribution and involves the extremities, face or scalp.

Linear scleroderma may affect the muscles and underlying bones, causing growth disturbance and ankylosis [14-16]. Children are more frequently affected than adults, but both sexes are affected equally.

The variable clinical evolution of localized scleroderma makes it difficult to manage. The physiatrist is often consulted because this lesion tendency to develop retraction, pain and stiffness [17].

Deformations of the foot and / or ankle, secondary to focal scleroderma, are rare in children. We have found 12 articles dealing with orthopedic deformities of Focal scleroderma, 9 of them relate to ankle or foot deformations (Table I).

Due to the rarity of this disease, localized scleroderma is not immediately recognized and delay diagnosis. Ultrasound, thermography, scintigraphy and several serological tests are sometimes used, with limited success [18]. The most frequent manifestations are: inequality of length of lower limbs, muscular atrophy and osteoarticular deformations [18-21]. All of these manifestations were present in our patient. Bone abnormalities, when exist, may occur radiologically in form of cortical thickening,

Subcutaneous calcifications [17], or simulate a mellorheostosis [18]. In our case articular deformity is caused by tissue retraction, without compromising bone structure radiologically proven.

Morphea may present tendency to progress and recur use, especially when the onset of disease occurs in childhood [22,23]; causing joint deformities [24,25], as described in our observation.

Nevertheless, localized scleroderma classically presents a self-limited course with a tendency to spontaneous regression after 3 to 5 years [26]. Plaque morphearearly progresses to generalized or debilitating forms.

The management of localized scleroderma is still unsatisfactory and there are very few randomized and controlled therapeutic studies [27]. Different therapeutic modalities have been suggested, including the use of topical medications, immunosuppressive pharmacological agents, physical therapy and phototherapy [28-30].

Treatment should be initiated at an earlier stage before complications occur due to the high morbidity of localized scleroderma, which leads to limitation of motion and deformities. In this case, treatment was delayed causing stiffness and deformities. Apparatus or physiotherapy is indicated if osteo-articular deformities are associated. Orthotics and plasters can slow down the Progression of joint deformities [17,31,32].

In this case, rehabilitation associated with correction of the inequality of lower limbs length and static disorders, gave a good functional result mainly on shoeing and walking.

Surgical procedure may be indicated to correct orthopedic deformation and restore or maintain joint mobility, through extensive surgical release of retracted soft tissues [18,23,33-35].

Table

Authors	Osteoarticular affection	Ankle or foot's affection	Deformation
Buckley et Skinner [17]	7	6	Calcaneo-valgus or varus-equine
Bernstein <i>et al.</i> [33]	4	5*	4 Metatarsus adductus: 1 varus' foot
Botoni et Reinker [18]	14	2	1 equine: 1 varus' foot
Goel et Shanks [32]	1	0	
Liu <i>et al.</i> [19]	18	1	Ankle : stiffness
Glockenberg <i>et al.</i> [20]	1	1	Plantar skin lesion
Chan et Shyur [24]	1	1	Stiffness first and second toes
Hatzis [14]	1	0	
Maurer <i>et al.</i> [25]	2	2	1 Equine: 1 Valgus
Katarincic <i>et al.</i> [35]	4	3	1 Equine: 2 valgus-equine
El Masri <i>et al.</i> [34]	1	1	Calcaneo-valgus

Table 1: Data from literature. (*Medial lesion of the foot)

Figures



Figure 1: linear scleroderma in the right thigh.



Figure 2: linear scleroderma in the right ankle and foot.



Figure 3: amyotrophic right lower limb.



Figure 4: an optic podoscopic examination: hollow feet grade 1 + bilateral calcaneal valgus.



Figure 5: Radiography of foot and ankle.

Conclusion

Foot localized scleroderma of a child is a rare disease. It may be associated with irreducible and evolutionary orthopedic deformations. Early diagnosis and suitable management can prevent these complications. Rehabilitation and prolonged apparatus are a part of the therapeutic arsenal.

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Authors Information: Zahi Siham*; Mahir Laila; Belhaj Karima; Lmidmani Fatima; El Fatimi Abdellatif

Physic medicine and rehabilitation department -CHU IBN ROCHD-CASABLANCA, MOROCCO.

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