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Original Investigation

Juvenile localized scleroderma: Is it a benign disease?☆

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ABSTRACT

Introduction: Juvenile localized scleroderma is a polymorphic disease. It is more prevalent in girls and has a significant morbidity. Extra-cutaneous involvement is common, and polyautoimmunity can reach 7%. The clinical characteristics of this disease in Colombian patients are currently unknown.
Objective: To describe the clinical characteristics, morbidity and outcomes in patients with juvenile localized scleroderma in different pediatric rheumatology clinics in Colombia.
Materials and methods: A descriptive, retrospective, and multicentre study was conducted on patients with juvenile localized scleroderma with a minimum of 1 year of disease onset, and 6 months of follow-up in 10 pediatric rheumatology clinics.
Results: The study included 88 patients, with a gender distribution of female 2.1: male 1. Mean age at disease onset was 7.1 years (0–14). Mean disease duration at diagnosis was 16.5 months (1–96). Sub-type distribution was, circumscribed (32.9%), mixed (31.8%), and linear (21.5%), that increased to 55% if linear lesions of the mixed subtype are included), generalized (11.4%), and pan-sclerotic morphea (2.3%). Esthetic compromise was detected in 91%, with growth disturbances in 41%, and joint functional compromise in 32%. Extra-cutaneous involvement occurred in 22.7% and polyautoimmunity in 12.5%.
Conclusions: Juvenile localized scleroderma is a polymorphic and unpredictable disease. It diagnosed late in most of the cases. Extra-cutaneous involvement suggests that is not a disease limited to skin. An early diagnosis, a dynamic treatment and a close follow-up helps to prevent, and detect, complications arising from the disease.

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Esclerodermia localizada juvenil: ¿es una enfermedad benigna?

Palabras clave:
Esclerodermia localizada
Morfea
Esclerodermia circunscrita

RESUMEN

Introducción: La esclerodermia localizada juvenil es una enfermedad polimórfica que ocurre con mayor frecuencia en niñas. Se acompaña de morbilidad importante. El compromiso extradérmico es frecuente y se reportan tasas de poliautoinmunidad de hasta 7%. Al momento, se desconocen las características clínicas de los pacientes colombianos con esta enfermedad.

Objetivo: Describir las características clínicas, morbilidades y secuelas en pacientes con diagnóstico de esclerodermia localizada juvenil, en múltiples centros de reumatología pediátrica en Colombia.

Materiales y métodos: Estudio descriptivo, retrospectivo y multicéntrico. Pacientes con diagnóstico de esclerodermia localizada juvenil con un mínimo de 1 año de evolución y 6 meses de seguimiento, en 10 centros de reumatología pediátrica mediante revisión de historias clínicas.

Resultados: El n = 88. La distribución por género fue: femenino 2,1; masculino 1. Edad promedio al inicio de la enfermedad 7,1 años (0-14). Promedio de duración de la enfermedad al diagnóstico 16,5 meses (1-96). La distribución por subtipos fue morfea circunscrita (32,9%), mixta (31,8%), linear (21,5%, asciende a 55% al incluir formas mixtas con lesiones lineares) generalizada (11,4%) y panesclerótica (2,3%). Se detectaron alteraciones estéticas en el 91%, alteraciones del crecimiento en 41% y compromiso funcional de articulaciones vecinas en 32%. Se presentó compromiso extradérmico en 22,7% y poliautoinmunidad en 12,5%.

Conclusiones: La esclerodermia localizada juvenil es una enfermedad polimórfica e impredecible. En la mayoría de los casos el diagnóstico es tardío. La tasa de compromiso extradérmico sugiere que no es una enfermedad limitada a la piel. Un diagnóstico temprano, tratamiento dinámico y seguimiento cercano permiten prevenir y detectar tempranamente complicaciones derivadas de la enfermedad.

Introduction

Scleroderma is an autoimmune, polymorphic disease, characterized by the presence of cutaneous sclerosis secondary to the excessive accumulation of collagen.[1,2] It is classified into 2 broad groups, localized scleroderma and systemic scleroderma. It is called localized scleroderma when the skin involvement is not accompanied by affectation of internal organs. In some cases it can involve neighboring structures or even originate distant symptoms, but unlike the systemic form, usually does not compromise vital organs and generates a different morbidity. Systemic scleroderma is characterized by involvement of internal organs and a worse prognosis.[1]

The classification criteria published by the Pediatric Rheumatology European Society (PRes) categorize the juvenile localized scleroderma (jLS) into 5 types according to the skin involvement, they are: linear, circumscribed, mixed, generalized and pan-sclerotic scleroderma. In turn, the circumscribed form is subdivided into superficial and deep forms. Linear scleroderma can compromise the trunk or the limbs (Fig. 1) or be located on the head, where it is called Coup de Sabre (CDS).[3] Localized scleroderma affects more frequently the female gender[4–8] and its presentation spectrum and clinical course are very varied.

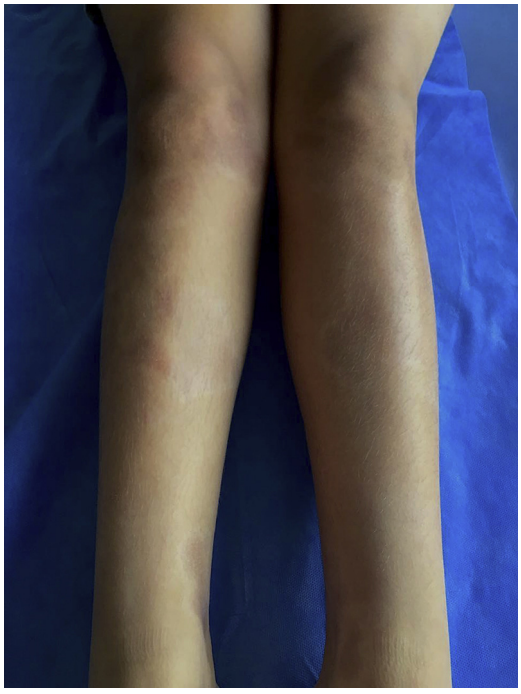

Fig. 1 – Chronic lesions of scleroderma in the lower limbs.