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Case Report

Linear scleroderma en coup de sabre in a teenager - a case report

Esclerodermia linear em golpe de sabre em adolescente – um relato de caso

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ABSTRACT: Scleroderma is an autoimmune disease identified by a progressive tightening of the skin. Scleroderma linear "en coup de sabre" is an unusual and uncertain cause, which in this form can cause progressive focal craniofacial atrophy. This disease is a rare condition that affects mainly children, in some cases besides that cutaneous findings, can result in comorbidity associated with the central nervous system. We describe the case report of a teenage boy (15 years old) with a complaint of the appearance of a hypochromic lesion with progressive growth in the frontal region. The diagnosis was confirmed by capillaroscopy, a dermatological physical exam of the affected area, incisional skin biopsy, and laboratory tests. Oral prednisone in association with topical clobetasol was used as a choice treatment, obtaining satisfactory clinical results with good response and a significant reduction in hypochromia. At the present, this is a rare disease with a lack of knowledge and some health professionals do not understand its difficulties, this report aims to demonstrate the importance of an early diagnosis and adequate clinical treatment to reduce active inflammation of the disease and morbidities, which can significantly reflect on the quality and life span of patients with this rare disease.

Keywords: Localized scleroderma; Morphea; Diseases of the skin and connective tissue; Skin diseases.

RESUMO: A esclerodermia é uma doença de pele, que tem uma origem autoimune, como o próprio nome sugere, ocorre um progressivo enrijecimento da pele. A esclerodermia linear em 'golpe de sabre' é uma variante incomum e de causa incerta que está associada a uma atrofia focal progressiva craniofacial. Essa doença se manifesta principalmente em crianças, em alguns casos além de achados cutâneos, pode ocorrer uma comorbidade no sistema nervoso central. Esse relato descreve o caso de um adolescente (15 anos) com queixa do surgimento de uma mancha hipocrômica em região frontal com crescimento progressivo. O diagnóstico foi confirmado por capiaroscopia, exames físicos dermatológicos da região frontal afetada, biópsia incisional da pele e exames laboratoriais. O tratamento de escolha envolveu o uso de prednisona oral, associado ao uso de clobetasol tópico, obtendo-se resultados clinicamente satisfatórios com boa resposta e melhora significativa da hipocromia. Por ser tratar de uma enfermidade rara, esse relato objetiva demonstrar a importância de um diagnóstico precoce e tratamento clínico adequado para reduzir inflamações ativas da doença e morbidade, o que pode refletir de forma significativa na qualidade de vida de pacientes portadores dessa doença rara.

Palavras-chave: Esclerodermia localizada; Morfeia; Doenças da pele e do tecido conjuntivo; Dermatopatias.

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INTRODUCTION

Scleroderma is a rare disease with different etiologies of the connective tissue that can manifest through cutaneous sclerosis and a large spectrum of systemic manifestation[1].

The pathogenesis of this disease is not yet fully understood, being a combination of vascular disease, autoimmunity, immune activation, and fibrosis. Clinical analyzes show that there is an imbalance between collagen production and destruction; thus, there is an increased synthesis of it, which leads to sclerosis; there is also damage to small blood vessels, activation of T lymphocytes, and rupture of connective tissue[1].

Localized scleroderma or morphea is one of the categories of this disease, it causes an inflammatory skin disorder that can affect soft tissues with devastating functional and aesthetic impairment with biological, social, and psychological consequences. The localized type is distinguished by its predominantly cutaneous involvement, but with the possibility of occasional involvement of the underlying muscles, while the internal organs are usually spared[2,3]. The variant that gives rise to hardened and linear alopecia plaques, sometimes pigmented, is called "en coup de sabre", a rare craniofacial subtype of localized scleroderma.

Treatment for scleroderma should be performed as early as possible to control inflammation. The options range from no medication to the use of corticosteroids, methotrexate, or other immunomodulatory drugs. Treatment should be supervised and prescribed by a rheumatologist and/or dermatologist.

In this context, due to the importance of obtaining an early diagnosis, it is essential to recognize and choose the appropriate clinical treatment, allowing to reduce the active inflammations of the disease. The purpose of this report is to present a case of scleroderma in "en coup de sabre" illustrating a successful option in the treatment of this disease.

CASE DESCRIPTION

GFOC, a 15-year-old black male, complained of the appearance of a hypochromic lesion with progressive growth in the frontal region for 1 year. The patient denied that the disease origin could be associated with exposure to chemicals, including silica, solvents, and hydrocarbons. Capillaroscopy revealed specific microangiopathy with giant loops and he had a known history of repetitive trauma in the region. The dermatological physical examination showed a hypochromic macula associated with atrophy in the left frontal region that extended to the scalp implantation area, with a diameter of 7.5 cm by 3 cm (Figure 1a). The incisional biopsy identified superficial and deep lymphomononuclear interstitial dermatitis, dermal fibrosis, and subcutaneous adipose tissue (Figure 2). Clinical laboratory tests showed non-reactive results for ANTI-CORE Factor and ANTI-SCL-70 antibody. Although classified in limited form or CREST, the patient did not present calcinosis, Raynaud's phenomenon, esophageal involvement, sclerodactyly, or telangiectasia. The case had a slower evolution with the hardening of the skin in the frontal region. Hands, fingers, distal part of the lower limbs, trunk, arms, and thighs were spared. The patient's clinical history, physical examination, laboratory results, and histopathological findings indicated that this was a case of linear scleroderma "en coup de sabre". After analysis of the exams, treatment with oral prednisone was started, dose set by the medical specialist associated with the use of topical clobetasol. This treatment plan showed good results, visually it was possible to observe a significant improvement in hypochromia in 30 days, but there was a worsening of atrophy in the lesion (Figure 1b). As recommended, low doses of corticosteroids with immunosuppressants are necessary in cases with recent cutaneous involvement and severe or progressive pulmonary fibrosis.

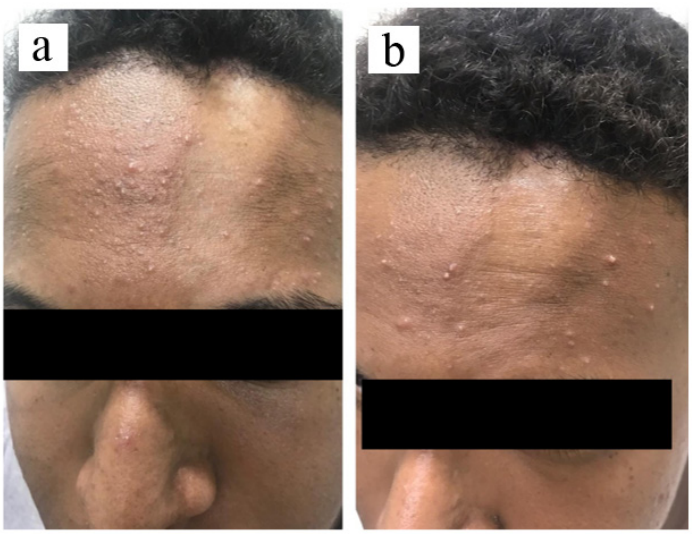

Figure 1. The hypochromic macula is associated with atrophy in the left frontal region. (a) Patient before and (b) after 30 days of treatment with oral prednisone associated with topical clobetasol.

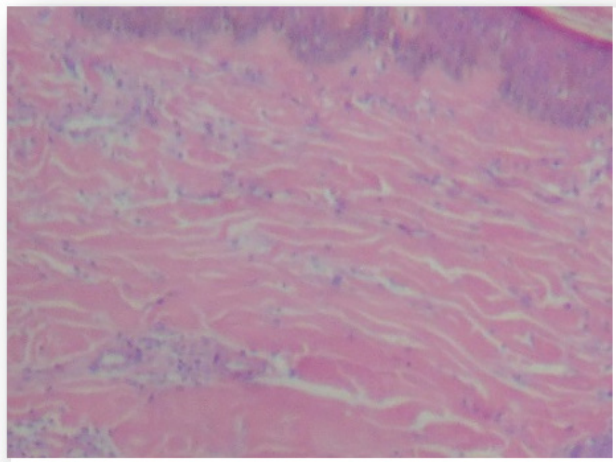

Figure 2. Incisional biopsy identified superficial and deep lymphomononuclear interstitial dermatitis, dermal and subcutaneous adipose tissue fibrosis.

DISCUSSION

The localized scleroderma, as the case of the study presented here, considering parameters such as clinical appearance and distribution, can be divided into five main subtypes: plaque, generalized morphea, bullous morphea, deep morphea, and linear morphea[4]. The linear scleroderma "en coup de sabre" is an even rarer subtype of the disease which is preferably located in a distinct region involving the forehead and scalp frontoparietal[5,6]. In this type it can cause atrophy and furrows of the skin, usually, it manifests as a linear fibrous plaque that involves lesions on the skin, underlying muscles, and bones that can also be involved, resulting in atrophy[7].

Localized scleroderma occurs in both children and adults, children are more susceptible to linear scleroderma on the face, being that the evolution of this abnormality is observed throughout life[8]. In children, about 90% of cases are diagnosed between 2 and 14 years of age[9,10].

The aspect of "en coup de sabre" disease is typically seen in children, although the rapid clinical diagnosis associated with the treatment can lead to softening or regression of the skin lesions, complete resolution rarely occurs, it's necessary to carry out evaluations within the 2 years, after the conclusion of treatment to avoid reactivation of the lesion[11]. Normally, the treatment of scleroderma in "en coup de sabre" is defined according to the prognostic factors of the disease, which allows an improvement in the aesthetic appearance of a patient's skin lesions and the prevention of complications and disabilities as well as deconstruct fear and prejudice that cause discrimination and psychological, moral and social damage to the patient.

In this report, although the treatment did not generate an aesthetic benefit in the atrophy of the lesion, there was an improvement in hypochromia, in addition to an improvement in the individual's quality of life and self-confidence and acts or thoughts. It is a consensus that people with high self-esteem are more willing in all areas of their lives, including self-care or work productivity and the quality of interpersonal ties. Medical management and interpersonal support were essential to improve the well-being physical and mental of adolescents and, consequently, contributed to reducing that initial fear associated with prejudice and discrimination because it is a lesion in the frontal facial region, such psychological problems are caused by self-image negative, anxiety and depression, caused by the social isolation of the adolescent.

On the other hand, if not diagnosed early, it can progress to bone atrophy, mandibular deformity, aesthetic damage by hemiatrophy, or deformity of the skull and central nervous system. These actions depend on the qualification of health professionals involved to identify suspicious signs and symptoms, explain to the patient and forward the patient to exams, define the appropriate treatment and, if necessary, carry out physical and psychological rehabilitation[11,12].

The objective of the localized scleroderma treatment is to obtain control of the lesion avoiding adverse effects, although the treatment, in more severe cases, can be challenging, currently, some therapeutic options can be used as a choice treatment for reduces the active lesions, such as phototherapy, immunosuppressive medications for example corticosteroids and methotrexate. Ultra-potent and oral topical corticosteroids may be useful to reduce inflammation from active superficial lesions of scleroderma.

Scleroderma en coup de sabre is a potentially disfiguring or unaesthetic, there are also surgical therapeutic options for correction of residual atrophy after controlling the inflammatory phase of the disease. Thus, to achieve a good esthetic result can be necessary a complementary procedure, such as the use of synthetic injectable fillers (hyaluronic acid, PMMA, calcium hydroxylapatite), lesion resection, and transplanted or autologous fat grafting[13,14,15]. Although these therapeutic alternatives are considered feasible to improve contours and fill furrows and depressions, fat grafting or autologous fat grafting, which consists of injecting the individual's fat under the skin, is the most used technique. It is possibly related to the low cost and minimal rates of morbidity and less scarring sequelae[16].

In this article, the surgical approach was not necessary, so control of the active inflations of the disease was performed with the use of an anti-inflammatory dose of systemic and non-immunosuppressive corticosteroids, which reduced side effects, revealing the efficiency of the treatment that provided a significant improvement in both hardening and skin atrophy. The disease evolution process can be prevented since the measures are applied at an early stage with appropriate treatment defined by qualified health professionals.

CONCLUSION

Scleroderma linear "en coup de sabre" is a rare disease of the connective tissue, it is developing in a distinct region involving the frontoparietal forehead and scalp. This report sheds light on the importance of early clinical diagnosis during the active phase of the disease. Furthermore, it should be noted that the proposed treatment was adequate and demonstrated a real benefit for the patient, as well as an improvement in the aesthetic appearance of skin lesions and prevention of complications and disabilities. The proposed treatment was adequate and demonstrated a real benefit for the patient, as well as an improvement in the aesthetic appearance of skin lesions and prevention of complications. Therefore, this disease is rare, and it is understood that investigative studies on pathology and more effective treatment options are important for these patients.

Author's contribution*: Marina Cardoso*: Literature review, Case analysis/assessment, and Article writing. *Marina Hernandes Carvalho*: Literature review and Case analysis/assessment. *Flaviane Wendland Fleck*: Literature review and Case analysis/evaluation. *Luana Rocco Pereira Copi*: Article writing, Critical Review and Approval of the Article.

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