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RESEARCH ARTICLE

LOCALIZED JUVENILE SCLERODERMA IN ITS LINEAR FORM: A CASE STUDY

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Abstract

Localized juvenile scleroderma is an autoimmune disease with an unknown cause, marked by skin fibrosis and sometimes involving the underlying fascia, muscles, and skeletal tissue. The disease's severity can vary from isolated skin hardening in one area to severe, disabling conditions affecting the skin, subcutaneous tissue, muscles, and bones if diagnosis is delayed. Here, we present a case of localized juvenile scleroderma from the Pediatrics Department at Mohammed VI University Hospital Center in Oujda.

Introduction:-

Localized juvenile scleroderma (LJS) is a rare autoimmune disease that occurs ten times more frequently than systemic sclerosis in children, with an estimated prevalence of 1 in 100,000 children[1]. Also known as morphea, localized scleroderma is characterized by skin thickening that may extend to underlying tissues, without Raynaud's phenomenon or internal organ damage. Some clinical manifestations can affect functionality and appearance, highlighting the importance of early intervention during the inflammatory phase[2]. The diagnosis of LJS is primarily based on clinical presentation, but a biopsy for histopathological examination is recommended when there is uncertainty. The course of the disease is unpredictable, often spanning a long period. Aesthetic and functional complications may develop, potentially worsening the severity of the condition [3]. Methotrexate has been widely used as a primary treatment for this condition [4]. The development of localized juvenile scleroderma may be influenced by familial and environmental factors [4].

Case Report

We report the case of a 9-year-old boy with no significant medical history, presenting with hyperpigmented, hard, and atrophying skin lesions of linear and smooth appearance on both upper limbs, extending to the axillary folds. The little finger of his left hand was fixed in flexion [Figure 1], with no other associated symptoms. Laboratory tests, including renal, hepatic, and inflammatory markers, were normal. X-rays of major joints, hands, feet, and thorax showed no abnormalities. A skin biopsy of the lesion confirmed a diagnosis of scleroderma [Figure 2]. Thus, the diagnosis of localized scleroderma in its linear form was established. The child was treated with methylprednisolone (30 mg/kg/day for 3 days, followed by prednisone 1 mg/kg/day), methotrexate at 2.5 mg/kg/week, potent dermocorticosteroids, and physical therapy sessions. Favorable progress was noted with treatment, and the child continues to be regularly monitored in outpatient consultations.

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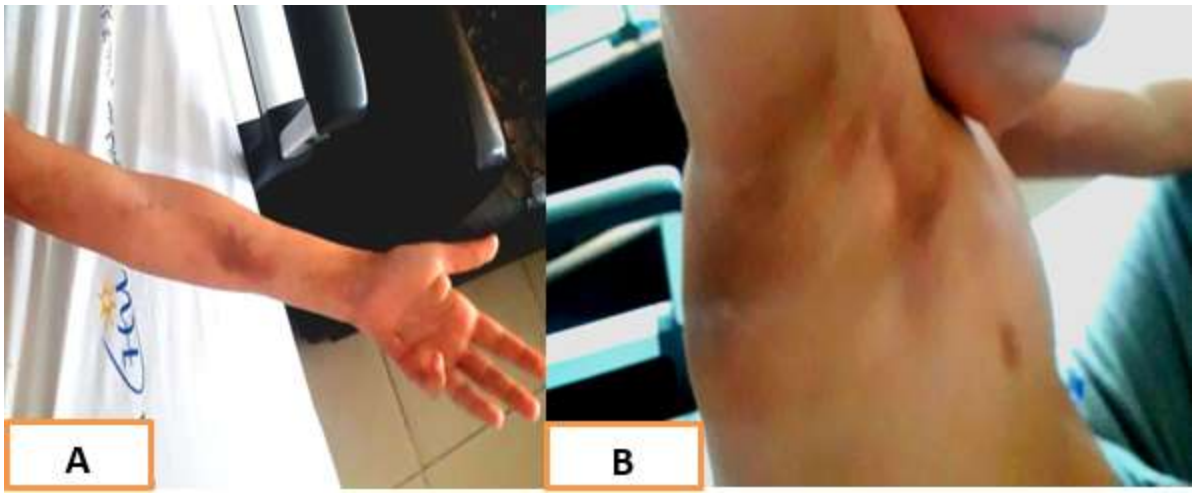

Figure 1 (A and B):- Skin lesions on the left upper limb and ipsilateral axillary fold, hyperpigmented, retractile, hard, and atrophic, presenting a linear and smooth appearance, associated with flexion blockage of the left little finger.

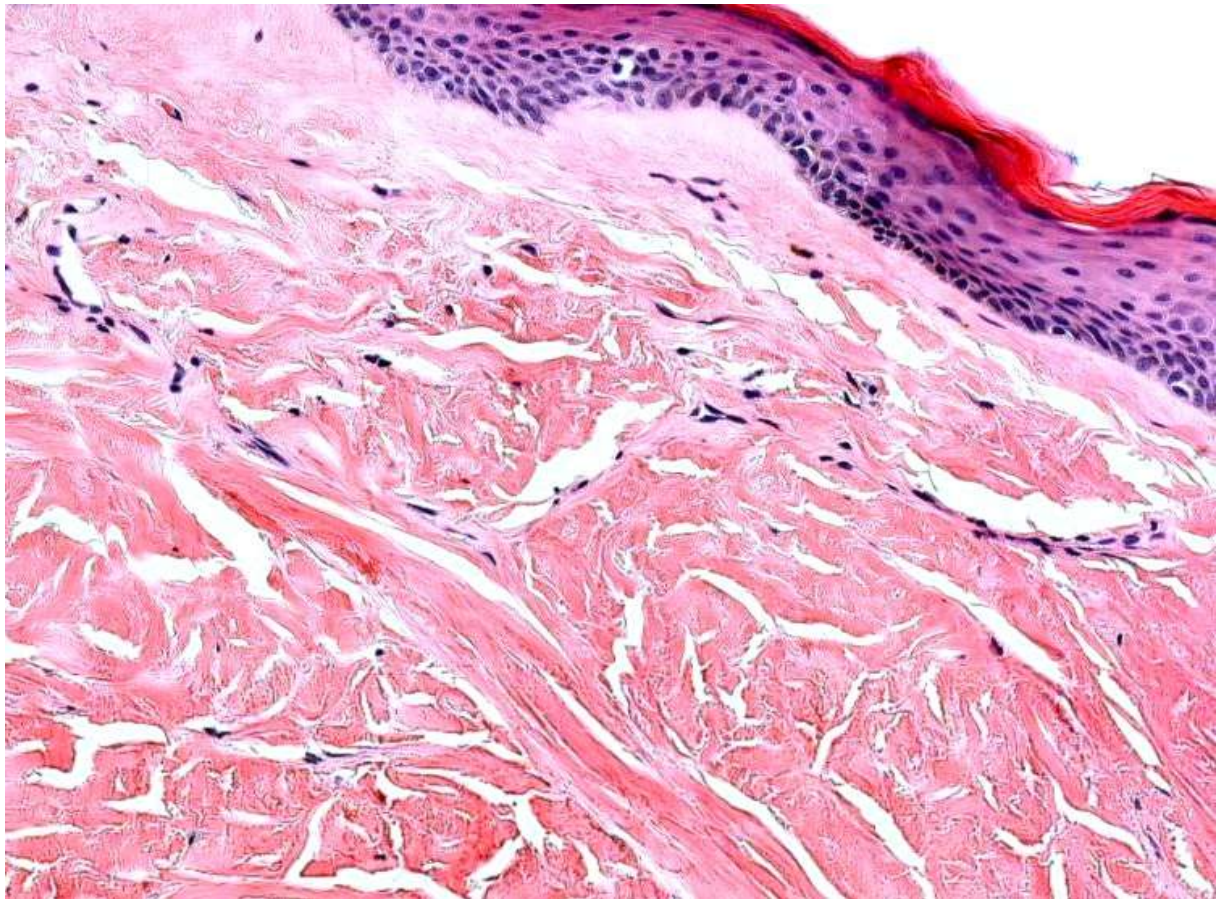

Figure 2:- Photomicrograph of the skin biopsy shows a sclerotic dermis with abundant collagen bundles, absent adnexal structures, and few inflammatory cells (H&E, x200).

Discussion:-

Localized juvenile scleroderma accounts for approximately 15% of all localized scleroderma cases. It primarily affects children aged 7 to 10 years, with a higher prevalence among girls. The incidence of LJS is estimated to be between 0.34 and 2.7 new cases per 100,000 children per year [5]. Linear localized scleroderma is a predominant

subtype of LJS, affecting 51 to 56% of children [6]. Linear localized scleroderma is the most common subtype of LJS, affecting 51 to 56% of children. In children, linear scleroderma of the limbs is notable for its severe progression and a higher risk of developing orthopedic complications compared to adult patients (30-50%) [7,8]. Commonly encountered abnormalities include growth disturbances, limb atrophy, joint movement limitations, and contractures [9]. The risk of bone and joint complications increases when lesions are located near joints [10]. In our patient, a complete flexion blockage was observed in the joints of the left little finger. Extra-cutaneous manifestations occur in 20 to 40% of patients and are the leading cause of death in localized juvenile scleroderma. These manifestations can be seen in all clinical presentations of LJS but are more frequent in its linear variant[11]. A retrospective study involving 750 pediatric patients revealed that the most common extra-cutaneous manifestations were musculoskeletal (19% of all patients, 50% of patients with linear limb scleroderma), neurological (5%), and ophthalmic (3.2%) [4]. Some studies highlight white uveitis as a characteristic symptom of extra-cutaneous localized juvenile scleroderma, primarily when localized to the head. White uveitis affects approximately 3.2 to 8.3% of patients and is characterized by ocular inflammation without clinical symptoms such as pain or redness [12]. Less than 2% of cases present complications affecting the gastrointestinal tract, respiratory tract, and kidneys [4]. Neurological involvement may present as headaches, seizures, neuropathies, behavioral disorders, and concentration difficulties [6]. According to experts, routine diagnostics for organ involvement in children with linear scleroderma are not recommended [12]. Children with localized scleroderma of the limbs should undergo regular rheumatological evaluation due to the potential risk of bone and joint complications [12,13].

The biological assessment of children with localized scleroderma should include tests such as a complete blood count, erythrocyte sedimentation rate, and C-reactive protein analysis. Elevated inflammatory markers are often observed during the early active phases of the disease, especially in cases of linear and deep juvenile scleroderma[6,12]. Patients with generalized or linear LJS often exhibit eosinophilia [6,8]. In cases involving muscle and joint symptoms, especially in scleroderma types such as linear, generalized, deep localized, or disabling pansclerotic scleroderma, it is advisable to check aldolase, creatinine kinase (CK), lactate dehydrogenase (LDH), and rheumatoid factor (RF) levels [6,12,14]. According to current knowledge, routine testing for antinuclear antibodies (ANA), extractable nuclear antigens (ENA), and antibodies against Borrelia is not recommended [8,12,14]. Treatment should be tailored based on the clinical subtype of localized scleroderma, disease activity or severity, age, and gender, as well as clinical subtype, severity of the disease, and extent of skin rash [8,12,14]. Localized juvenile scleroderma is often diagnosed with a delay of one to two years, compromising the efficacy of subsequent therapeutic treatments [5,7]. Studies indicate that localized scleroderma onset in childhood shows more severe progression into adulthood [7]. Children with localized skin involvement in localized juvenile scleroderma respond well to local treatment [12]. Preferred medications include calcineurin inhibitors and calcipotriol, recommended for monotherapy or in combination, administered twice daily for a period of 3 months [5,8]. Topical glucocorticoids (GCS) effectively treat active inflammatory areas of the disease. However, experts advise limiting their use due to the risk of skin atrophy and frequent non-compliance with medical recommendations regarding medication use [12]. Severe forms of localized scleroderma, characterized by extensive tissue involvement (linear, deep, generalized, and mixed types), require immediate aggressive treatment upon diagnosis [12,14]. To achieve optimal therapeutic outcomes, early treatment initiation is crucial, before signs of tissue damage (such as depigmentation, cutaneous, and subcutaneous atrophy) manifest [12].

Due to differing opinions among specialists on first-line treatments, it is recommended to annually organize a joint conference in pediatric dermatology and rheumatology. This conference would focus on pharmacotherapy for severe forms of localized juvenile scleroderma, where pediatric rheumatologists often prefer methotrexate (MTX), while dermatologists favor local treatments [12]. Pediatric rheumatologists recommend introducing methotrexate for patients with localized scleroderma showing significant aesthetic damage or localized rashes around the joints [13]. In 2011, the first double-blind randomized study demonstrated the therapeutic effects of methotrexate in treating severe localized juvenile scleroderma [4]. Zulian et al. reported that with methotrexate, the Computerized Skin Score (CSS) decreased from 1 to 0.79, while with placebo, it decreased from 1 to 1.1 [4]. Moreover, among the 46 patients treated with methotrexate, only 15 (32.6%) experienced disease recurrence, while among the 24 patients receiving placebo, 17 (70.8%) experienced recurrence [4]. According to the Childhood Arthritis and Rheumatology Research Alliance (CARRA) recommendations, methotrexate can be used alone or in combination with orally or intravenously administered corticosteroids [8,14]. The recommended methotrexate dose for children is 15 mg/m2 per week (with a maximum of 25 mg per week), administered subcutaneously or orally, for at least 12 months [8,12,14]. Corticosteroid dosage should be adjusted based on administration method and child's weight. For orally administered prednisone, the recommended dose is 0.5 to 2 mg/kg per day, divided into 2 to 3 doses (maximum of

60 mg per day), for 2 to 4 weeks, followed by a gradual dose reduction. For intravenous methylprednisolone, the recommended dose is 30 mg/kg per day (maximum of 1,000 mg per day), using one of two regimens: either 3 consecutive days every 3 to 6 months or once weekly for 12 months, followed by a gradual dose reduction [5,12]. Combined therapy with corticosteroids (GCS) and methotrexate (MTX) is effective when initiated early in treatment but fails to prevent relapses [15]. Additionally, supplementation with folic acid at a dose of 0.4 to 1 mg per day is recommended during methotrexate treatment [5]. If no clinical improvement is observed after 3 months of treatment or if signs of disease activity persist (such as new lesions or expansion of existing lesions), treatment should be discontinued. After 6 months, consideration should be given to changing treatment [12]. In cases of methotrexate inefficacy or intolerance, mycophenolate mofetil (MMF) is recommended as a second-line treatment for severe localized scleroderma [5,14]. Physical therapy helps prevent functional sequelae, while psychosocial support is essential in treating these chronic conditions. Depressive states are common, affecting between 17% and 50% of cases [3].

Conclusion:-

Localized juvenile scleroderma is a rare disease, classified as a connective tissue disorder characterized by skin sclerosis that can extend to subcutaneous tissues without affecting internal organs. Several forms of localized scleroderma exist based on their clinical manifestations and histological characteristics. Methotrexate has been widely used as the primary medication to treat this condition. Its progression is very slow yet unpredictable. Esthetic and/or functional complications may arise, contributing to the severity of the disease. Long-term outcome data and recurrence rates available in the literature are limited.

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