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Research Article

MODERN TREATMENT AND CORRECTION OF THE CONSEQUENCES OF LOCALIZED SCLERODERMA

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ABSTRACT

Localized scleroderma (LS) is an inflammatory sclerosing disease of the skin and subcutaneous tissues associated with their atrophy. Depending on the subtype, severity of the disease and localization of the lesion, involvement in the pathological process of adipose tissue, muscles, joints and bones, but not internal organs, is noted. The annual primary incidence of drugs in childhood is 3.4 cases per 1,000,000 child population; in females, the disease occurs 2.6–6 times more often. The wide range of clinical manifestations of the disease has led to the emergence of a large number of different classifications, which take into account the severity, prevalence and depth of the fibrosis process, as a result of which five main clinical forms of LS are distinguished: limited, generalized, linear, deep and mixed. The forms of drugs are not mutually exclusive, since the same patient may experience different manifestations of the disease. One of the most common forms of LS in childhood is the linear form, which is observed in approximately 40–70% of children.

KEYWORDS

localized scleroderma, lipofilling, maxillofacial area.

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INTRODUCTION

The linear form of LS is characterized by the presence of one or more linear bands of compaction, which can affect the skin, subcutaneous tissue, muscles and underlying bone tissue [5]. It is usually a single, unilateral lesion with a linear distribution, affecting the extremities, face, or scalp. Lesions are often located along Blaschko's lines (lines of normal development of skin cells that are invisible under normal conditions, but may appear as pathological rashes with a linear or segmental distribution across the skin). When localized on the scalp, a linear lesion appears, often atrophic and slightly depressed, the skin is smooth, shiny, sometimes pigmented. The linear form tends to deform bone structures, causing depressed lesions, and when localized on the face, it can spread to the zygomatic and nasal areas, and the upper lip [6].

In the case of complete damage to half of the face, the process is classified as Parry-Romberg syndrome (progressive facial hemiatrophy) [7]. The disease has a slow, progressive course and usually develops between the ages of 2 and 20 years. It is characterized by unilateral facial atrophy with damage to the skin, subcutaneous tissue, muscles and underlying bone structures, most often the dermatomes of one or more branches of the trigeminal nerve are affected. Atrophy may be preceded by cutaneous induration and discoloration of the affected skin, such as depigmentation or hyperpigmentation, and scarring alopecia is sometimes observed in affected areas of the scalp [8].

In 40% of cases, progressive facial hemiatrophy is combined with linear scleroderma of the "saber strike" type (en coup de sabre). Currently, many authors combine these forms of drugs into one [9, 10]. The course of LS of the "saber blow" type is usually slowly progressive, and the pathological process is usually limited to damage to one half of the face. This form often begins with swelling of the affected area, followed by the formation of a depressed groove in the frontoparietal region, which can then linearly spread to the scalp with the development of cicatricial alopecia. The groove can reach the nose, upper lip and sometimes the gum, which leads to pronounced deformation of these structures; the distance between the teeth and their direction can be changed. The pathological process may involve the bones of the skull, including the facial region. Jaw deformation can lead to malocclusion, poor implantation and atrophy of tooth roots, as well as a delay in their appearance and development [11].

The hypothesis of the genetic origin of LS as a systemic autoimmune disease is supported by the association of cases with a family history of autoimmune diseases and the presence of common HLA types with rheumatoid arthritis. The systemic nature of the disease is also

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indicated by the presence of autoantibodies and increased concentrations of chemokines and cytokines associated with T-helper cells circulating in the blood [10].

Along with the signs of a systemic disease, drugs are characterized by signs of a disease caused by inflammatory fibrosis, namely the formation of a lymphocytic and macrophage infiltrate with the deposition of collagen and fibroblasts [11]. Fibrosis is associated with high concentrations of transforming growth factor beta and interleukin 4 [13]. The tendency to replace normal tissues during fibrosis and destruction of adipose tissue leads to phenotypic changes, including facial atrophy and depigmentation [8].

Assessing the activity of a lesion in LS is crucial in choosing therapeutic tactics. For this purpose, various instrumental methods are used, such as infrared thermography, magnetic resonance imaging, Doppler flowmetry, ultrasound examination (ultrasound), as well as multifactorial assessment systems. Among the latter is the modified Localized Scleroderma Skin Severity Index (mLoSSI), which is equivalent to the modified Rodnan Skin Score (mRSS), used for systemic sclerosis. On a scale of 0 to 3, the Rodnan Index evaluates erythema, skin thickening, and new lesions in 18 different anatomical sites and can be used in both adults and children. To assess active inflammatory lesions in LS, as well as the therapeutic effect, the LoSDI skin damage index (Localized Scleroderma Skin Damage Index) was developed, which evaluates cutaneous and subcutaneous atrophy, as well as the degree of dyspigmentation. The combination of LoSDI and the Physician's Global Assessment (PGA) is designated the Localized Scleroderma Cutaneous Assessment Tool (LoSCAT), which helps the practitioner evaluate both active and inactive lesions [1] .

Therapeutic tactics for managing patients with LS depend on many factors: the activity of the pathological process, localization of rashes (foci of local inflammation (erythema, edema) followed by the formation of sclerosis and/or atrophy of the skin and underlying tissues), form of the disease, age patient. Local therapy usually includes glucocorticosteroids (GCS), calcineurin inhibitors, vitamin D analogues, and phototherapy. Systemic therapy, in turn, is effective for common and severe forms of the disease. The most commonly used systemic approach is a combination of corticosteroids and methotrexate [5]. When planning treatment, it should be borne in mind that clinical effects sometimes appear no earlier than 3 months after the start of therapy [2]. Mycophenolate mofetil (MPM) has been proposed as an alternative immunomodulatory agent in cases of methotrexate resistance [1-4]. In vitro studies have shown that MPM suppresses the proliferation of lymphocytes, as well as other types of mesenchymal cells, including smooth

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muscle cells and fibroblasts [4]. Using a series of cases of drugs resistant to methotrexate therapy, it was shown that the use of MPM leads to a decrease in the degree of skin sclerosis and inflammation (according to infrared thermography and clinical assessment) [6]. In recent studies, the combination of GCS and methotrexate/MFM showed inconsistent effectiveness; several cases of drugs were presented in adolescents who did not respond to such treatment [7]. The effectiveness of an alternative treatment method for drugs in adults and children has been reported using the drug abatacept, a recombinant fusion protein that blocks T-cell activation, approved in the USA and the Russian Federation for the treatment of rheumatoid arthritis [10] .

RESULTS

Particular attention is paid to the search for treatment methods for drugs that can eliminate the consequences of the disease, namely gross cosmetic defects of the face that negatively affect the harmony of the physical and psycho-emotional development of the child. Recently, fat grafting has become the focus of attention (due to the effectiveness of the method in recreating volume and improving skin quality). Fat transfer, including adipocytes, adipose stem cells, endothelial cells and vascular smooth muscle cells, has been shown to reduce inflammation as well as fibrosis by limiting the synthesis of extracellular matrix proteins and promotes increased collagenase activity, as well as providing structural support. support due to proliferation and differentiation of stem cells [11] .

Below is a description of a clinical case of LS in order to demonstrate the possibilities of correcting skin defects in children.

CLINICAL CASE STUDY

Patient A., 1 7 years old, complained of skin atrophy in the forehead area. In 2019 , the diagnosis was made: “Localized scleroderma, linear form.” The skin pathological process at the time of diagnosis was linear in nature and was represented by a focus of atrophy of the skin and underlying flesh-colored tissues. The lesion was localized on the skin of the forehead with transition to the skin of the scalp and supraorbital region. The size of the skin atrophy is 6 cm in height and 3 cm in width. When performing an ultrasound of the skin and soft tissues of the supraorbital region, forehead, and scalp, thinning of the skin and subcutaneous fat in the supraorbital region, as well as in the forehead on the right, was noted.

The patient received methotrexate therapy for two years with positive dynamics—a decrease in the clinical activity of the disease and stabilization of the skin pathological process (reduction in the severity of inflammation) were noted. After 6 months she was hospitalized for further examination. A general assessment of the state of health and monitoring of the titer of antibodies to single-stranded DNA

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(negative test) and to the Scl-70 antigen (negative test) were carried out. The patient was recommended for contouring with autologous fat (lipofilling), which was performed in the plastic surgery department.

After treating the surgical field under intravenous anesthesia using a cannula with a diameter of 2.7 mm and a length of 23 cm, syringe lipoaspiration was performed in the area of the inner thighs. Fat was obtained in a volume of 30 ml. A cannula was used to perform a blunt detachment of scar tissue from the underlying structures. The introduction of adipose tissue into the recessed areas was carried out using a cannula with a diameter of 1.6 mm in a volume of 23 ml: in the scalp area - 3 ml, in the frontal area - 15 ml, supra-orbital area - 5 ml. The remaining 7 ml of fat graft were treated with nanotransfer (Tulip nano system, USA), a cell fraction was obtained, which was injected intradermally into the affected area. The postoperative period was uneventful. Methotrexate therapy was discontinued.

CONCLUSION

Pathogenetic therapy of drugs does not have a significant effect on the elimination of cosmetic defects accompanying the disease. In this regard, the most promising method for correcting a skin defect is contouring with autologous fat (lipofilling), which allows you to recreate the natural contour and fullness in the affected area. Autologous fat grafting may be an effective therapeutic alternative in patients with LS. The presented clinical observation demonstrates the effectiveness of using the method in a teenager - leveling the cosmetic defect of the soft tissue structures of the facial skull.

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