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Lichen Sclerosus of Anogenital Localization in Children.

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ABSTRACT

Lichen sclerosus (synonyms: lichen sclerosus et atrophicus, morphea guttata, von Zumbusch's disease, white spot disease, lichen sclerosus type, atrophic spot leukoderma, etc.) was first described by Hallopeau H. (1887). Chronic autoimmune dermatosis is one of the clinical options for circumscribed scleroderma, characterized by severe focal atrophy of skin and mucous membranes of the external genital organs. The article presents modern literature data on the etiology and pathogenesis, and localization features, clinical manifestations and treatment approaches in children with lichen sclerosus of anogenital localization.

Keywords: circumscribed scleroderma, lichen sclerosus, anogenital localization, vulvar lichen sclerosus, penile lichen sclerosus, lichen sclerosus of anogenital localization in children.

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Lichen sclerosus is a clinical option of circumscribed scleroderma. Lichen sclerosus was first described by H. Hallopeau at the end of the 19th century in 1887. Subsequently, a number of synonyms for this pathology appeared in the literature: morphea guttata, von Zumbusch's disease, white spot disease, lichen sclerosus type, atrophic spot leukoderma, etc. [1-5]

Lichen sclerosus is a steadily progressing chronic autoimmune dermatosis, characterized by severe focal atrophy of skin and mucous membranes of the external genital organs [1-5,6]. Around the world, there has been an increase in the number of patients with dystrophic diseases of the external genital organs, if earlier this pathology was mainly found in women of menopausal and postmenopausal age, then at present vulvar lichen sclerosus is often diagnosed in children and women of reproductive age. [6]

Two peaks of the incidence of lichen sclerosus in children are distinguished. So, in girls, lichen sclerosus occurs most often in the neutral period of puberty (88%), but can also occur in the prepubertal period (12%). The average onset age of the disease in girls is 5.4 y. [7] The incidence in childhood is about 0.1% of the number examined ones [8]; the prevalence of lichen sclerosus in girls is 1:900 [79].

The etiology and pathogenesis of lichen sclerosus has not been fully studied, despite numerous studies that examined the polygenic nature of inheritance and the multifactorial nature of the process (various exogenous and endogenous triggers: infectious, endocrine, physical, chemical). [9]

The development of lichen sclerosus is associated with a number of endogenous factors, in particular, with a genetic predisposition, neuroendocrine and dishormonal disorders, microcirculatory disorders, stress, and chronic inflammatory processes. The exogenous effects affect the nature of the clinical picture and the course of the disease: hypothermia, mechanical damage, viral and bacterial infections. [10-12]

According to some authors, a significant role in the development of the pathogenesis of lichen sclerosus is played by the autoimmune mechanism, as evidenced by the frequent association with autoimmune diseases, such as autoimmune thyroiditis, alopecia areata, vitiligo, autoimmune gastritis, systemic lupus erythematosus, pernicious anemia, diabetes mellitus. [13,14]

However, the endocrine hypothesis, according to which low endogenous estrogen levels are often determined in patients with anogenital lichen sclerosus, plays an important role in the development of lichen sclerosus of anogenital localization. [15]

The literature widely discusses the participation of estrogens and progesterone, as well as some other hormones in the synthesis of collagen and other components of connective tissue in the development of lichen sclerosus. According to studies, a number of authors revealed a low level of estrogen synthesis and a low level of estrogen saturation of target organs in girls with lichen sclerosus of anogenital localization, which led to later puberty and symptoms of menstrual dysfunction. [16,17,18] The role of hormone imbalance in the development of the disease emphasizes the fact that resolving of foci of lichen sclerosus in girls occurs mainly in the prepubertal period of puberty — on average in 55.6% of cases, in other children — with the onset of menarche. With age, the level of estradiol increases, as a result of which, in some cases, regression of pathological foci is possible [17,18]. In a prospective study of S. Smith et al. [19] 75% of the examined patients of 12 patients with genital lichen sclerosus after puberty had an active course of the disease.

In the development of lichen sclerosus of anogenital localization in children, the role of infectious factors in the development of the disease is also discussed. So, in a study of scientists under the supervision of which there were 48 girls, it was noted that 60% revealed infectious diseases of the genitourinary system, among which there were: Candida albicans, Chlamydia trachomatis, mycoplasma and ureaplasma infections, human papilloma virus, herpes simplex virus, Epstein–Barr virus. [19,20,21]

A possible factor in the occurrence of lichen sclerosus can be chronic traumatic injury. Helminthic invasions, allergic manifestations, which are accompanied by itching, can act as traumatic agents in the genital area in children. [22]

Damage to the anogenital zone of lichen sclerosus can be either isolated or widespread on the skin, spreading to the neck, shoulders, mammary glands, trunk, extremities. In girls with lichen sclerosus of the

anogenital region, the zones of the external genital organs are mainly affected: labia majora and labia minora, clitoris, posterior commissure with a transition to the anus, forming a specific outline (in the form of a figure eight). (fig. 1) Features of localization in the vulva area are the appearance of white papules or plaques in the anterior portion of the vulva and around the clitoris, while the plaques appear hypopigmented and their surface resembles tissue paper. Gradually, the number and size of papules increase, they soon merge, then the epidermis becomes thinner and atrophy occurs. Subjective sensations are characterized by paresthesia – prickling sensation, goose bumps, mild itching, provoking scratching and trauma to the vulva (often in a dream, unconsciously). [22,23,24] Lichen sclerosus progression proceeds against the background of inflammation and swelling of the mucous membrane and perineum tissues, the process of tissue sclerosis gradually begins, atrophy of the external genital organs appears, which leads to the loss of the vulva structure and its reduction. [24,25]

A feature of the localization of lichen sclerosus in boys is the location of papules circularly around the penis, as well as on the inner and outer leaf of the foreskin, including on the skin of glans penis. [25] With a similar localization of lichen sclerosus rashes, the development of phimosis and paraphimosis is possible [25,26]. Atrophic white spots or plaques are a typical manifestation of lichen sclerosus on glans penis; lilac, slightly exfoliative spots with telangiectasias and weak purpura are also described. At first, the course of lichen sclerosus is almost asymptomatic, but in the future there may be itching, burning and soreness. The progression of lichen sclerosus leads to a smoothing of the frenulum of glans penis, and the process also captures urethral tissue. Urethral stenosis can lead to problems with urination and dysuria, urethral disease can be a serious complication. [25,26,27]

The following polymorphic clinical forms of lichen sclerosus of anogenital localization in children are also distinguished: papular, erythematic-edematous, vitiliginous, atrophic, erosive-ulcerative. Papular form is represented by isolated flat papules, leaving behind atrophy sites. Rashes in girls, as a rule, are located on the inner surface of the labia majora without involving the perianal region, as a result of the resolving, whitish areas of superficial atrophy remain at the site of the rashes (subjective sensations are usually mild); Erythematous-edematous form is presented in the form of whitish atrophy sites against the background of obvious hyperemia and edema of the labia majora, prepuce, less often with spread to the perianal region. In girls, it is often accompanied by vulvovaginitis; Vitiliginous form is a common form, manifested by foci of superficial atrophy, cracks, depigmentation, localized in the posterior commissure, the glans penis, and foreskin. Subjective sensations are absent. Doctors of related specialties often mistakenly regard these manifestations as leukoplakia or vitiligo; Bullous form is represented by subepidermal vesicles with serous and/or hemorrhagic contents against the background of atrophy and hyperemia of the mucosa. Subjectively patients have itching, burning, soreness. The most commonly described clinical picture is regarded as herpes. The bullous form of lichen sclerosus is characterized by a persistent course, resistance to ongoing therapy; Atrophic form is characterized by foci of circumscribed atrophy of the mucous membrane according to the type of “tissue paper” and frequent involvement of the perianal region; Erosive-ulcerative form is characterized by spontaneous formation of bleeding painful erosions or ulcerative defects against the background of hyperemia and atrophy of the mucosa without previous blistering. Patients are concerned about itching and soreness in the lesions. [28,29]

Lichen sclerosus of anogenital localization, regardless of the clinical picture, is characterized by a long chronic course of the disease, as well as repeated exacerbations up to the puberty.

A typical histological picture of lichen sclerosus is atrophy of the epidermis, hydropic degeneration of the cells of the basal layer, edema and pronounced lichenoid infiltrate in the area of the dermo-epidermal junction. Homogenization of collagen fibers is detected in the papillary dermis (Fig. 2). [30]

Peculiarities of the localization of lichen sclerosus on the genital organs makes it difficult to conduct a differential diagnosis with nonspecific balanoposthitis, atrophic form of lichen planus, vitiligo, contact dermatitis, etc. [1,7,8,9,12,17,24,25,30], and the development of severe complications in the form of urethral stricture, phimosis and paraphimosis of the penis, atrophy of the vulvar lips and vaginal mucosa, gross cosmetic defects in the anogenital zone.[23,24,25,26,27]

The means that act on the pathogenetic aspects of the mechanism of development of the disease are mainly used in the treatment of children with lichen sclerosus of anogenital localization. The main purpose of

therapy is to slow the progression of the disease, to achieve stabilization of the process, and in the future, to regress the clinical picture. The treatment should be multi-course and comprehensive, with the mandatory use of penicillin family antibiotics, hyaluronidase-based products, microcirculation-improving products, immune regulators, vitamins, enzymes, and physiotherapeutic methods. [30,31,32,33] Ointments stimulating reparative processes that have anti-inflammatory effects are used as the main external agents for lichen sclerosus of anogenital localization. [32,33,34]

Thus, the systematization of the clinical manifestations of lichen sclerosus of anogenital localization in children is undoubtedly of interest to both dermatologists and pediatricians. Timely diagnosis of lichen sclerosus of anogenital localization determines not only the right choice of therapeutic approach to the disease, the absence of progression of the process, but also the prevention of a number of complications, such as urethral strictures, phimosis and paraphimosis of the penis, atrophy of the vulvar lips and vaginal mucosa, gross cosmetic defects in the anogenital zone.

Fig. 1. Lichen sclerosus with lesions of labia majora, labia minora, clitoris and perianal region

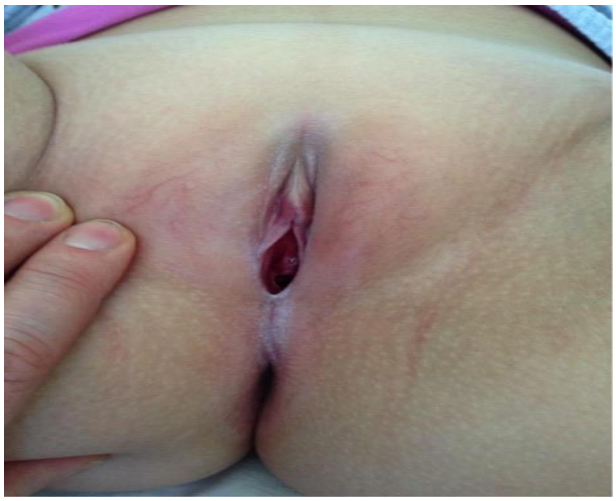
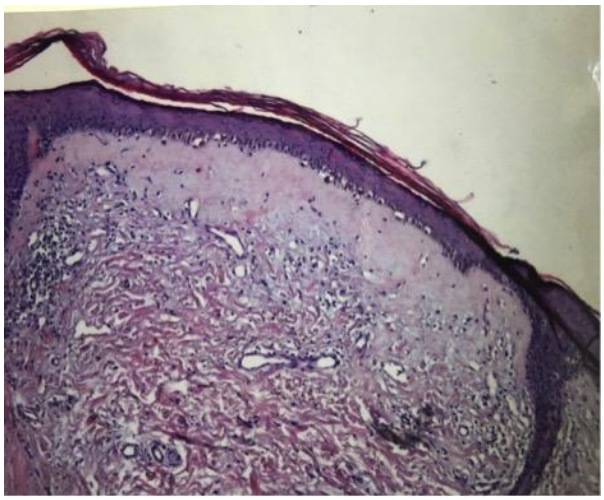


Fig. 2. Histologic pattern of lichen sclerosus et atrophicus of anogenital localization in children



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