Patient suffering from lichen sclerosus together with partial labial fusion (adhesion) and inflammatory infection of the vulva, vagina and anal area

G. Jarząbek-Bielecka1, M. Mizgier2, J. Boroch1, M. Pisarska-Krawczyk3, M. Pawlaczyk1, E. Jakubek3, W. Kędzia5

1Division of Developmental Gynecology and Sexology, Department of Perinatology and Gynecology, Poznan University of Medical Sciences, Poznan 61-701
2Department of Morphological and Health Sciences, Dietetic Division, Faculty of Physical Culture in Gorzów Wlkp., Poznan University of Physical Education, Gorzów Wlkp. 66-400
3The President Stanisław Wojciechowski Vocational State School in Kalisz, Kalisz 62-800
4Department of Geriatrics and Gerontology, Faculty of Health Sciences, Poznan University of Medical Sciences, Poznan 61-701
5Department of Law, Organization and Management in Healthcare, Faculty of Health Sciences, Poznan University of Medical Sciences, Poznan 61-701
6Department of Perinatology and Gynecology, Poznan University of Medical Sciences, Poznan 61-701 (Poland)

Summary

The etiology of lichen sclerosus is not fully known, due to its chronic and progressive nature. The predisposing factor is physiological hypoestrogenism, hence the condition concerns girls during their developmental phase considered as estrogen silent (even as early as at the age of 6 months). The paper presents a 5-year-old patient coming from a rural area and suffering from lichen sclerosus together with partial labial fusion (adhesion) and inflammatory infection of the vulva, vagina and anal area. These symptoms intensified after antibiotic therapy the girl had undergone due to a respiratory tract infection. There were changes in the skin of the labia majora and buttocks indicating lichen sclerosus - swelling, traces of scratches, secondary redness, partial fusion of the labia minora on the side of the crotch known as the fourchette, and a runny yellow-white discharge from the vagina.

Key words: Inflammatory infection; Lichen sclerosus; Labial fusion.

Case study

The patient AB, age 5, came with her mother to the gynecologist because of persistent, pain-prone itching of the vulva and anal area, frequent recurrent urinary tract infections and a disconcerting vaginal discharge. These symptoms intensified after antibiotic therapy for her respiratory tract infection. Changes in the skin of the labia majora and buttocks indicated lichen sclerosus - swelling, traces of scratches, secondary redness, partial fusion of the labia minora on the side of the crotch called the fourchette, and a runny yellow-white vaginal discharge.

According to Tanner scale (A1Th1P1: height 110 cm, body weight 21 kg), the girl was well developed in terms of her sex and age: internal genital organs palpation and ultrasonography unmodified - uterus infantilis with linear endometrium, and ovaries of normal size with small blisters with a diameter of max. 4 mm. A 4th bacteriological swab was collected - no acid-producing bacteria, and numerous pathological bacteria present, as well as leukocytes and fungi (yeasts). Such findings are rare when it comes to young girls, and these are due to previous antibiotic therapy while treating the respiratory tract infection.

The patient underwent vulvar biopsy under general anesthesia at the hospital and the histopathological result confirmed the suspicion of lichen sclerosus.

Diseases of the vulva and vagina in gynecology of the developmental age

Inflammation of the vulva and vagina is a gynecological problem, regardless of the patient’s age, from as early as the neonatal period to senectitude. In the gynecology of the developmental age, such inflammation is most commonly observed between the ages of 3 and 10. The etiopathogenesis of vulvovaginitis in this girl’s case is related to both her age and her systemic diseases. The importance of premature sexual activity or the problem of sexual harassment is also emphasized. Before menarche, inflammatory changes occur more often in the area of the external genital organs, while after menarche, it is mainly the internal genital organs that are inflamed.

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According to the literature, 25-60% of girls with vulvovaginal inflammation do not show any pathogen in the bacteriological examination (the cause of symptoms may be, for example, atopic dermatitis), and in 60% with genital infections along with urinary tract infections (a urine test should be an inherent element of STI diagnosis regarding girls). In etiopathogenesis, transmission pathways are crucial. These include the respiratory system - upper respiratory tract infections, strep throat (Streptococcus spp., Staphylococcus spp.), gastrointestinal tract - fecal contamination (Enterococcus spp., E. coli), urinary system infections, and congenital malformations (Gram (-), Pseudomonas spp.), home environment (care processes), the sexual path (for sexually active girls) and the vertical path. Fungal infections of the genital organs are very rare in girls, especially before puberty (during what is called 'hormonal silence'). They occur in about 0.5-1.5% of cases, mostly as multifocal invasions. Nonetheless, factors such as diabetes, long-term antibiotic therapy, immunosuppression, congenital immunodeficiency, allergic diseases and allergic rhinitis predispose to the presence of fungal infections among girls. The etiological factors of vulvovaginal inflammation in girls are also parasitic infections pinworms, human roundworms, pubic lice, scabies, and infectious mollusk (9-12% of cases of girls’ genital infections include a pinworm infection) [1, 11].

Treatment of vulvovaginitis in girls should be matched to the result of the bacteriological examination and antibiogram. For girls with severe local and / or generalized symptoms and a persistent infection, it is recommended to use general antibiotic therapy consistent with the result of the antibiogram, whereas with a mild clinical course of infection, local antibiotics (ointments, creams, drops, stamens, vaginal globules) are recommended. During therapy, it is crucial to use antihistamines orally and topically, avoid allergenic products, in case of allergic conditions (i.e. seafood, chocolate, nuts), and cleanse the vulva regularly, using half-baths and refraining from long baths with hot water. Particular attention should be paid to maintaining personal hygiene; taking special care of the perinaeum area (2-3 times a week), and using topical soothing products such as olive emulsions or glucocorticosteroids. It is advisable to wear loose, cotton underwear and use emollients and neutral ointments containing vitamins A, D and fatty acids, among others, to prevent labial fusion (adhesion) once more. Unfortunately, regardless of the method of treatment, frequent recurrences of the symptoms are observed (about 10-15% of cases).

**Etiology of lichen sclerosus**

The etiology of lichen sclerosus is not fully known – it is chronic and progressive. The predisposing factor is physiological hypoestrogenism, hence the condition concerns girls during their estrogen silent developmental phase (even as early as the age of 6 months). Symptoms imitate recurrent inflammation of the genital organs. It is difficult to determine the incidence of lichen sclerosus, as many girls suffering from this problem before puberty do not go to specialists. It is thought that about 1 in 900 girls might have this condition. It may also be the case that a full diagnosis is made late, at about the age of 10. Some caregivers mistakenly identify the condition as the result of sexual harassment.

The girl examined in our case had symptoms typically associated with lichen sclerosus, not differing significantly from a bacterial and fungal infection of the external genitalia and anus (intensifying symptoms i.e. pruritus, vulvar burning, dysuria and bowel dysfunction. According to the literature, in 10% of cases, the disease may be asymptomatic (in the case described, there was an additional bacterial and fungal superinfection after antibiotics were used earlier for an infection of the respiratory tract). The authors state that in 8-10% of cases, LS changes also occur outside the genital area - on the skin of the chest, chin, neck, wrists and face. However, the girl had changes observed only in the genital area and anus. When it comes to adolescent girls, the prognosis is optimistic, as during their puberty, spontaneous remission may occur and oncogenesis is rare. With respect to adult women, the prognosis is not so good - chronic treatment is usually required.

Once lichen sclerosus is histopathologically confirmed, the use of topical steroid preparations (ointments, emulsions) is recommended as the first-line therapy. Data from the literature indicate the importance of using 0.05% clo-
betasol propionate, which is characterized by the highest efficiency. Of course, personal hygiene, wearing cotton underwear, avoiding tight underwear and injuries around the vulva and crotch are important. According to the literature, in the case of girls, the use of subtherapeutic doses of topical steroids is a mistake in the treatment of lichen sclerosus, which is associated with ineffective treatment and the need to extend therapy. The authors state that in the absence of desirable effects from first-line therapy, alternative methods may be attempted: photodynamic therapy, which has particularly good effects in this age group, and tacrolimus, a calcineurin inhibitor (0.1% Protopic) applied twice a day for 4 months and then twice a week for 6 months as maintenance treatment. As the authors emphasize, the use of tacrolimus preparations in the treatment of LS takes place only in the phase of clinical trials, but the effectiveness is much higher than in the case of topical steroid preparations, especially when it comes to young children. The use of this therapy, as the authors point out, does not cause undesirable effects such as labia atrophy, as is the case with local steroid therapy. To quote the authors, recommendations for LS treatment include local anti-inflammatory substances (on average 3-4 months) and long-term follow-up (at least once/year), due to the high risk of recurrence and the possible development of vulvar cancer in adulthood. In general, the prognosis for the treatment of LS with regard to children is good. With rapid diagnosis and correct therapy, complete remission is often attained. It is also believed that if in the course of LS the children fail to achieve remission until menarche, the prognosis becomes less favorable (difficulties in achieving complete remission). Moreover, anatomical and physiological conditions of the genital organs - moisturizing and the proximity of the urethra, anus and the rectum - are conducive to the development of inflammation.

**History of vaginal ecosystem research**

- 1892 Doderlein describes lactobacilli as a microbiota of normal vaginal discharge
- 1914 Curtis confirms the dominant role of lactobacilli, at the same time stating that the vagina is inhabited by other bacteria
- 1938 Weinstein reveals the presence of anaerobic bacteria in normal vaginal discharge
- around 1930-1940 Cruickshank and Sharman describe the vaginal microbiota in the uterine cavity (sterility), newborns, pre-menarche girls, sexually active and post-menopausal women.

Inflammation of the genital organs is most often caused by hygienic negligence, mechanical injuries or infections (viruses, chlamydia, fungi, and protozoa). A certain group of these inflammations also occurs on an allergic basis (such as an allergy to cleaning products).

A newborn female that is under the influence of estrogens from intrauterine life has acidic pH of the vagina as a result of colonization with acid-bearing bacteria Lactobacillus acidophilus (lactobacilli are a normal component of the vaginal bacterial flora). In the following months of the girl’s life, the estrogen concentration gradually decreases; the vaginal epithelium becomes thin and consists only of the basal and ground layer, without the content of glycogen in cells, which determines the development of lactobacilli. The acid reaction of the vagina changes to neutral until early puberty. During the physiological absence of estrogens, or the lack of defensive biochemical mechanisms, a mixed bacterial flora composed of kernels and sticks can be established and thrive in the vagina. A few millimeters of vaginal light, the adherence of its walls, the presence of hymen and the so-called palmate folds (plicae palmate) in this period of life mechanically compensate for the lack of antibacterial acid pH. During puberty, the concentration of estrogen gradually increases and is visible in the light of the hymen as opalescent, pearly-white mucosal walls, due to the fact that the vaginal epithelium becomes multilamelar and the blood vessels are invisible. The cells of the intermediate layer already contain glycogen, which allows the development of lactobacilli and during this period prevents the settlement of pathogenic microorganisms [1-4, 11].

In the late period of puberty, further defense mechanisms develop, i.e. intensification of exfoliative processes of the genital mucosal epithelium, formation of active secretion removed by the growth of the epithelial recruitment within the uterine cavity and fallopian tubes or passively with microorganisms and cell debris and acidic pH in individual sections of the genital organs.

Proper biocenosis of the genital environment determines the protection against the development of inflammatory processes. When it comes to girls, the protection of the female genitalia against infection determines the mutual complementation of the subsequent local defense factors.

The vaginal environment is an independent, stable biocenosis, in which the most important component are Lactobacillus acidophilus, in particular L. acidophilus, L. fermentum, L. plantarum, L. delbrueckii, L. rhamnosus. These gram-positive bacilli are facultative anaerobic or microaerophilic, rod-shaped, non-spore-forming bacteria that come in various forms.

Vaginal biocenosis is a stable system, however, it is subject to continuous changes, albeit minor in quantity. Not only does the number of lactobacilli change, but also bacteria from the gastrointestinal tract which appear and with a properly functioning protective barrier die. Thus, there is constant struggle for a place within the vaginal mucosa and in the physiological state small quantities of other bacteria may appear next to lactobacilli. The maintenance of the physiological state (normal vaginal biocenosis) is affected by the concentration of estrogen in different periods of life, i.e. puberty, pregnancy, sexual cycle, menopause. Moreover, it is affected by surgical procedures, compliance with hygiene rules, sexual habits, and uncontrolled antibiotic therapy [5, 6, 7, 11].

In order to estimate the vaginal biocenosis correctly, one
can look at images showing vaginal discharge stained by the Gram method, then distinguish and classify it according to the appropriate degree of purity of the vagina. In Poland Kuczyńska’s modified scale is used to visualize the current condition of vaginal biocenosis with the determination of the physiological state, inflammation and ratio between groups of microorganisms. The basic criterion for the evaluation of biocenosis, according to Kuczyńska, is the presence or absence in the vaginal content of lactobacilli. Dysfunction of normal microbiota creates the possibility of infection by strains of high invasiveness and pathogenicity, leading to inflammation of the vulva and vagina, ascending to the internal genital organs. The effectiveness of defense mechanisms and their mutual compensation varies depending on the periods of the woman’s life, which are hormonally conditioned. Some particularly common factors that facilitate inflammation are: anatomical structure (close vicinity of the vulva and anus), not fully developed labia, lack of pubic hair, low estrogen concentration affecting vaginal mucosal sensitivity to irritants and infectious agents, exposure to various irritating agents (such as bath liquids or rough towels), foreign bodies in the vagina, and neglectful hygiene of a child older than 1 year (infants are routinely bathed every day, in contrast to older children).

Vulvovaginitis among adolescent girls

75% of all gynecological visits paid by adolescent girls before their menarche are due to inflammatory infection of vulva and vagina - vulvovaginitis. However, there is a need to differentiate inflammatory diseases of the vulva from dermatoses. The symptoms of vulval diseases, which are reported by patients, often require consultation and dermatological treatment. Dermatoses are not only located on the genitals (the vulva is only one of the places covered by the disease process), so it is worth examining all the patient’s skin. Nonspecific vulvovaginitis accounts for 25-75% of all vaginitis in this age group.

Symptoms of vulvovaginitis with regard to adolescent girls:
- pruritus,
- swelling and redness of the labia,
- abnormal vaginal discharge,
- fractures, a characteristic feature of chronic inflammation,
- traces of scratches and abrasions on the vulva skin,
- diarrheal symptoms (infection of the urethra and perianal glands),
- urinary tract infection.

For girls who have not experienced sexual intercourse, inflammation of the internal sexual organs is relatively rare. Nonetheless, in the literature, vaginitis was reported as associated with purulent inflammation of the palatine tonsils or teeth. Moreover, continuous and long-lasting appendicitis, which has been altered and inflamed, may cause the inflammation to penetrate into appendages on the right side, eg. in the form of plastron. Plastron or peri-opical abscess caused by the perforation of the appendix in the phase of an acute inflammatory process covers the surrounding tissues and does not spread to the entire peritoneal cavity. The symptom of abscess formation is extremely painful, often perceptible by means of an abdominal tumor, in McBurney’s point, accompanied by symptoms of appendicitis. Treatment of the abscess is initially conservative, with antibiotics and low-fat diet being used, and after the end of the inflammatory process, appendectomy may be considered. Another disease with unclear etiology occurring in this age group is lichen sclerosus, which causes pruritus of the skin and secondarily, by the compulsion to scratch in this area, the child experiences an infection of the vulva, vagina and full-blown vulvovaginitis. Treatment is a symptomatic preparation of antibiotics, antihistamines, ointments with vitamin A and estriol cream, which significantly reduces the symptoms [8-12].

Etiological factors of genital inflammation

Among the etiological factors of genital inflammation, the following should be mentioned:

ENDOGENIC FACTORS:
- hypoestrogenism,
- atrophic changes,
- dermatosis,
- avitaminosis,
- psychosis,
- systemic diseases (e.g. diabetes),

EXOGENOUS FACTORS:
- abiotic (mechanical, thermal, chemical, hygiene),
- biotic (bacteria, viruses, fungi, chlamydia, protozoa, pinworms and others).

Conclusions

To conclude, lactobacilli play an important role in protecting sexual organs against infections and inflammations as they:
- provide acidic pH which hinders the spread of pathogenic microorganisms (lactic acid produced maintains the pH of the vagina within the range of: 3.8 – 4.5),
- produce substances resembling bacteriocins (protein compounds that inhibit the growth of bacteria),
- stimulate the immune system,
- compete with pathogenic bacteria for receptors, nutrients or attachment sites on the surface of the epithelial cell,
- inhibit the multiplication of other potentially pathogenic bacteria by producing hydrogen peroxide (H₂O₂).

The increase in estrogen concentration positively affects the acidic pH of the vagina.

Another local mechanism that prevents the development of infection works due to the fact that the mucus covering the vaginal epithelium contains substances that inhibit the development of pathogens (lysozyme, lactoferrin, zinc, fibrinectin, complement proteins).
In response to specific pathogens, the concentration of IgA antibodies increases.

Another problem (non-inflammatory) associated with adolescence is the appearance of bacterial vaginosis episodes, which affect 3-7% of sexually inactive and 4-15% of sexually abused girls aged 13-18. This is important because the microorganisms that build the vaginal flora in the state of bacterial vaginosis are the source of cervical inflammation, endometritis (including often in subclinical forms), and pelvic organ inflammation (PID).

Conflict of interest

The authors declare no competing interests.

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Corresponding Author:

MALGORZATA MIZGIER, PHD

Faculty of Physical Culture in Gorzów Wlkp.
Poznan University of Physical Education
Estkowskiego 13
66-400 Gorzów Wlkp. (Poland)
e-mail: m.mizgier@awf-gorzow.edu.pl