Pediatric lichen sclerosis: what can we learn from typical disease course?

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Case photos are available for health care providers on the request via e-mail: anna.torres@umlub.pl

1. Introduction

Lichen sclerosis (LS) is a chronic autoimmune inflammatory skin condition with predilection for anogenital area [1]. LS presents more frequently in females, with bimodal peak incidence in premenarchal girls and in menopausal women, with mean age of onset at 5.4 years in girls [2]. LS is one of the most common diagnoses in girls with persistent vulvar and anogenital complaints, including itching, pain, dysuria, and constipation. [2,3,4].

The aim of this case report is to present the typical symptoms and course of disease during treatment with ultrapotent topical corticosteroid (UPTC). The uniqueness of the case is the presentation of the 10-month follow-up enriched with photos documenting gradual regression of the disease and its relapse after 6 months of cessation of therapy.

2. Case description

Patient information and primary concerns and symptoms

A 5-year old patient initially presented to PAG Outpatient Clinic of the Lublin University Children's Hospital due to "whitish coating" of the vulva, as it was described by the child's mother, which was first discovered about a year before. In addition, the child also experienced disturbing itching.

The past year, to the initial visit at our Clinic, the child visited three different health care providers. The girl has received antiparasitic medication and was treated with local and oral antifungal medications. The mother did not notice any improvement with the prescribed treatment. About 4 weeks before the visit to our outpatient clinic, the mother started to use feminine hygiene products for child's vulva, which made the itching worse.

Patient's medical, family, and psycho-social history was not contributing.

Clinical findings during initial visit, diagnosis and treatment

On physical examination the patient had Tanner stage 1 breast and pubic hair development. General physical examination and inspection of oral mucosa did not reveal any abnormal findings.

Examination of vulvar area revealed several typical symptoms typical of lichen sclerosis, including: ivory-like looking skin around vagina and anus (figure-of-eight" distribution) with "cigarette paper" appearance, that was mainly confined to labia majora and clearly demarcated from surrounding skin. In addition, a 1 cm subcutaneous hemorrhagic area was present between left labia majora and the clitoral hood, small ecchymoses and telangiectasias were also visible around the anus. Active fissures could be found in typical areas, above clitoris, in interlabial sulci and on the perineal area (Photos 1-2).

The clinical findings and symptoms were typical of lichen sclerosis and the diagnosis was made without further investigations. The mother was educated about the disease and necessity to comply with therapy and follow-up visits.

The patient was prescribed clobetasol propionate 0.05% ointment applied twice daily to affected areas during first four weeks of treatment due to the severity of signs.

The mother obtained written instructions including the amount of ointment to be applied and information regarding possible side effects. She was discouraged to using the intimate hygiene gel and received written information about proper hygiene measures.

Clinical findings during 1st follow-up visit, 4 weeks after initial presentation

The itching had disappeared two weeks after the start of the therapy. The child was complaint with UPTC and no side effects occurred. The proper hygiene measures were followed.

Although, the vulvar signs were still present, some improvement could be seen: less demarcation of affected areas from normal skin, less area with "cigarette paper" appearance, significant decrease in subcutaneous hemorrhagic area adjacent to clitoral hood and disappearance of ecchymoses around the anus.

The UTPC therapy was tapered to once daily for another 4 weeks. The images of the affected areas were presented in the photos 3-4.

Clinical findings during 2nd follow-up visit, 8 weeks after initial presentation

No symptoms were present and no side effects were noted. Proper hygiene measures were maintained. The improvement was clearly visible. Almost all previously visible vulvar skin changes resolved, only one affected area of 1 x 2 cm located above the clitoris still consisted of ivory-white skin with a healing fissure at 12:00 o'clock. The images of the affected areas were presented in the photos 5-6. The therapy was tapered to once every other day for another 4 weeks.

Clinical findings during 3rd follow-up visit, 16 weeks after initial presentation

The child was asymptomatic and the therapy was continued. On physical inspection of the vulva, a slightly pale area measuring about one square centimeter above the clitoris (Photo 7) remained visible. The therapy with UPTC was discontinued. The mother was instructed to use of emollient to protect the vulvar skin daily. The next follow-up was scheduled in 3 months or earlier should the symptoms reoccur.

Clinical findings during 4th follow-up visit – the relapse

The fourth follow-up visit occurred after six months from the previous visit. According to child's mother, she did not think it was necessary to come to scheduled appointment. She came because the itching returned. The proper hygiene measures were continued, however the emollient was not used.

Genital examination revealed a relapse of typical lichen sclerosis signs however the presentation was less severe as compared to the initial visit, with ivory-like looking skin and "cigarette paper" appearance limited to labia majora and perineal areas, and the healing fissure at 12:00 o'clock (Photo 8). The therapy with clobetasol propionate 0.05% ointment (UTPC) applied once daily to affected areas was restarted. On the follow-up in four the weeks the itching was not present and improvement in signs was visible (photo 9), however the once daily treatment was maintained until the follow-up visit scheduled in another 4 weeks.

Ethical issues

Signed written patient's care giver consent was obtained for educational and publishing purposes of all photographs used in the description of the presented case.

3. Summary and Conclusion

Described case presents a classical presentation of lichen sclerosis and typical course of the disease during treatment. In addition, the inclusion of photos taken during follow-up visits makes it easier to understand not only the initial diagnosis but also assessment of treatment effectiveness, and can be of help to healthcare providers, who are not familiar with this disease.

The case also underlies two important issues connected with lichen sclerosis: the delay in diagnosis, the importance of compliance with follow-up and prolonged maintenance therapy. It has been underlined in literature that symptoms and signs of LS could be very diverse, may be nonspecific or discrete. Therefore, the correct diagnosis is often delayed by at least one to two years. The average age of onset varies between 5-7 years in literature and the delay to diagnosis is reported between 1-3 years [3, 5, 6]

In the presented case the signs and symptoms were quite typical of LS, however the diagnosis was delayed by at least one year despite multiple visits to pediatrician, adult gynecology or general practitioner. This underscores the need for education of health care providers about lichen sclerosis in girls and also include the knowledge of presence, presentation and treatment of lichen sclerosis in girls as an important educational goal in undergraduate curriculum.

Lichen sclerosis is a chronic condition and although in previous years it was believed it could often resolve during puberty, recent studies have proven that it often persists throughout reproductive age [7, 8]. This natural course of the disease needs to be clearly explained to caregivers so they understand the importance of compliance with prolonged maintenance therapy [9]. Various regimens of follow-up and treatment were proposed in the past. In the presented case after initial treatment and reduction of signs and symptoms resolution, the

close follow-up with only emollient was introduced. The disease recurrence occurred within the 6 months period. According to literature recurrence rates of prepubertal lichen sclerosis after medical therapy have been reported to range from 44% to 82% [5, 8, 10, 11]. Therefore, it seems prudent to prolong the maintenance therapy in order to prevent recurrences and long-term sequel [12].

References:

1. Smith SD, Fischer G. Paediatric vulval lichen sclerosus. Australas J Dermatol 2009 ;50:243-8.

2. Maronn ML, Esterly NB. Constipation as a feature of anogenital lichen sclerosus in children. Pediatrics 2005;115:e230-2.

3. Powell J, Wojnarowska F. Childhood vulvar lichen sclerosus: an increasingly common problem. J Am Acad Dermatol 2001;44:803-6.

4. Fischer G, Rogers M. Vulvar disease in children: a clinical audit of 130 cases. Pediatr Dermatol 2000;17:1-6.

5. Lagerstedt M, Karvinen K, Joki-Erkkilä M, Huotari-Orava R, Snellman E, Laasanen SL. Childhood lichen sclerosus--a challenge for clinicians. Pediatr Dermatol 2013;30:444-50.

6. Ismail D, Owen CM. Paediatric vulval lichen sclerosus: a retrospective study. Clin Exp Dermatol 2019;44:753-758.

7. Morrel B, van Eersel R, Burger CW, Bramer WM, Ten Kate-Booij MJ, van der Avoort IAM, Pasmans SGMA. The long-term clinical consequences of juvenile vulvar lichen sclerosus: A systematic review. J Am Acad Dermatol 2020;82:469-477.

8. Smith SD, Fischer G. Childhood onset vulvar lichen sclerosus does not resolve at puberty: a prospective case series. Pediatr Dermatol 2009;26:725-9.

9. Ellis E, Fischer G. Prepubertal-Onset Vulvar Lichen Sclerosus: The Importance of Maintenance Therapy in Long-Term Outcomes. Pediatr Dermatol 2015;32:461-7.

10. Cooper SM, Gao XH, Powell JJ, Wojnarowska F. Does treatment of vulvar lichen sclerosus influence its prognosis? Arch Dermatol 2004;140:702-6.

11. Focseneanu MA, Gupta M, Squires KC, Bayliss SJ, Berk D, Merritt DF. The course of lichen sclerosus diagnosed prior to puberty. J Pediatr Adolesc Gynecol 2013;26:153-5.

12. Simms-Cendan J, Hoover K, Mararthe K, Tyler K. NASPAG Clinical Opinion: Diagnosis and Management of Lichen Sclerosis in Pediatric and Adolescent Patients. J Pediatr Adolesc Gynecol 2022;35:112-120.