Childhood lichen sclerosus is a rare but important diagnosis

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ABSTRACT

INTRODUCTION: Lichen sclerosus (LS) is a chronic skin disorder with a predilection for the anogenital area. The disease is mostly seen in prepubertal and postmenopausal females. The lesions present as sharply demarcated white plaques encircling the vagina and anus. The atrophic form can lead to scarring of the affected area.

MATERIAL AND METHODS: Retrospective analysis of hospital records of children (aged 1-18 years) seen at the Department of Dermatology and Allergy Centre in Odense from October 1998 to November 2010 with a definite clinical diagnosis of anogenital LS with/without a confirming biopsy.

RESULTS: A total of 35 girls and one boy were diagnosed with anogenital LS. The diagnostic delay was 17 months. Pruritus, dysuria, bleeding and constipation were the dominant complaints, while one patient was asymptomatic. Referral was made by general practitioners, private dermatologists and paediatricians. Sexual abuse was suspected in five cases. Ten patients underwent biopsy confirming LS. Before a definite diagnosis was given, many children were extensively treated with various topical and oral agents. In our outpatient clinic, 30 children were treated with potent/ultra-potent corticosteroids and five patients were treated with calcineurin inhibitors.

CONCLUSION: General practitioners may overlook this disorder despite characteristic clinical features and effective symptomatic treatment. Diagnostic delay is a significant problem for both patient and family, and the lesions may mimic the findings of sexual abuse. Potent corticosteroids are very effective in symptomatic treatment.

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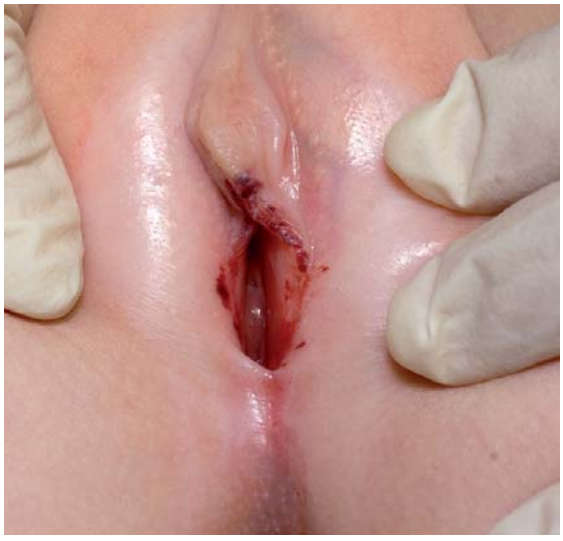
Lichen sclerosus (LS) is a chronic skin disorder of unknown aetiology. The inflammatory dermatosis has a predilection for the anogenital area, and affects the female gender more frequently than males [1]. The disease has two peak ages of presentation – prepubertal and postmenopausal, the latter being the most common.

On examination there is a characteristic white shiny skin plaque in a figure-of-eight distribution around the vagina and anus. Other observations may be atrophy, hyperkeratosis and haemorrhagic areas within the lesions. The atrophic form of LS may result in stenosis of the vaginal introitus, resorption of the labia minora, anal stenosis and phimosis in boys. Most affected children experience bothersome itching and soreness of the vulvar area. Some children have dysuria or constipation as their main complaint, which may be related to painful fissuring in the anogenital area.

The prevalence of vulvar LS has been reported to be one in 900 girls [2], although the condition may be underdiagnosed and underreported. The purpose of this article is to draw attention to LS in children as a highly symptomatic and treatable disorder which may be encountered in various specialties.

MATERIAL AND METHODS

A retrospective analysis was conducted in children aged 1-18 years seen at the Department of Dermatology and Allergy Centre in Odense between October 1998 and November 2010 with the diagnosis of anogenital LS. The inclusion criteria were diagnosis of LS based on typical clinical features with or without a confirming biopsy and exclusion of differential diagnoses. The following data were obtained from the hospital records: referring physician, referral diagnosis, age, gender, diagnostic work-up, symptoms (with focus on itching, dysuria, bleeding and constipation), treatments and follow-up. The child's



Girl with haemorrhagic lichen sclerosus lesions which can raise concerns of possible child abuse.

ORIGINAL ARTICLE

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TABLE 1

Frequency of symptoms in children with anogenital lichen sclerosus.

Symptoms	No. of patients (%)
Itching	31 (86)
Dysuria	23 (64)
Bleeding	22 (61)
Constipation	10 (28)
Enuresis or incontinence	3 (8)
Abdominal pain	2 (6)

age was established at the time the LS diagnosis was made. The delay in diagnosis was recorded as the time between the patient-recorded onset of symptoms and definite diagnosis.

Trial registration: not relevant.

RESULTS

In the study period, 52 children were registered with the diagnosis of LS. Five patients with extragenital LS were excluded as were 11 children who did not meet the inclusion criteria.

A total of 36 children, 35 girls and one boy, with a mean age of 7.5 years (range: 1-18 years) were diagnosed with anogenital LS.

The mean delay in diagnosis was 17 months (0-72 months), and the symptoms are summarized in **Table 1**. Three children aged seven, eight and nine years suffered from enuresis or incontinence. One patient was asymptomatic.

Before referral, the children had all been seen by a minimum of one physician from various specialties. Referral sources and referral diagnoses are listed in **Table 2**.

Ten of the 36 children had the clinical diagnosis of LS confirmed by a skin biopsy.

A total of 16 patients (44%) had been treated with topical antifungals, and 11 patients (31%) had been treated with oral antibiotics before referral. Other topical treatments included antibiotics, antiseptics, tar products, oestrogen and corticosteroids of various potencies.

In our department, 30 children (83%) were treated with potent/ultra-potent topical corticosteroids (potent corticosteroid equals class III and ultra-potent corticosteroid equals class IV), and five patients were treated with calcineurin inhibitors (tacrolimus ointment or pimecrolimus cream). Five patients reported adverse effects such as irritation and burning sensation and one had atrophy of the skin after long-term use of topical corticosteroids.

At the time of the chart review, six patients had experienced complete remission and were discharged after a mean period of 1.5 years. In all, 15 patients were still being treated in the outpatient clinic (mean follow-up period of 1.3 years) to adjust the topical treatment regimen and support the child and family in case of psychosocial issues. Seven patients were being followed by their family practitioner, a practicing dermatologist or a department of paediatrics. Eight patients were lost to follow-up after a mean period of 2.9 years.

DISCUSSION

LS is a benign skin disease mostly affecting the genital skin in prepubertal and postmenopausal females. We found one boy with genital LS out of 36 children, which is in accordance with the literature reporting a skewed gender distribution with a male:female ratio of 1:10 [1]. Hormonal factors possibly play a role in the pathogenesis of LS, but this theory has yet to be proven [3]. LS has been associated with thyroid disease, vitiligo and alopecia areata [2, 4, 5]. We did not screen these children actively for autoimmunity, but found no documentation of known autoimmune diseases in their medical records. The incidence of LS is unknown and treatment is shared between specialties including dermatology, gynaecology, paediatrics, urology and family medicine.

Anogenital LS can present with a variety of different symptoms and signs, but can also be asymptomatic as reported in 7-10% of cases [1, 2]. As a referral is needed for consultation in our outpatient clinic, it is unusual to meet asymptomatic patients here. However, we diagnosed one asymptomatic patient when she was seen with a genital nevus.

The most commonly reported symptoms in genital LS are vulvar itching and soreness [2, 6, 7], and 86% of our patients complained of pruritus. The symptom can worsen around bedtime and nightly genital rubbing can lead to exhausted children during the day. Rubbing or mild trauma tends to tear the fragile skin and cause bleeding. 61% of our patients reported bleeding, which is in accordance with a study by Maronn & Esterly [6], but considerably higher than the 26% reported by

TABLE 2

Referring physician and referral diagnosis.

Referring doctor/department	Number in the group	Lichen sclerosus as correct referral diagnosis, %
General practitioner	11[a]	64
Private dermatologist	4	100
Private paediatrician	4	100
Department of paediatrics	16	100
Venereal clinic	1	0

a) Three of the 11 children had also been examined by a dermatologist, paediatrician or gynaecologist.

Cooper et al [7]. Bleeding can emerge from atrophic and haemorrhagic skin lesions as well as from fissures.

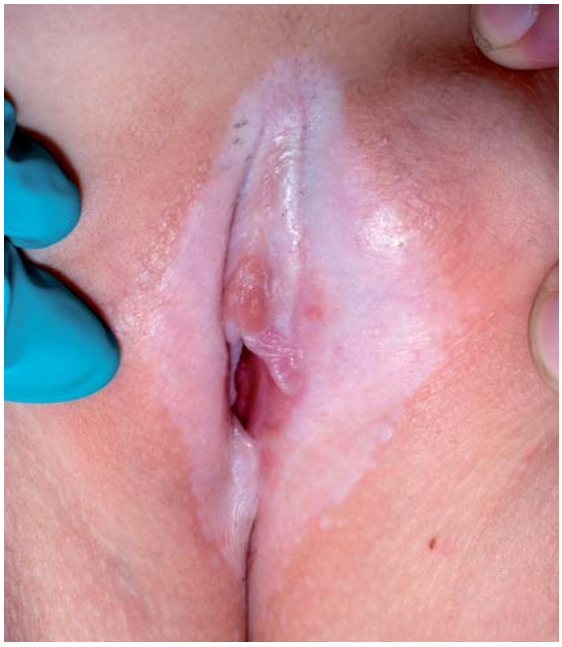
Dysuria is a frequent complaint in childhood LS and was recorded in 64% of our patients. The frequency of this symptom in the literature is 16-50% [2, 6-8]. We noted a correlation of bleeding and dysuria in our patient cohort. Constipation is another well-known complication of anogenital LS [9], and about one fourth of our patients suffered from constipation requiring long-term treatment with laxatives. Periodical stomach pain was reported by two other patients. Published studies have reported gastrointestinal symptoms in 12-67% of patients with genital LS [2, 6-8]; a finding which supports our data. The varying frequencies reported could be related to the parents' detection rate in smaller children and openness of older children, who can be embarrassed to report obstipation. Anal stenosis following perianal LS can cause pencil-like stools and therefore a stool description can suggest LS [10].

A weakness in this retrospective study is that different doctors have evaluated the patients, and it is unknown whether all symptoms have been revealed during the individual disease history.

The clinical history and appearance of the lesions are usually very characteristic. A biopsy is rarely needed in children, unless atypical or neoplastic changes are suspected or if the disease fails to respond to adequate therapy [9]. Nevertheless, ten out of 36 patients in this study had undergone biopsy in diagnosing their skin disease.

Twenty-two children had bleeding, and the symptoms in five children raised concerns of underlying sexual abuse. Sexual abuse should always be considered whenever a child presents with unexplained genital injury or bleeding and therefore, girls with anogenital LS can easily be mistaken as victims of sexual assault [8, 11]. The emotional consequences of misdiagnosis are severe so one should be cautious when suggesting sexual abuse. On the other hand, it is important not to overlook sexual assault. In none of the suspected five children was sexual abuse proved.

There is no cure for LS, so treatment is aimed at alleviating symptoms and preventing anatomical changes like phimosis, narrowing of the vaginal introitus, atrophy of the labia minora and pseudocyst of the clitoris. We found no permanent anatomical changes in our material. Squamous cell carcinoma (SCC) may develop in areas affected with LS in up to 5% of adults [1, 12, 13]. So far, no studies have revealed the incidence of SCC in children with LS. Theoretically, treatment of LS may prevent transformation into malignancy, but, nevertheless, long-term follow up is advisable in affected children as well as adults. In 11 medical records, it was specified that information had been given about cancer risk. The remaining



Girl with classic presentation of vulvar lichen sclerosus: porcelain-white and sharply demarcated plaques.

patients/parents might have been informed, but this was not documented. Some doctors might hold back this information as it may create fear and worries.

In 2010, updated guidelines for managing LS were published by the British Association of Dermatologists [9]. The mainstay of treatment is ultra-potent topical corticosteroids based on smaller case series and observational studies. This treatment is considered to be safe and effective. Calcineurin inhibitors have been reported to have some efficacy in LS, but are not recommended as first-line therapy. In this study, five patients were treated with calcineurin inhibitors as the effect or compliance of corticosteroids was inadequate. Many parents and physicians fear side effects of topical corticosteroids with thinning of the vulnerable skin of anogenital LS. Nevertheless, the scarring evolving from untreated LS lesions seems to be the major problem. In our study, five patients reported adverse effects to topical treatment which is in accordance with other studies [14, 15].

Some children were followed for years, while others only had a few visits. Patients who experienced complete remission were discharged after a mean period of 1.5 years of follow-up, whereas those who were more resistant to treatment or needed psychosocial support were followed for a mean period of 2.9 years. Some children were discharged to their family practitioner, while others were advised to seek a physician in case of refractory symptoms or changing skin signs. The British guidelines suggest follow-up visits three and six months after the initial consultation. Primary care physicians can be consulted once a year if the patient continues to use topical corticosteroid. If patients stop their health care

visits, they should be instructed in self-monitoring and warning signs of vulvar cancer [9].

The clinical course of LS has been unclear and there has been a widely held belief that the disease is likely to remit at menarche/puberty. LS disappearing after puberty, however, is probably restricted to extragenital lesions [1]. Two studies have evaluated the symptoms of girls with genital LS after puberty and both conclude that LS is a remitting and relapsing disease, as patients may experience improvement after puberty, but total remission is uncommon [16, 17].

Failure of the general practitioner to recognize skin signs of LS and associated symptoms like pruritus, dysuria and constipation may lead to a delay in diagnosis and treatment. In our study, the delay of diagnosis was 17 months, although there is some uncertainty about the precise time of disease onset. In the literature, the average time period from onset of symptoms to a diagnosis of LS ranges from eight months to two years [2, 6, 14, 16, 18]. A heightened awareness of childhood LS in primary care will reduce such delay. Vulvar itching and constipation due to LS can be severely distressing, and delay in the diagnosis is therefore a significant problem. Parents often express considerable anger and frustration due to the diagnostic delay when given the correct diagnosis. Also, the delay in treatment can lead to scarring of the affected skin. Many of the patients referred from primary care physicians had been treated with inadequate topical remedies such as antifungals, antibiotics and milder topical corticosteroids. Fungal infection is a common misdiagnosis, and in a study by Maronn & Esterly almost half of the patients with genital LS had been wrongly diagnosed with a yeast infection [6]. Other differential diagnoses of LS include genital eczema, atopic dermatitis, skin infections and morphea. To avoid a delay of diagnosis, referral should be made if treatment with topical agents does not readily improve the patients' symptoms.

In conclusion, anogenital LS in children is a rare diagnosis mostly seen in girls. To avoid diagnostic delay, symptoms of anogenital pruritus, bleeding, dysuria and obstipation should lead to an examination of the anogenital skin and LS should be considered, also in prepubertal children. If children have persistent anogenital complaints or skin changes despite medical treatment, they should be referred to a dermatologist. Delay in diagnosis and treatment can lead to severe scarring of the anogenital area. Potent corticosteroids are very effective in treatment of this disease.

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CONFLICTS OF INTEREST: none

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