



Case Report

Lichen Sclerosus et Atrophicus

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Abstract

This report presents two cases of Lichen Sclerosus et atrophicus. Patients were prepubertal females having itchy white plaques in the vulvar region with on and off burning micturition. No extragenital involvement was seen. Diagnosis was confirmed by histopathology. They were treated with local application of corticosteroid for a variable time period. Both the cases resolved by the age of menarche. These cases are being reported here in the context of its rare occurrence in children and to the best of our knowledge has not yet been reported in children in Bangladesh. The etiopathogenesis, diagnosis and treatment modalities of the disease are reviewed from the literature.

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Introduction

Lichen sclerosus et atrophicus (LSA), first described in 1889 by Hallopeau,¹ is an infrequent, chronic atrophic disease most commonly affecting the vulva of postmenopausal white women,^{2,3} although it may appear in any age, in all races and both sexes⁴ When it occurs in children, it affects mainly girls.⁴ The disease manifests as white, flat papules with an erythematous halo and black, hard follicular plugs. In advanced cases, the papules tend to coalesce into large, white patches of thin, shiny and itchy skin. Grooves, cracks and ecchymoses may appear due to atrophy and shrinkage of the skin.⁵

The condition is progressive and the atrophy does not regress. Lesions are most common in the vulvar and perianal areas but may be seen anywhere with the most common extragenital sites

being on the trunk and limbs. The characteristic lesions are hypopigmented plaques in a figure of - 8 pattern surrounding the vulva and anus and often involving the natal cleft.⁶ The affected hypopigmented skin is sharply demarcated from the normal skin. The incidence of the disease is more in females than in males. LSA in females has two peak ages of presentation. The first of these occurs in prepubertal girls⁷ and may resolve or continue beyond the menarche.⁸ The other peak of incidence is in postmenopausal women;⁷ although this suggests a hormonal influence, hormone replacement therapy neither improves existing disease nor provides any protection against the development.⁹ The incidence of LSA in male is lower than in female but there is also a bimodal onset, with peaks of disease presentation occurring in young boys and then again in adults.¹⁰

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LSA in Childhood

Incidence of LSA in childhood is much less than that in adulthood. It generally occurs prior to age 6.¹¹ It is very similar to the disease seen in adults, as it is more common in the anogenital region but there are often extragenital involvements as well. The lesions are similar to those in adult women but ecchymosis may be very striking and potentially mistaken as evidence of sexual abuse.¹² Among the children also it is much more common in girls and may present with the typical symptoms of itching and occasional vaginal discharge which lead to excoriation, bleeding and ulceration. More than half have these symptoms and occasional constipation is a feature as well.

Pathogenesis

The etiology of LSA is uncertain but there is mounting evidence to suggest that autoimmune mechanisms are involved in its pathogenesis;¹³ there is an increased incidence of tissue-specific antibodies¹⁴ and associations with other autoimmune diseases in patients with LSA,¹⁵ as well as positive associations with HLA class II antigens.¹⁶ Approximately 21% of patients have an autoimmune disease, most commonly a thyroid disorder.⁹ Twenty-two percent of patients have a family history, and 44% have one or more autoantibodies. Patients with a genetic predisposition or with associated autoimmune disorders do not differ clinically from those without such a background.¹⁷ The role of dehydrotestosterone is often considered as a cause in adult and postmenopausal women. Role of infection in the genesis of LSA has also been looked at recently. Cell wall deficient type of mycobacteria are suggested as one of causative agents according to some authors.¹⁸

Diagnosis

The diagnosis of LSA in most patients is usually made clinically, but a confirmatory biopsy is helpful in cases where there is some clinical doubt about the diagnosis and to document any atypical features. The classical histological features of uncomplicated LSA include a thinned epidermis

with hyperkeratosis, a wide band of homogenized collagen below the dermoepidermal junction and a lymphocytic infiltrate beneath the homogenized area. There may be small focal areas where the inflammatory infiltrate is close to the dermoepidermal junction, similar to Lichen Planus.¹⁹ The length of time that LSA has been present cannot be determined accurately using histological parameters.^{19,5}

The main differential diagnoses include Lichen Planus, mucous membrane pemphigoid and genital psoriasis. A skin biopsy is not always practical in children and it is preferable to initiate their treatment without histological confirmation. A biopsy is essential in all cases that fail to respond to adequate treatment.⁹

Lichen sclerosus et atrophicus mimics sexual abuse and has led to false accusation and investigations.⁶ Suspicious features include LS in older prepubertal girls, the presence of associated infection (especially infections that are characteristically sexually transmitted), or other symptoms or signs of abuse.

Management

Patients with LSA typically present with thin, parchment like skin, which is a poor barrier to the loss of moisture. Patients should avoid excessive drying of this skin after bathing. Bland emollients should be used to improve moisture retention.

As definitive therapy the recommended and accepted treatment is an ultrapotent topical corticosteroid ointment. Clobetasol propionate 0.05% ointment may be applied once daily¹⁷ The regimen recommended by the authors is clobetasol propionate initially once a night for 4 weeks, then on alternate nights for 4 weeks and, for the final third month, twice weekly. A 30-g tube of clobetasol propionate should last 12 weeks and the patient is then reviewed. If the treatment has been successful the hyperkeratosis, ecchymoses, fissuring and erosions should have resolved but the atrophy and colour change will remain. The clobetasol propionate is then continued and used as and when required. Most patients seem to require 30-60 g annually.

Topical oestrogen is reported to be beneficial in girls, improving the histological features and itch.²⁰ However, the magnitude of benefit is uncertain as this report stated that the overall clinical improvement was 20%, and no comparative trials are available. Promising results have been reported from treatment with oral retinoids in patients with severe LSA of the vulva.²¹

Difficult cases refractory to the usual therapies require consultation with a dermatologist and, on occasion, a plastic surgeon. Multidisciplinary management is helpful in such patients. Surgery, laser, photodynamic therapy and cryotherapy may be tried.^{9,17}

Case Reports

Case I

A 8 year old female hailing from Rajshahi city presented with itchy, ivory white plaques in the vulvar region (Fig-I) with on and off burning micturition and curdy white vaginal discharge since one year. No extragenital site was found to be involved. She was diagnosed clinically as Lichen sclerosus et atrophicus of vulva with superadded candidal infection. Her diagnosis was confirmed by biopsy. Microscopic examination of vaginal discharge showed candidal infection. On light microscopy the histological sections of skin revealed hyperkeratosis and atrophy of the epidermis with hyalinized superficial dermis and underlying lymphocytic infiltration. On routine examination patient was found to be anaemic which was hypochromic microcytic in nature. Her ESR was 50 mm in first hour of observation and urine culture was negative for bacterial growth. Ultrasonography of abdomen was unremarkable. She was treated with oral antihistaminics and local combination of 1 percent clotrimazole with 0.1% betamethasone, "one finger-tip unit" (about 0.5 gm) twice daily and was followed up for four weeks. Her symptoms were reduced and then the dose was given once a day for another four weeks and thereafter every alternate day for another two months. Her clinical appearance did not reverse, even though the symptoms were relieved. She was

advised to use and discontinue betamethasone as and when required. She was followed up for another six months and was found to be apparently free from the symptoms.



Fig. 1: Ivory white plaques in vulval region.

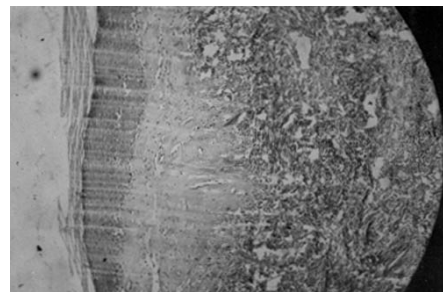


Fig. 2: Histological feature of LSA.

Case II

A 10 year female hailing from Rajshahi city attended a private clinic of the city with complaints of vulvar discolouration with intense itching and burning micturition. Her mother stated that she developed a whitish lesion in vulvar area at the age of 6 which gradually spread away affecting the whole genitalia. She often used to scratch it. She was treated by local doctors with oral and topical antibiotics. Mother also stated that the baby is a constipator and have to take laxatives for bowel movement. On local examination, her whole vulvar area was whitish in colour with sharp demarcation with the surroundings. The lesion has also surrounded the anus to make a figure of 8 pattern. The whole area was covered with fowl-smelling serosanguinous discharge. There was no extragenital involvement. The case was diagnosed clinically as Lichen sclerosus et

atrophicus and was confirmed histologically (Fig-II). A lichenoid infiltrate in the dermal-epidermal junction, compact hyper-keratosis with stratum corneum and remarkable edema in the papillary (upper) dermis were noted. Routine examination revealed mild anaemia with a high ESR (60 mm. in first hour) and urine culture showed growth of *Esch. coli* with a colony count $>10^5$ /ml. An antibiogram was performed. Ultrasonography of abdomen showed unremarkable features. She was first treated with an antihistaminic and an appropriate antibiotic for two weeks. The urine culture was repeated and was found to be free from bacteria. She was then prescribed 0.1% betamethasone ointment twice daily for four weeks. No symptomatic relief was observed. Betamethasone was then replaced by 0.5% Clobetasol propionate ointment once a day for four weeks. Antihistaminic was also continued. On further follow up no improvement was noticed. Treatment was continued for some more weeks with very negligible relief of her symptoms. Patient discontinued treatment. The disease however did not progress into any further complication and has resolved spontaneously within next two years.

Discussion

Lichen sclerosus (LS) is a chronic inflammatory dermatosis that results in white plaques with epidermal atrophy. It is a multifactorial disease commonly seen among aged female. Genetic aspect seem to play an important role as the LSA are reported in successive generations usually mother and daughter.² It has also been seen in sisters, mother and son.²² Our patient did not report such family history. There was no associations of autoimmune disease with these girls., although autoimmunity is often implicated as a common cause of LSA. Extragenital involvement is often reported in childhood LSA, though our cases did not develop any such lesion except the vulva.

Although most common in postmenopausal women, this disease appears in children, most frequently in girls.²³ No single factor can be implicated in the development of LSA in children. Manifestations are similar to those seen in adult women⁴ but with a better prognosis. It occurs in the anogenital area of girls, and less frequently in boys. About two-thirds of cases improve or undergo involution before or around the menarche.²³ Those that do not spontaneously resolve may go onto have trouble with atrophy of the labia minora and clitoris. Some requires surgical intervention to correct gross atrophy of the introitus. Our first patient was suffering for three years, her lesion was gradually spreading to occupy the whole vulval area. She responded well to topical steroid therapy. Our second patient was suffering from four years. Her vulval area had undergone marked atrophic changes but no vulvar stenosis was seen. She did not show a good response to steroid treatment. Her condition however improved slowly even after withdrawal of therapy.

Vulvar LSA may progress to gradual obliteration of the labia minora and stenosis of the introitus. It is widely reported that prepubertal LSA in girls may resolve spontaneously although some of these patients may go on to suffer from various types of vulvodynia in adulthood. Our first case responded well to conventional steroid treatment. Our second patient did not respond to steroid but even after discontinuation of treatment her lesion did not progress into any complication either. She rather recovered spontaneously. Both the patients became free from the disease by the age of menarche.

LS has no associated increased mortality unless the patient develops a malignancy in the area. The likelihood of development of Squamous cell carcinoma is however, more in older women.¹⁷ The relationship of LSA to the development of malignancy varies greatly in different studies. An increased risk of squamous cell carcinoma may exist in vulvar disease, but the precise increase in

risk and what cofactors (HPV or prior radiotherapy) may be involved are not yet defined. In a review of 1356 patients with LSA, the coincidence with cancer was found to be 4.1%.²³ Lichen sclerosis et atrophicus was detected in up to 53% of patients with invasive vulvar carcinoma.²³ Such an association was not reported in our cases. The risk of malignancy in uncomplicated genital LS that has been diagnosed and treated appropriately is very small.²⁴ Though the lesions had resolved in both of our cases we had kept them under a long term follow-up to look for recurrence or further complication.

References

- Hallopeau H. Lichen plan sclereux. *Ann Dermatol Syphiligr* 1889; 10:447-449.
- Shirer JA, Ray MC. Familial occurrence of lichen sclerosis et atrophicus. *Arch Dermatol* 1987; 123: 485-488.
- Friedrich EG, Karla PS. Serum levels of sex hormones in vulvar lichen sclerosis and the effect of topical testosterone. *N Engl J Med* 1984; 310:488-491.
- Wallace JH. Lichen sclerosis et atrophicus. *Trans St Johns Hosp Dermatol Soc* 1971; 57:9-30.
- Marren P, Millard PR, Wojnarowska F. Vulval lichen sclerosis: lack of correlation between duration of clinical symptoms and histological appearances. *J Eur Acad Dermatol Venereol* 1997; 8: 212-16.
- Loening-Baucke V. Lichen sclerosis et atrophicus in children. *Am J Dis Child*.1991Sep; 145(9):1058-61.
- Wallace HJ. Lichen sclerosis et atrophicus. *Trans St John's Dermatol Soc* 1971; 57: 9-30.
- Ridley CM. Genital lichen sclerosis (lichen sclerosis et atrophicus) in childhood and adolescence. *J R Soc Med* 1993; 86: 69-75. 18
- Meffert JJ, Davis BM, Grimwood RE. Lichen sclerosis. *J Am Acad Dermatol* 1995; 32: 393-412.
- Lipscombe TK, Wayte J, Wojnarowska F et al. A study of clinical and etiological factors and possible associations of lichen sclerosis in males. *Australian J Dermatol* 1997; 38: 132-6.
- Robert DL, Tremaine MD, Robert AW, Miller MD. Lichen sclerosis et atrophicus. *Int J Dermatol* 1989; 28(1):10-15.
- Handfield Jones SE, Hinde FR, Kennedy CTC. Lichen sclerosis et atrophicus in children misdiagnosed as sexual abuse. *Br Med J* 1987; 294: 1404-5.
- Dickie RJ, Horne CHW, Sutherland HW. Direct evidence of localized immunologic damage in lichen sclerosis et atrophicus. *J Clin Pathol* 1982; 35: 1395-9.
- Goolamali SK, Barnes EW, Irvine WJ, Shuster S. Organ specific antibodies in patients with lichen sclerosis. *Br Med J* 1974; iv: 78-9.
- Harrington CI, Dunsmore IR. An investigation into the incidence of autoimmune disorders in patients with lichen sclerosis et atrophicus. *Br J Dermatol* 1981; 104: 563-6.
- Marren P, Charnock FM, Bunce M et al. The associations between lichen sclerosis and antigens of the HLA system. *Br J Dermatol* 1995; 132: 197-203.
- Clark JS, Muller SA. Lichen sclerosis et atrophicus in children. A report of 24 cases. *Arch Dermatol* 1967; 95:474-481.
- Cantwell AR. Histologic observations of pleomorphic variably acid fast bacteria in scleroderma, morphea and lichen sclerosis et atrophicus. *Int J Dermatol* 1984; 23:45-52.
- Hewitt J. Histologic criteria for lichen sclerosis of the vulva. *J Reprod Med* 1986; 31: 781-7.
- Lascano EF, Montes LF, Mazzini MA. Tissue changes in lichen sclerosis in children following local application of oestrogens. *Br J Dermatol* 1964; 76: 496-7.
- Virgili A, Corazza M, Bianchi A et al. Open study of topical 0.025% tretinoin in the treatment of vulval lichen sclerosis. One year of therapy. *J Reprod Med* 1995; 40: 614-18.
- Murphy FR, Lipa M, Haberman HF. Familial vulvar dystrophy of lichen sclerosis type. *Arch Dermatol* 1982; 118:329-331.
- Tremaine RDL, Miller RAW. Lichen sclerosis et atrophicus. *Int J Dermatol* 1989; 28:10-16.
- Brenner, Sarah, and Yael Politi. Dermatologic Diseases and Problems of Women Throughout the Life Cycle. *Int J Dermatol* June 1995; 34 (6): 369-379.

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