

Acute Lichen Sclerosus in a 25 Years Young Female- A Case Report

NAHID YASMIN¹, NAZNEEN KABIR², TAHMINA AFRIN DAISE³, SHAHRIN AHMED⁴, FARHANA SHARMIN⁵

Abstract:

Lichen sclerosus (LS) is a disease of unknown cause that results in white patches on the skin, which may cause scarring on and around genital skin¹. Several risk factors have been proposed, including autoimmune diseases, infections and genetic predisposition^{2,3}. There is evidence that LS can be associated with thyroid disease⁴. Women are more commonly affected than men (10 to 1 ratio), particularly prepubertal girls and after menopause. The condition most commonly occurs on the vulva and around the anus with ivory-white elevations that may be flat and glistening. There may be marked itching or the condition may be without any symptoms. There may also be thinning and shrinkage of the genital area. This condition is presented here as an younger woman presented with acute form of lichen sclerosus.

Introduction:

Lichen sclerosus is the commonest non neoplastic epithelial vulval disorder^{5,6}. It is most often seen in post menopausal women but can occur in young women^{7,8,9}. It appears as white, glistening sheets with clearly defined margins and involves the labia, the perineum and the perianal region^{10,11,12}. There may also be thinning and shrinkage of the genital area that may make coitus, urination, and defecation painful. Lichen Sclerosus is not contagious; it cannot be caught from another person¹³. Although it is not clear what causes LS, several theories have been postulated. Theories are genetic, autoimmunity, infection, hormones and local skin changes. Lichen sclerosus may have a genetic component. Higher rates of lichen sclerosus has been reported among twins^{14,15} and families¹⁶. Autoimmunity is a process in which the body fails to recognize itself and therefore attacks its own cells and tissue. Specific antibodies have been found in LS. Furthermore, there seems to be a higher prevalence of other autoimmune diseases such as diabetes mellitus type 1, vitiligo and thyroid disease¹⁷. Both bacterial as well as viral pathogens have been implicated in the etiology of LS. A disease that is similar to LS, acrodermatitis chronica atrophicans is caused by the spirochete *Borrelia burgdorferi*¹⁸ evidenced by investigation. Viral

involvement of HPV¹⁹ and hepatitis C²⁰ are also suspected. Since LS in females is primarily found in women with a low estrogen state (prepubertal and postmenopausal women), hormonal influences were postulated. To date though, very little evidence has been found to support this theory. Some findings suggest that LS can be initiated through scarring²¹ or radiation,^{22,23} although these findings were sporadic and very uncommon. A biopsy of the affected skin is often done to confirm diagnosis.

There is no definitive cure for LS²⁴. Behavior change, such as good hygiene and minimizing scratching of the affected area, is an important part of treatment²⁵. LS is also usually treated with potent topical steroids, like clobetasol propionate or mometasone furoate²⁴. These can relieve symptoms and prevent scarring²⁶. However, LS is a chronic disease so topical steroids may need to be continued as maintenance therapy²⁷.

Case report:

A 25 yrs old multiparous lady from low socioeconomic family was admitted in Institute of child and mother Health on first week of october 2013 with the complaints of itching of the vulva and gradually whitening of vulval skin for last three months. She also complained of cracking in the lesion after intercourse which become painful but healed spontaneously.

-
1. Associate Professor, GYNAE, ICMH, Matuail, Dhaka.
 2. Head of Dept. of GYNAE, ICMH, Matuail, Dhaka.
 3. Registrar, GYNAE, ICMH, Matuail, Dhaka.
 4. Internee Doctor, Bangladesh Medical College.
 5. DGO student, ICMH, Matuail, Dhaka.

She has no history of diabetes mellitus, eczema or any other medical, surgical or skin diseases. She had history of taking ciprofloxacin, metronidazole and topical antifungal ointment 15 days back.

She has regular cycle of average flow and duration. On admission, patient was healthy with average built, wt-52kg, mildly anaemic and normotensive. No abnormality was detected on systemic examination.

Local examination of vulva revealed multiple whitish area of variable in size and shape in upper part of inner aspect of left side of labia majora (1.5cm x 1.25cm), middle part of inner aspect of right side of labia majora (3cm x 2cm), lower part of labia majora covering the perimum (2cm x 1.5cm). Vagina, cervix and uterus were healthy.



Figure: *Lichen Sclerosus of Vulva*

All investigation reports were within normal limit, blood group is 'A' positive. Multiple wedge biopsies were taken and histopathological examination report was Lichen sclerosus of vulva.

General management was -to avoid the use of local cosmetics, to use non irritant soap and dry carefully without rubbing, to use cotton underwear or nothing at all. Topical use of clobetasol propionate 0.05% was advised to apply at night daily for 1 month followed by alternate night for the 2nd month and then after twice weekly for 1 month along with oral antihistamine as specific management.

Patient was discharged four days later with advice to come for follow up at 15 days interval for 3 months.

Discussion:

Lichen sclerosus et atrophicus was first described in 1887 by Dr. Hallopeau²⁸. Since not all cases of lichen sclerosus exhibit atrophic tissue, *et atrophicus* was dropped in 1976 by the International Society for the Study of Vulvovaginal Disease (ISSVD), officially proclaiming the name *lichen sclerosus*²⁹. Lichen sclerosus was most commonly observed in postmenopausal women (18, 69.2%), followed by women in reproductive age group (5, 19.23%), and prepubertal girls (3, 11.5%). All patients presented with ivory white atrophic plaque similar to this patient. Warmth and moisture, chronic mechanical irritation, deficiency of iron, vit A, folic acid, vit B₁₂, riboflavin and other essential factors are responsible for these changes^{30,31}. Female genital lesions may be confined to the labia majora but usually involve and eventually obliterate the labia minora and stenose the introitus. Often, an hourglass, butterfly, or figure-8 pattern involves the perivaginal and perianal areas, with minimal involvement of the perineum in between. Only labia majora was involved in this case. Presentation of lichen sclerosus may be acute-manifested by erythema and edema of vulval skin, lichenification, hyperkeratosis, erosion/ulceration and subepithelial haemorrhages.

Presentation of chronic lichen sclerosus are- wrinkled, white skin appearance, agglutination of labia minora and clitoris, introital stenosis, involvement of the perianal region. This patient had similarity with acute presentation. Clobetasol propionate was prescribed for three months and asked for follow up. In case of poor response long-term antibiotic will be followed. Another small study has shown long-term antibiotic treatment to be effective in patients who had poor response to steroids³².

Lichen sclerosus usually does not cause skin cancer. However, skin that is scarred by lichen sclerosus is more likely to develop skin cancer. The high rate of squamous cell carcinoma in women with lichen sclerosus in 3-5% cancer develop mainly in women who continue to suffer from vulval itching or neglect treatment³³.

Conclusion:

Lichen sclerosus is one of the premalignant conditions of the vulva specially in presence of scarring. This patient received effective treatments within three months of the appearance of the disease there is very

little chance of developing malignancy but follow up every six to 12 months is necessary.

Reference:

- Pugliese JM, Morey AF, Peterson AC. "Lichen Sclerosus: Review of the Literature and Current Recommendations for Management". *J Urol* 2007; 178 (6): 2268–2276.
- Yesudian PD, Sugunendran H, Bates CM, O'Mahony C. "Lichen sclerosus". *Int J STD AID* 2005; 16 (7): 465–473.
- Regauer S. "Immune dysregulation in lichen sclerosus". *Eur J Cell Biol* 2005; 84 (2–3): 273–277.
- Birenbaum, DL; Young, RC (). "High prevalence of thyroid disease in patients with lichen sclerosus". *J Reprod Med* 2007; 52 (1): 28–30.
- Funaro D. Lichen sclerosus: A review and practical approach. *Dermatol Ther* 2004;17:28-37.
- Burns T, Breathnach S, Cox N, Griffiths C, editors. The genital, perianal and umbilical regions. Rook's textbook of dermatology. 7th ed. Blackwell Science: Oxford;2004. p.68.1-68.104.
- Neill SM, Tatnall FM, Cox NH: British Association of Dermatologists. Guidelines for the management of lichen sclerosus. *Br J Dermatol* 2002;147:640-9.
- Meffert JJ, Davis BM, Grimwood RE. Lichen sclerosus. *J Am Acad Dermatol* 1995;32:393-416.
- Wallace HJ. Lichen sclerosus et atrophicus. *Trans St Johns Hosp Dermatol Soc* 1971 ;57 :9-30.
- Powell J, Wojnarowska F, Childhood vulvar lichen sclerosus: An increasingly common problem. *J Am Acad Dermatol* 2001;44:803-6.
- Meyrick Thomas RH, Ridley CM, McGibbon DH, Black MM. Lichen Sclerosus et atrophicus and autoimmunity: A study of 350 women. *Br J Dermatol* 1988;118:41-6.
- Tasker GL, Wojnarowska F. Lichen sclerosus. *Clin Exp Dermatol* 2003;28:128-33.
- Thomas M, Kennedy CT. "The development of lichen sclerosus et atrophicus in monozygotic twin girls.". *The British journal of dermatology* 1986; 114 (3): 377–9.
- Cox NH, Mitchell JN, Morley WN. "Lichen sclerosus et atrophicus in non-identical female twins.". *The British journal of dermatology* 1986; 115 (6): 743.
- Sherman V, McPherson T, Baldo M, Salim A, Gao XH, Wojnarowska F). "The high rate of familial lichen sclerosus suggests a genetic contribution: an observational cohort study.". *Journal of the European Academy of Dermatology and Venereology : JEADV* 2010; 24 (9): 1031–4..
- Thomas M, Ridley CM, McGibbon DH, Black MM ("Lichen sclerosus et atrophicus and autoimmunity—a study of 350 women". *Br J Dermatol* 1988; 188 (1): 41–46. .
- Eisendle K, Grabner TG, Kutzner H). "Possible Role of *Borrelia burgdorferi* Senu Lato Infection in Lichen Sclerosus". *Br J Dermatol* 2008; 144 (5): 591–598.
- Drut RM, Gomez MA, Drut R, Lojo MM. "Human papillomavirus is present in some cases of childhood penile lichen sclerosus: an in situ hybridization and SP-PCR study". *Pediatr Dermatol* 1998; 15 (2): 85–90.
- Yashar S, Han KF, Haley JC). "Lichen sclerosus-lichen planus overlap in a patient with hepatitis C virus infection". *Br J Dermatol* 2004;150 (1): 168–169.
- Pass CJ). "An unusual variant of lichen sclerosus et atrophicus: delayed appearance in a surgical scar". *Cutis* 1984; 33 (4): 405.
- Yates, VM; King, CM; Dave, VK (1985). "Lichen sclerosus et atrophicus following radiation therapy". *Arch Dermatol* 121 (8): 1044–1047.
- Chi, CC; Kirtschig, G; Baldo, M; Lewis, F; Wang, SH; Wojnarowska, F. "Systematic review and meta-analysis of randomized controlled trials on topical interventions for genital lichen sclerosus.". *Journal of the American Academy of Dermatology* 2012; 67 (2): 305–12.
- "ACOG Practice Bulletin No. 93: diagnosis and management of vulvar skin disorders.". *Obstet Gynecol* 111 (5): 1243–53. May 2008.

25. Goolamali, SK; Goolamali, SI (1997). "Lichen sclerosus". *Journal of obstetrics* Tasker GL, Wojnarowska F (2003) Lichen sclerosus. *Clin Exp Dermatol* 28:128-133.
26. Casabona, F; Priano, V; Vallerino, V; Cogliandro, A; Lavagnino, G). "New surgical approach to lichen sclerosus of the vulva: The role of adipose-derived mesenchymal cells and platelet-rich plasma in tissue regeneration". *Plastic and reconstructive surgery* 2010; 126 (4): 210e–211e.
27. Li, Y; Xiao, Y; Wang, H; Li, H; Luo, X (Aug 2013). "Low-concentration topical tacrolimus for the treatment of anogenital lichen sclerosus in childhood: maintenance treatment to reduce recurrence.". *Journal of pediatric and adolescent gynecology* 26 (4): 239–42.
28. Hallopeau, H (1887). "Du lichen plan et particulièrement de sa forme atrophique: lichen plan scléreux". *Ann Dermatol Syphiligr (Paris)* (8): 790–791.
29. Friedrich Jr., EG. "Lichen sclerosus". *J Reprod Med* 1976; 17 (3): 147–154.
30. Marren P, Millard P, chia Y et al. () Mucosal lichen sclerosus/lichen planus overlap syndromes. *Br J Dermatol* 1994; 131;118-123
31. Power JJ, Wojnarowska F. Lichen sclerosus. *Lancet* 1999; 353;1777-1783.
32. Shelley, W. B.; Shelley, E. D.; Amurao, C. V. "Treatment of lichen sclerosus with antibiotics". *International Journal of Dermatology* 2006; 45 (9): 1104–1106.
33. "Fast Facts About Lichen Sclerosus". *Lichen Sclerosus*. National Institute of Arthritis and Musculoskeletal and Skin Diseases, Retrieved June 2012.