

Dermoscopic Evaluation of Extragenital Lichen Sclerosus et Atrophicus

Sunanda Arun Mahajan¹, Jayati Shailesh Dave¹

¹ Department of Dermatology, Seth G.S. Medical College and K.E.M Hospital, Parel, Mumbai

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Corresponding author: Jayati Shailesh Dave, MD, Department of Dermatology, OPD 117, Seth G.S. Medical College and K.E.M Hospital, Parel, Mumbai, 400012 Telephone number: 9987054564. E-mail address: jayatidave20@gmail.com

ABSTRACT **Introduction:** Lichen sclerosus (LS), is an uncommon inflammatory dermatosis with preferential involvement of anogenital region. Diagnosis of LS is mainly clinical, but clinical differentiation from conditions like vitiligo, morphea may be a difficult task at times that often requires histological analysis. Dermoscopy is one such non-invasive tool which can help diagnose the disease. There is paucity of Indian data on dermoscopy of LS.

Objectives: To evaluate clinical, dermatoscopic patterns of LS and correlate them with histopathology.

Methods: The study was conducted in a tertiary hospital after obtaining consent from 20 patients. OITEZ e-scope digital microscope was used to evaluate the lesions. Both polarized and nonpolarized mode were used and skin biopsy was done to confirm diagnosis.

Results: Based on morphology, LS was classified as scleroatrophic lesions (61.5%), guttate lesions (30.8%) and hyperkeratotic lesions (7.7%). Dermoscopic analysis revealed structureless white to yellow areas as most common finding (100%) followed by chrysalis like structure (80.8%). Linear irregular vessels were seen in 61.5% lesions and perifollicular scaling in 50.0% lesions. Keratotic plugs were seen in 50.0% lesions. A new characteristic finding, “rosettes” was seen in 38.5% lesions has never been reported with LS before. Non polarized mode was particularly useful for identifying texture changes, keratotic plugs and minute scales which were not visible otherwise.

Conclusions: Dermoscopy is a simple diagnostic tool that helps in the early diagnosis of LS with specific pattern which can avoid invasive procedure like biopsy. Both non-polarised and polarized dermoscopy must be done to visualize the changes of LS well.

Introduction

Lichen sclerosus (LS), is an uncommon inflammatory dermatosis with preferential involvement of anogenital region. Isolated extragenital LS is seen in 6% of cases, and association with genital LS seen in 15-20% cases [1]. Women are affected more commonly than men [2]. Extragenital LS - found to be relatively common on the neck and shoulder - is generally asymptomatic, but can occasionally be pruritic [2]. Extragenital LS presents as white polygonal porcelain atrophic papules that coalesce to form plaque studded with comedo-like plugs or evenly spaced dells which corresponds to appendageal ostia [2].

Diagnosis of LS is mainly clinical, and sometimes clinical differentiation from other hypopigmentary conditions such as vitiligo and scleroatrophic disorder like morphea may be a difficult task that often requires histological analysis. Dermoscopy is one such non-invasive tool which can help develop a pattern to diagnose the disease.

Objectives

To study dermoscopic patterns of extragenital LS in series of patients with brown skin and correlate these patterns with histopathology.

Methods

Patients

This was an observational case series study conducted in the Department of Dermatology of a tertiary hospital, from July 2018 to January 2020. A total of 20 patients with clinical features of LS were enrolled in the study. A complete history and dermatological examination were performed after obtaining written informed consent from the patient. Demographic data, such as age, gender, and clinical variables in terms of site of lesions and disease duration were documented. Lesions with less than 2 years and more than 2 years duration were arbitrarily termed as early and late lesions. A first biopsy was taken from clinically active lesion and a second biopsy was taken if patient had genital LS or different morphology of LS.

Dermoscopic examination

OITEZ e-scope digital microscope [DP-M17 filter e-scope pro (optical 200x)] with 20x and 200x magnification was used for dermoscopy and an alcohol-based sanitizer was used as immersion fluid. Both polarized and non-polarized modes were employed in the study. The dermoscopic images were saved and the findings noted on an excel sheet after evaluation by two individual independent authors. Parameters such as vessels, scales, follicular findings, background color, morphology and specific findings if any were noted.

Inclusion criteria

Untreated patients of extragenital LS willing to give consent with histopathology suggestive of the disease were included in the study.

Exclusion criteria

Patients already on treatment for the disease, or not willing for biopsy were excluded from the disease.

Statistical analysis

Statistical analysis was performed using descriptive tools such as percentage and frequency. Results were statistically described as types of dermoscopic patterns.

Results

There were 20 patients (12 females, 8 males) and 26 lesions included in the study. The patients with extragenital LS with mean duration of disease of 20.4 months (minimum 2 months and maximum 6 years) and mean age of 37.8 years (youngest 12 years and oldest 71 years) presented with hypopigmented lesions at different sites. The lesions were asymptomatic in 15 patients and pruritic in 5 patients. On examination the maximum number of lesions were seen on the trunk (101, 38.5%) followed by lower extremities (6, 23.1%), upper extremities (9, 34.2%) and the least on face (1, 3.8%). Koebner phenomenon was seen in 4 patients. Patients presented with classical ivory colored small polygonal papules coalescing to form plaques or hypopigmented atrophic plaques. Four patients also had multiple guttate macules of LS. LS of the genitalia was seen in 3 patients and 2 patients had biopsy-proven morphea in addition. Lesions were classified into 3 groups based on morphology namely scleroatrophic plaque (16) (Figure 1A), guttate (8) (Figure 1B) and hyperkeratotic having predominantly keratotic plugs with underlying sclerosis (2) (Figure 1C).

All lesions (Table 1) of extragenital LS revealed patchy structureless white to yellow areas (WSA) (Figure 2A) which were most commonly associated with irregularly arranged linear vessels (16, 61.5%), linear branching vessels (10, 38.5%) or dotted vessels (4, 15.4%) and glomerular vessels (4, 15.4%). The linear vessels were of varying calibers, whereas dotted vessels were arranged in a random fashion. Erythematous areas were seen in 10 (38.5%) lesions which were accompanied by vessels. Chrysalis like structure which is shiny white streaks was seen in 21 (80.8%) patients (Figure 2B). Comedone-like opening (CLO) defined as ovoid or round craters filled with brownish-black material were seen in 9 (34.6%) lesions whereas keratotic plugs defined as invaginations filled by yellowish material were seen in 13 (50.0%) sites (Figure 3, A and B). Peppering blue grey dots was seen in 9 (34.6%) lesions (Figure 3, C and D).



Figure 1. (A) Two shiny porcelain white colored atrophic plaques on axilla typical of scleroatrophic plaque of Lichen sclerosus (LS). (B) Multiple discrete hypopigmented rain drop like slightly atrophic macules typical of guttate lesions of LS. (C) Multiple hypopigmented sclerotic guttate lesions coalescing to form plaques studded with multiple Comedone-like openings typical of hyperkeratotic LS.

Table 1. Morphology and duration wise distribution of dermoscopic findings

Dermoscopic findings, N (%)	Total (N)	Scleroatrophic plaques (N = 16)	Guttate (N = 8)	Hyperkeratotic plaques (N = 2)	Duration ≤ 2 years (N = 20)	Duration >2 years (N = 6)
White structure-less areas	26	16 (100%)	8 (100%)	2 (100%)	20 (100%)	06 (100%)
Chrysalis structures	21	13 (81.3%)	6 (75.0%)	2 (100%)	16 (80.0%)	05 (83.3%)
Linear irregular vessels	16	10 (62.5%)	5 (62.5%)	1 (50.0%)	13 (65.0%)	3 (50.0%)
Perifollicular scales	13	8 (50.0%)	4 (50.0%)	1(50.0%)	9 (45.0%)	4 (66.7%)
Keratotic plug	13	6 (37.5%)	5 (62.5%)	2 (100%)	10 (50.0%)	3 (50.0%)
Erythematous areas	10	6 (37.5%)	2 (25.0%)	01 (50.0%)	9 (45.0%)	1 (16.6%)
Rosettes	10	5 (31.3%)	5 (62.5%)	1 (50.0%)	8 (40.0%)	2 (33.3%)
Comedo-like openings	9	5 (31.3%)	2 (25.0%)	2 (100%)	6 (30.0%)	3 (50.0%)
Peppering of pigment	9	5 (31.3%)	4 (50.0%)	0	06 (30%)	3 (50.0%)

Peri-follicular scales were observed in 13 (50.0%) lesions, generalized white colored scales in 12 (46.2%) lesions and collarette like in 2 (7.7%) lesions (Figure 4). Pigment network was seen in 1 (3.8%) patient. Fibrotic beams and

erosions were absent in our patients. Hemorrhagic spots were seen in a single patient.

We found that non-polarizer mode was particularly useful for identifying texture change (due to sclerosis), CLO and minute

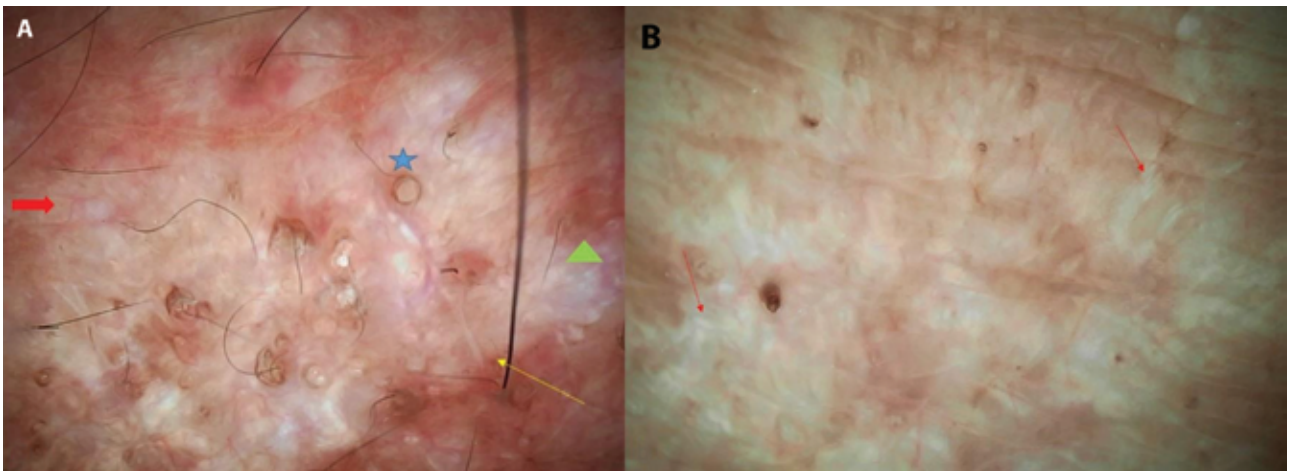


Figure 2. (A) Dermoscopy of scleroatrophic lesion with OITEZ e-scope digital microscope (20x magnification) suggestive of white to yellow structureless areas (yellow arrow) studded with multiple keratotic yellow–white plugs (blue star), chrysalis like structure (green triangle) and linear irregular vessels (red arrow). (B) Chrysalis structures (red arrow) seen on polarizer mode (20x) characterized by bright white orthogonal linear streaks seen only on polarized dermoscopy is suggestive of underlying dermal collagen homogenization.

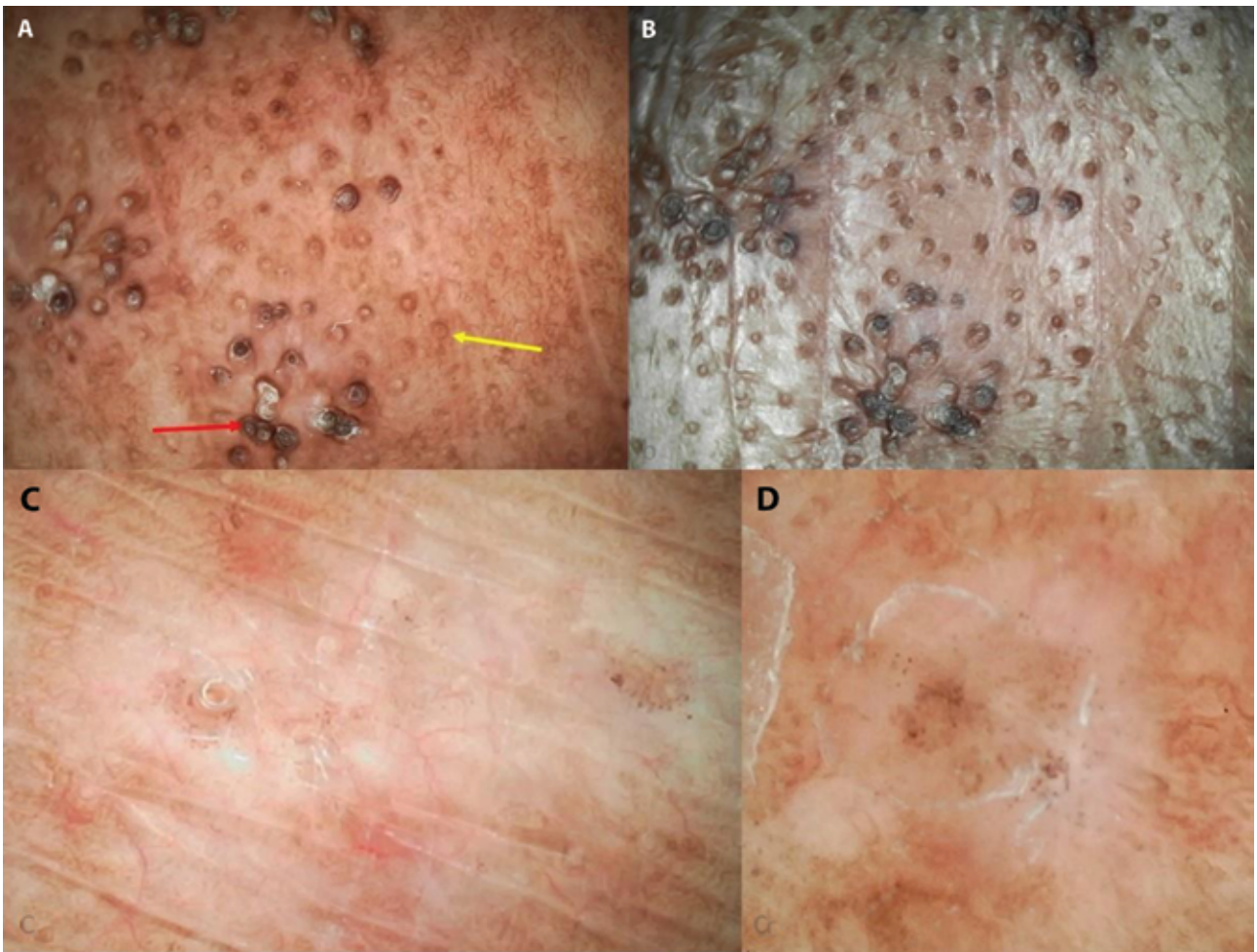


Figure 3. (A) Comedone-like openings (red arrow) and yellowish-white keratotic follicular plugs (yellow arrow) seen on polarizer mode (20x). (B) Better appreciated on non-polarizer mode. (C,D) Peppering blue-gray dots suggestive of melanin incontinence seen on polarizer mode (100x).

scales which were not visible otherwise (Figure 3, B and D). In addition, we observed stretched eccrine openings and rosettes on polarized mode. Rosettes defined as 4 white points,

arranged as a 4 leaf clover were seen in 10 (38.4%) lesions (Figure 5). In few patients with multiple CLO, we noticed that there was central clustering with radial arrangement (Figure 6).

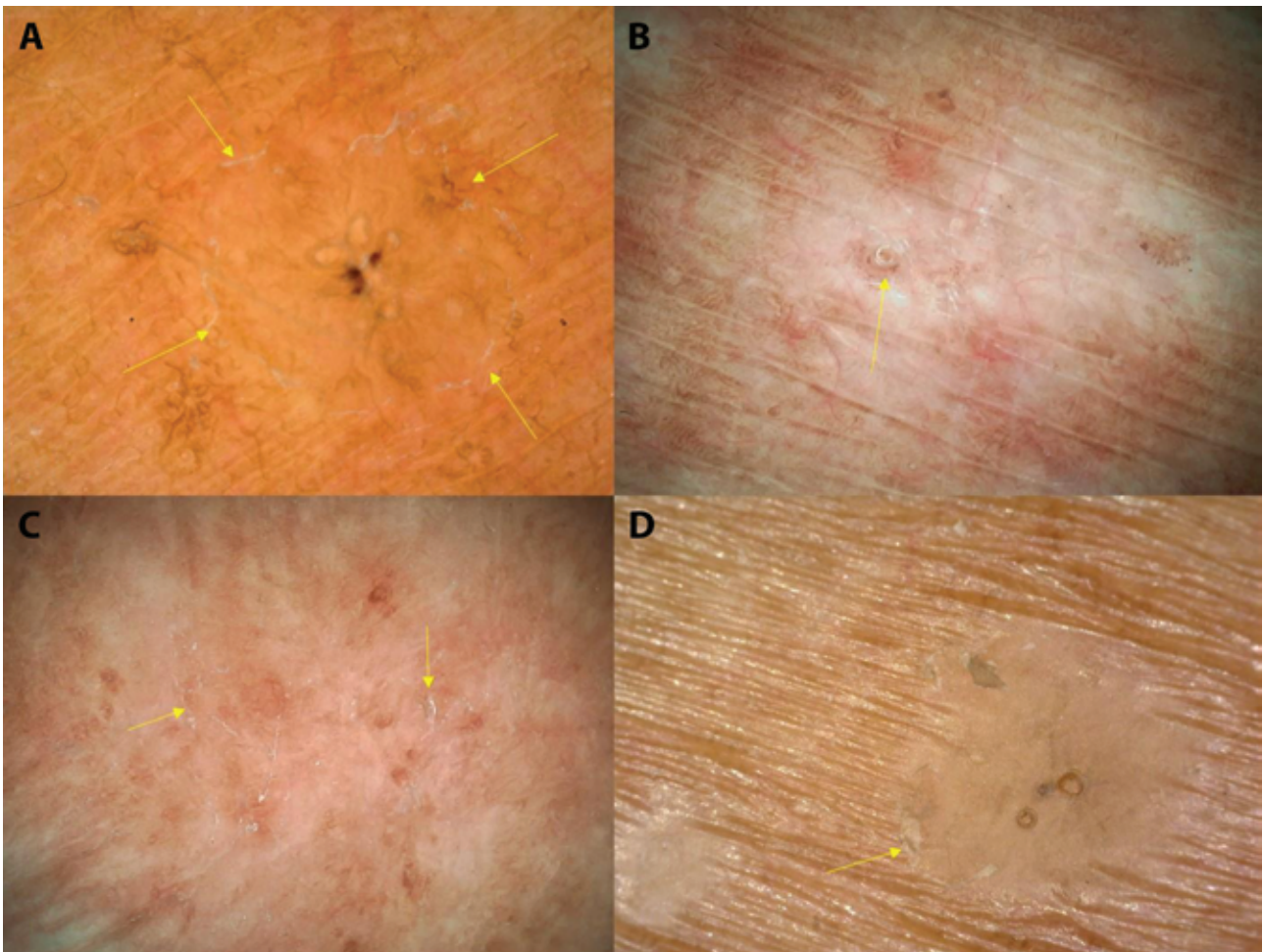


Figure 4. (A) Collarette scaling. (B) Peri-follicular scales. (C) generalized scaling appreciated on 20x polarizer mode. (D) Non polarized mode was particularly useful to identify minute scales not visible to naked eyes.

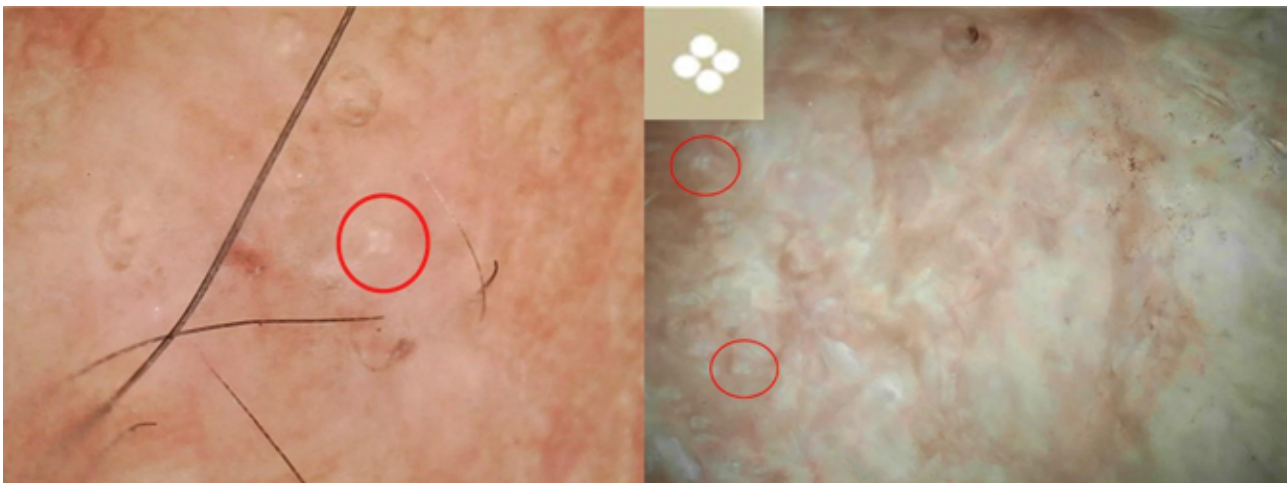


Figure 5. Rosette formation is also called as four-dot clods, seen on polarized dermoscopy (200x) as white shiny structures resembling a 4 leaf clover (red circle).

Histopathological examination findings of 21 biopsied lesions are enlisted in Table 2 (Figure 7).

Conclusions

Dermoscopy-aided algorithm of inflammatory disorders are defined by their characteristics based on background color,

vessel morphology and distribution, follicular involvement, surface changes such as scales and disease specific additional clues. Dermoscopy hallmarks of LS on polarizing mode include WSA with linear and dotted vessels suggestive of active lesion [3]. Keratotic plugs suggestive of follicular plugging [3]. Chrysalis like structures are seen as shiny, bright white orthogonal linear streaks which are also commonly observed

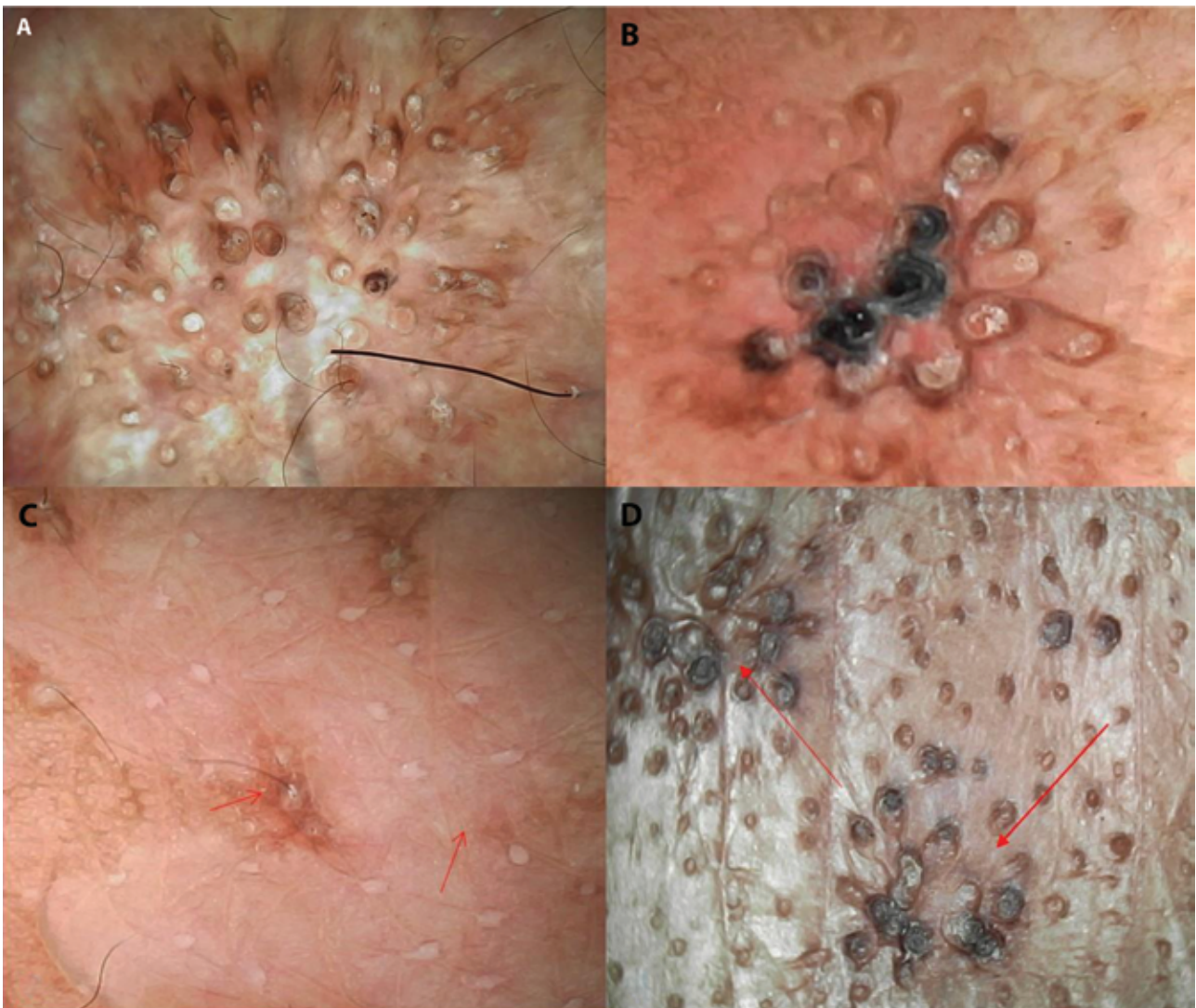


Figure 6. (A,B) central clustering with radial arrangement of comedone-like openings (CLO) and keratotic plugs. (C) Stretched eccrine openings suggestive of stretched acrosyringia due to upper dermal sclerosis and atrophic epidermis seen on polarizer mode (200x). (D) Grouping of CLO in coalescing atrophic papules of LS described as corymbiform pattern seen on non-polarizer mode (200x).

Table 2. Histopathology findings (N = 21)

Histopathology findings, N (%)	
Hyperkeratosis	21 (100%)
Collagen homogenization	17 (80.9%)
Perivascular infiltrate	13 (61.9%)
Epidermal atrophy	13 (61.9%)
Dilated lymphatics	12 (57.1%)
Vacuolar interface	10 (47.6%)
Follicular plugs	10 (47.6%)
Melanophages	9 (42.9%)
Band like infiltrate	7 (33.3%)
Papillary dermal edema	6 (28.6%)
Dilated blood vessels	4 (19.0%)
Perifollicular infiltrate	3 (14.3%)

in dermatofibroma, Spitz nevus, melanoma and basal cell carcinoma [4].

Dermoscopic features of LS correlate with histopathological findings such as WSA and correspond to epidermal atrophy and dense diffuse hyalinization of superficial dermis [5]. CLO is suggestive of dells which correspond to appendageal ostia [5,6]. Chrysalis structures are due to increased collagen deposition [4]. Scaling seen in LS lesions corresponds to hyperkeratosis while the peppering blue gray dots correspond to melanin incontinence seen after vacuolar interface dermatitis. Telangiectasia and dotted vessels represent the easy visibility of vessels due to inflammation from an atrophic epidermis [5]. Rosettes are a form of white shiny structures or are white shiny lines and white shiny areas. White shiny areas and lines have been correlated to dermal

fibrosis whereas exact correlation of rosette is unknown. It has been suggested that interaction of the polarized light with narrowed or keratin-filled adnexal openings could be the morphological correlation toward formation of rosettes. Rosettes have been linked histologically with alternating focal hyperkeratosis and normal corneal layer and keratin-filled openings. Haspeslagh et al proved on transverse sections that smaller rosettes are mainly caused by polarizing horny material at infundibular level in adnexal openings and larger rosettes mainly by concentric perifollicular fibrosis. We believe that rosette in case of LS will be due to same reasons [7]. Dermoscopy of LS on non polarizing mode is a useful adjunct to observe scaling, tiny CLO and keratotic plugs which aid in diagnosis. The closest mimicker of LS clinically and histopathologically is morphea, which can be differentiated by dermoscopy. Shim et al found a statistically significance for WSA and CLO in LS whereas fibrotic beams crossed by spreading telangiectasia for morphea [8].

WSA were seen in all patient of our study similar to Liu et al and Borges et al whereas it was seen in 88.6% patients in a study by Errichetti et al [6,9,10]. Shim et al reported much lesser percentage (66.7%) of WSA [8]. CLO was seen in 34.6% of patients which was lesser than Shim et al (77.8%) [8]. The most common vessel morphology observed in our study was linear irregular vessels (61.53%) which was less compared to Liu et al (72.8%) and higher than Errichetti et al (25.7%) [6,9]. Dotted vessels were observed in 15.4% patients which was similar to Liu et al (16.8%), Borges et al (13.3%), lesser as compared to Errichetti et al (28.6%) and higher as compared to Shim et al (5.6%) [6,8,9,10]. Chrysalis-like structure suggestive of collagen deposition was seen

in 80.8% study participants which was comparable to study by Liu et al (84%) and lower in study by Errichetti et al (40%) and Borges et al (26.7%) [6,9,10] (Table 3).

Scaling particularly peri-follicular (50.0%) represented a very common feature of LS in our study which was less as compared to previous study [6]. Early lesions of LS show keratotic plugs on dermoscopy whereas older ones demonstrate chrysalis like structure [10]. However, in our study there was not much difference in these 2 parameters, instead we saw that vascular findings like irregular linear vessels, and erythematous areas were more frequent in early lesions. Peppering blue-gray dots suggestive of melanin incontinence was seen more frequently in old lesions of LS probably indicating lesion inactivity (Table1).

Rosettes are a particular finding we want to highlight that has not been described before with LS. Stretched eccrine openings represent stretched acrosyringia of the eccrine glands due to sclerosis of the upper dermis and atrophy of the epidermis. The grouping of CLO in coalescing atrophic papules of LS better visualized on dermoscopy has been described as cribriform or corymbiform [2].

On dermoscopy keratotic plugging was seen in more number compared to follicular plugging on histopathology. This could be because the keratin filled craters correspond to adnexal (eccrine and follicular openings) which may not always be visualized in a particular section of skin biopsy. Broader band of hyalinization was associated with prominent chrysalis like structures.

Limitations of our study were the small sample size, the fact we couldn't study multiple lesions on the same patient and do their biopsies to correlate their results with dermoscopic findings.

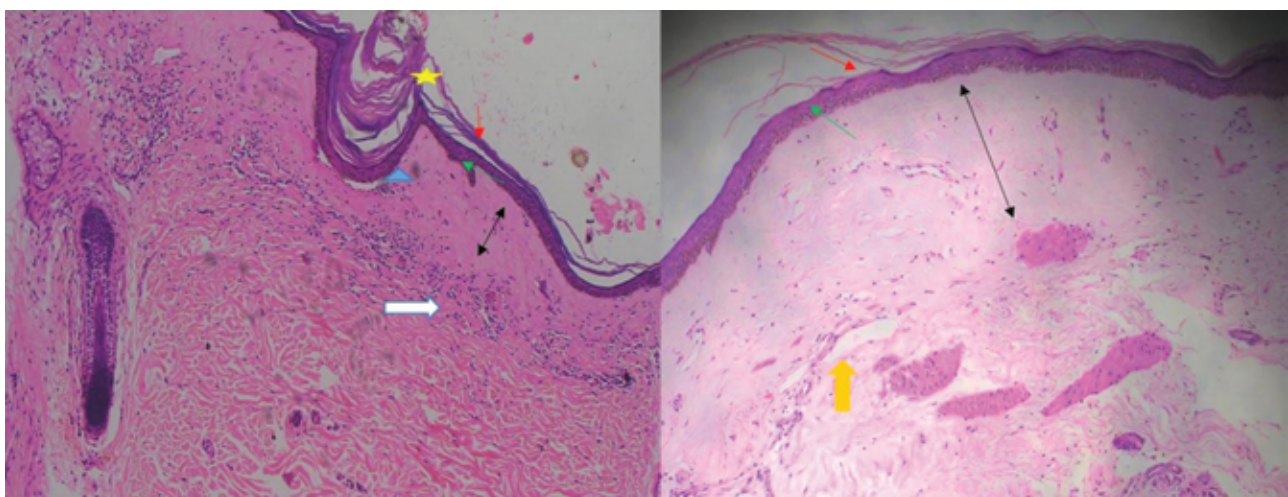


Figure 7. Histopathology examination of Lichen sclerosus (H&E, 20x magnification) shows hyperkeratosis (red arrow), follicular plugging (yellow star), atrophy of epidermis with flattening of rete ridges (green arrow), focal basal cell degeneration (blue triangle), subepidermal band of homogenization (black arrow) corresponding to the sclerosis with a lichenoid infiltrate of lymphocytes beneath it (white arrow) and dilated lymphatics (orange arrow).

Table 3. Dermoscopic profile of Lichen sclerosis and comparison with other published data

Dermoscopic features, N (%)	Present study	Errichetti et al [6]	Shim et al [8]	Liu et al [9]	Borges et al [10]
White structureless areas	26(100)	31 (88.6)	12(66.7)	124(99.2)	15(100)
Chrysalis like structure	21(80.8)	14 (40.0)	--	105(84.0)	4(26.7)
Linear irregular vessels	16(61.5)	9 (25.7)	--	91(72.8)	--
Yellowish-white keratotic follicular plugs	13(50.0)	28(80.0)	--	63(50.4)	5(33.3)
Scales (perifollicular)	13(50.0)	15 (42.9)	--	--	9 (60.0%)(type not specified)
Linear branching vessels	10(38.5)	1(2.9)	9(50.0)	--	5(33.3)
Comedone-like openings	9(34.6)	1(2.9)	14(77.8)	--	--
Dotted vessels	4(15.4)	10(28.6)	1(5.6)	21(16.8)	2 (13.3)

In conclusion, LS is a rare disorder that can be difficult to differentiate clinically from a number of disorders of hypopigmentation and sclero-atrophic disorders, and hence requires skin biopsy. Dermoscopy is a simple diagnostic tool that helps in the early diagnosis of LS with specific and characteristic patterns which can avoid invasive procedure like biopsy. Both non-polarized and polarized dermoscopy must be done to visualize the changes of LS well. Larger sample size study is required to ascertain the newer dermoscopic findings.

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