

# Histochemical differentiation of localized morphea-scleroderma and lichen sclerosus et atrophicus

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Dermatologic literature has debated the occurrence of concomitant morphea-scleroderma (M-S) and lichen sclerosus et atrophicus (LSA) for sometime. Presentation of a case which has the appearance of both M-S and LSA creates a diagnostic dilemma frequently unresolved even by histopathology. Routine hematoxylin and eosin stained sections may add to the confusion and the difficulty of the differentiation, but examination for the presence or absence of elastic fibers in the upper corium of the lesions affords a definitive separation of these two conditions.

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The clinical presentation of some cases of localized morphea (M) or small plaques of scleroderma (S) appears very similar to lesions of lichen sclerosus et atrophicus (LSA). At times the surface of M-S lesions gives the impression of atrophic changes seen with LSA; and occasionally LSA cases feel indurated on palpation as one would expect with M-S. These examples have led to reports of association of M-S and LSA and LSA-like circumscribed scleroderma (1-3).

The purpose of this paper is not to agree with or reject the idea of concurrence of M-S and LSA, but to offer a way to differentiate the unusual cases of these two conditions and to eliminate the problem of misdiagnosis.

## Material and methods

Twenty consecutively received biopsies which were diagnosed as LSA at our laboratory, according to the accepted diagnostic criteria (4, 5) together with 23 consecutive cases of M-S, similarly diagnosed were reviewed. Hematoxylin and eosin (H&E) and Pinkus' acid orcein and Giemsa (O&G) stained sections of these cases were studied. In addition silver-

stained sections of Cases 15 and 19 of the LSA group and Case 5 of M-S group were prepared and studied.

The information supplied by the clinician sending the biopsy to the laboratory is presented in Tables 1 and 2.

## Results

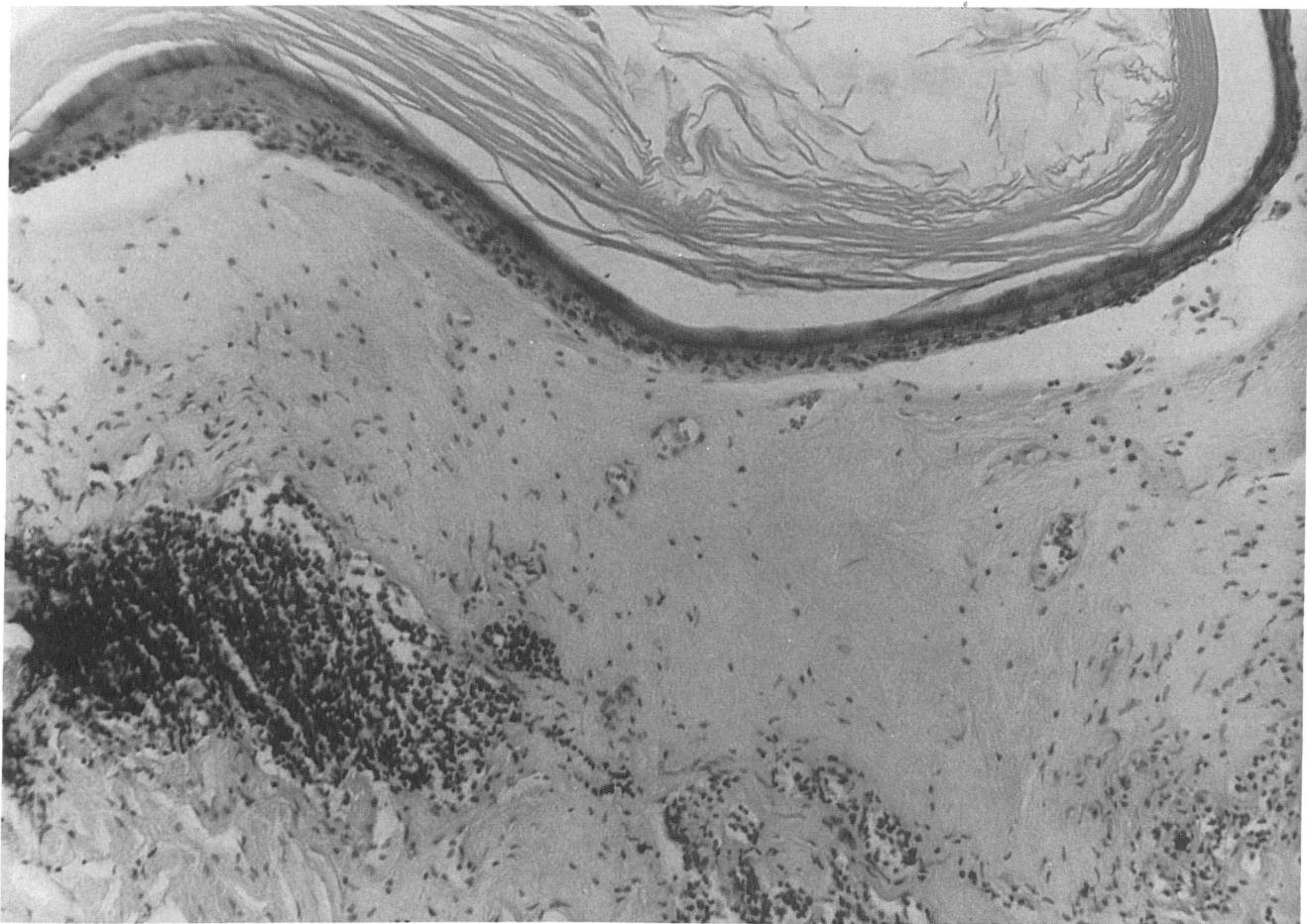
### Clinical data

LSA cases ranged in age from 10 to 79 years. The patients in the study were women except for three men. Although 9 patients had lesions in the perineal and inguinal areas, other regions of the trunk and extremities were also involved. In the morphea-scleroderma cases ages ranged from 14 to 69 years; with the women to men ratio of 17:6. The M-S lesions were not prevalent in any location on the body.

The most interesting and fully predictable finding was that the primary clinical differential diagnosis for LSA was morphea or guttate scleroderma and vice versa. Clinical presentation at times was compatible with both conditions.

### Light microscopy

Light microscopy of most of the cases (18 of LSA and 22 of M-S groups) showed typical findings of



*Fig. 1.* Lichen sclerosus et atrophicus: characteristic atrophy of the epidermis covered with orthokeratotic keratin and homogenized and edematous upper dermis with lymphocytic infiltrate (H&E,  $\times 100$ ).

LSA (Fig. 1) or M-S (Fig. 2). The examination of O&G stained sections was very revealing and supported the basis for this report which is that for differentiation of questionable cases one can depend on the visualization of the presence or absence of elastic fibers in the upper dermis. A typical LSA case (Fig. 3) shows very few, if any, elastic fibers in the edematous upper corium, whereas characteristic M-S (Fig. 4) shows abundant elastic fibers throughout the thickened dermis.

Occasionally (one case among the 23 M-S group in the present study) there are cases of M-S which clinically are not clear-cut and histopathologically also show some features of LSA (Fig. 5). In these cases there may be some edema and appearance of LSA-like changes in the upper dermis although the rest of the dermis shows the changes that one expects to see with M-S. On examination of O&G stained sections (Fig. 6) the presence of elastic fibers throughout the dermis, especially in the upper parts

and right below the epidermis attests to the correct diagnosis being M-S rather than LSA.

On the other hand, at times a very early lesion of LSA (Fig. 7) presents confusing clinical appearance and histopathologic examination also gives the impression of increased fibrosis of the dermis with the characteristic findings for LSA sparse or missing (two cases out of the 20 LSA group in the present cases). In these instances examination of O&G stained sections (Fig. 8) shows the absence of elastic fibers in the uppermost areas of the dermis, leading to the diagnosis of early LSA. The silver stained sections of the unusual cases of LSA (2 cases) and MS (1 case) showed no evidence of spirochetes.

### Comment

When this study was started the impression was that the author had come across an original obser-

Table 1. Clinical data of 20 cases of lichen sclerosus et atrophicus (diagnosis based on histopathologic findings).

Age/Sex	Location	Clinical diagnosis
1. 67/F	(L) chest	Isolated plaque of morphea, R/O LSA
2. 65/F	Posterior neck	LSA vs. morphea
3. 77/F	Left inguinal area	R/O LSA, R/O Lichen simplex chronicus
4. 73/F	(R) posterior shoulder	Atrophoderma vs. LSA vs. morphea
5. 63/F	(R) inguinal region	?
6. 42/F	Labia	R/O leukokoratosis – kraurosis vulvae
7. 79/F	(L) forearm	LSA ?
8. 56/F	(R) perirectal	LSA ?
9. 51/M	Penile prepuce	R/O LSA, Bowenoid papulosis, squamous cell carcinoma
10. 34/F	Labia majora	Atopic vaginitis, R/O leukoplakia
11. 34/F	Vaginal introitus	R/O leukoplakia
12. 10/F	(L) knee	LSA
13. 27/M	(L) side of back	LSA, R/O BCE
14. 33/F	Vulva	R/O LSA, leukoplakia
15. 36/M	(R) buttock	? LSA
16. 77/F	Upper back	? LSA
17. 19/F	Lower neck	Scar, R/O morphea, R/O LSA
18. 32/F	Back	LSA, morphea
19. ?/F	(R) shoulder	LSA
20. 55/F	(L) upper back	LSA, R/O lichen planus, atrophoderma

Table 2. Clinical data of 23 cases of morphea-scleroderma (diagnosis based on histopathologic findings).

Age/Sex	Location	Clinical diagnosis
1. 34/F	(L) upper arm	Morphea
2. 53/F	(L) shoulder	R/O scleroderma, granuloma annulare, drug eruption
3. 26/M	Back	Localized morphea
4. 37/F	(R) back	? connective tissue nevus
5. 65/F	(R) axilla	Morphea
6. 21/F	Hip	R/O morphea, LSA
7. 47/F	(L) ant. tibia	? erythema induratum ? localized morphea
8. 14/F	Back	Morphea
9. 70/F	(L) leg	R/O scleroderma
10. 25/F	(L) trunk	Fixed drug eruption vs. parapsoriasis
11. 53/F	Neck	R/O guttate morphea-LSA
12. 14/F	(R) back	Morphea ? R/O LSA
13. 29/M	(R) temple	Morphea
14. 36/M	(L) lateral back	Diffuse cutaneous scleroderma- morphea
15. 27/F	(L) breast	Morphea, R/O breast ca.
16. 35/M	(L) ext. arm	? localized morphea or LSA
17. 54/F	(L) calf	R/O Morphea-LSA, parapsoriasis
18. 48/M	Upper back	LSA vs. morphea variant
19. 26/F	(L) upper back	LSA vs. scleroderma
20. 69/F	(L) thigh	LSA, morphea
21. 20/F	L.U.Q. abdomen	LSA vs. morphea
22. 24/F	(R) foot	LSA, R/O morphea
23. 49/M	Abdomen	R/O morphea-LSA



Fig. 2. Morphea-scleroderma: characteristic changes of the dermis with sclerosis and thickening of the collagen and absence of intact follicular structures. There are small foci of inflammatory infiltrate and the overlying epidermis is normal (H&E,  $\times 100$ ).

vation; namely the great diminution or absence of elastic fibers in the upper corium characterized cases of LSA and persistence or even increased elastic fibers was seen in M-S. Review of the dermatologic literature proved that this was personal rediscovery of previously known facts (*vide infra*).

The intent of this presentation is not to discuss the accuracy of the belief on the part of some observers that cases of LSA and M-S occur together or that some lesions have features of both diseases. Although it seems that, considering the typical clinical and histopathologic presentations of these diseases, they must have a very different *modus operandi*, and there is no good reason to think that the two diseases result from one set of events in the same patient.

Increased amounts of collagen fibers in cases of M-S is well known, and recent data indicate that the dermal thickening is most likely due to an increase in Type III collagen (6). No decrease of elastic tissue in scleroderma has been reported by some (7), while other investigators have reported various changes in the elastic fibers in scleroderma (8–9)

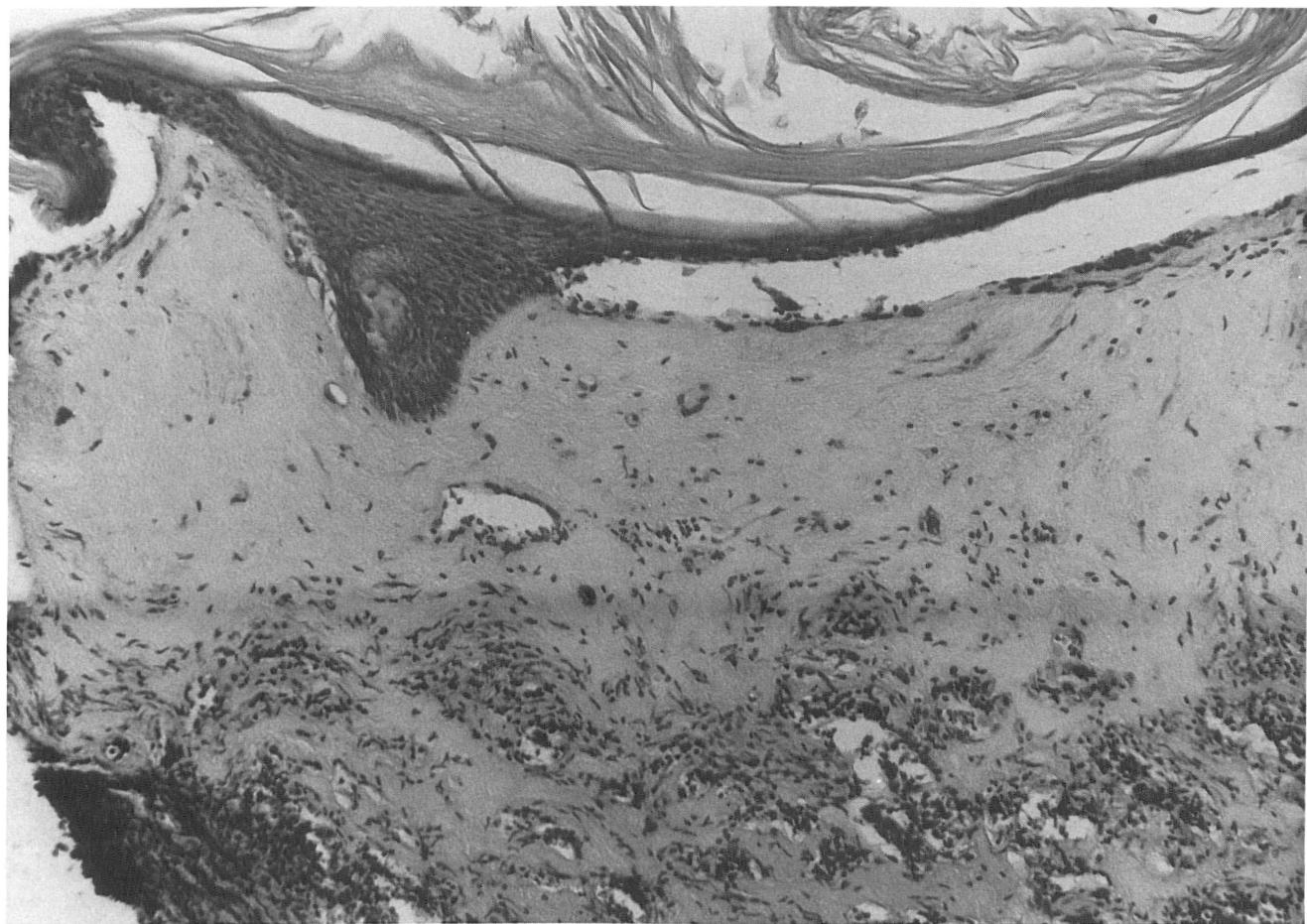


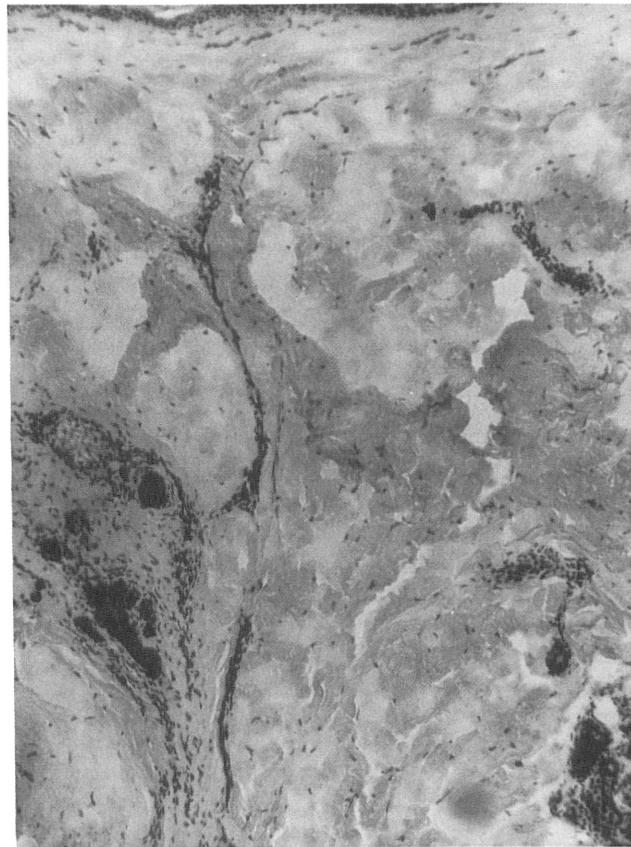
Fig. 3. Lichen sclerosus et atrophicus: characteristic findings as in Fig. 1 with complete absence of elastic fibers in the upper dermis (acid orcein & Giemsa,  $\times 100$ ).

but no absence of these fibers. Some observers have stated that elastic fibers proliferate in scleroderma (10).

On the other hand the disappearance of elastic fibers in the upper dermis has been observed in LSA (11–14), but this fact has not been emphasized or been disseminated well. Romppanen et al. (15) point up this lack of emphasis in their light microscopic and electron microscopic studies of LSA. Their immunoelectron microscopic findings in LSA show that there was only occasional positive reaction for elastic material in the superficial dermis. Dupré and Viraben (16) also report that their electron microscopic examination of LSA of the glans penis showed the basal lamina to have no anchoring fibers. This suggests other structural defects besides the lack of elastic fibers in the upper dermis in LSA. Relative to this, Kint and Geerts (17) found collagen fibrils in the epidermal intercellular spaces as Mann and Cowan (18) had observed and suggested

that the dermoepidermal junction in LSA was not normal. They (17) also observed that: "the elastin fibers were rare" in the upper dermis in LSA.

From these immune and electron microscopic studies it would appear that the absence or great diminution of elastic fibers in the upper dermis in lesions of LSA is well documented corroborating what is observed with light microscopic examination. Use of elastic tissue stains which show the fine elastic fibers in the upper dermis can be helpful in resolving the questionable cases of M-S-like LSA or lesions of M-S with some features of LSA; furthermore by studying more of these cases we may ultimately arrive at the resolution of the question concerning possible concurrence of LSA and M-S in the same patient.



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*Fig. 4.* Morphea-scleroderma: elastic fibers are present throughout the dermis, same case as in Fig. 2 (acid orcein & Giemsa,  $\times 100$ ).

*Fig. 5.* Morphea-scleroderma: changes reminiscent of LSA with atrophic epidermis and homogenization of the collagen. There is sclerosis of the collagen bundles and atrophy of pilosebaceous structures (H&E,  $\times 100$ ).

*Fig. 6.* Morphea-scleroderma: elastic fibers are present throughout the dermis and there is thickening of collagen bundles (acid orcein & Giemsa,  $\times 100$ ).



Fig. 7. Lichen sclerosus et atrophicus: early case without epidermal atrophy and the upper dermis does not show definitive lymphedema and homogenization (H&E,  $\times 100$ ).

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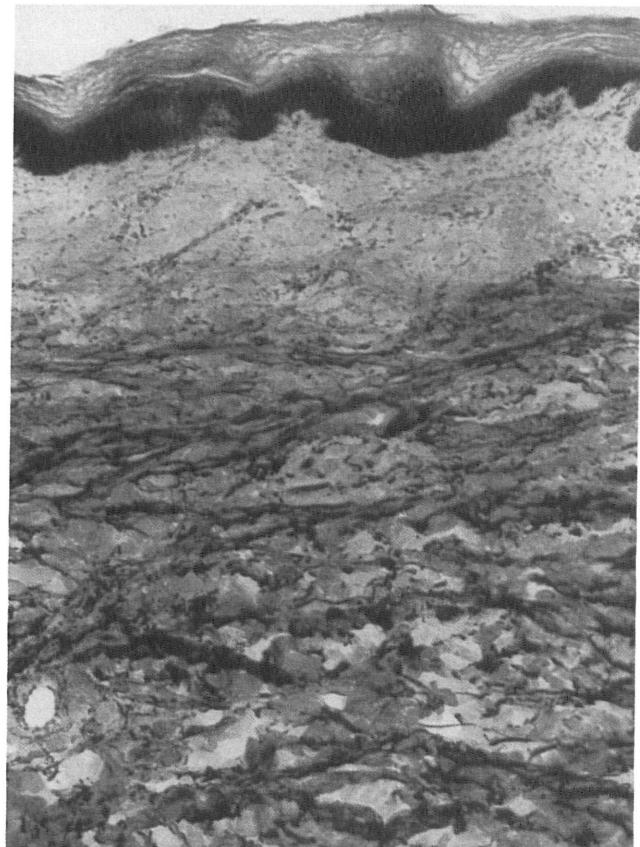


Fig. 8. Lichen sclerosus et atrophicus: same case as in Fig. 7. The upper dermis is devoid of elastic fibers and the early homogenization and lymphedema are easily observed (acid orcein & Giemsa,  $\times 100$ ).

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