



Coexisting Lichen Sclerosus and Morphea

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BACKGROUND

For decades, dermatopathologists claimed that lichen sclerosus and morphea could not occur simultaneously. However, over the years, sporadic cases of coexisting lichen sclerosus and morphea have been reported in the literature.

Lichen sclerosus is a chronic inflammatory dermatitis that is characterized by white, slightly raised, atrophic papules and plaques on the skin. Histologic features include hyperkeratosis with follicular plugs, atrophy of the Malpighian layer with vacuolar degeneration, edema, homogenization of the upper dermal collagen, and a mid-dermal inflammatory infiltrate.

Morphea is a type of localized scleroderma. This inflammatory fibrosing disease presents with round, erythematous to violaceous patches and plaques with a white sclerotic center. Histologically, there is a dense, sclerotic appearance to the dermal collagen, particularly in the mid-to-deep reticular dermis.

CASE PRESENTATION

A 33-year-old male with no previous medical history presented with a two year history of an asymptomatic plaque on his ribcage. The patient was not taking any medications except for a daily multi-vitamin.

Physical examination:

- 4 x 3 cm white plaque on the left ribcage
- No other cutaneous or mucosal lesions present

Hematoxylin and eosin stained sections from the punch biopsy revealed the following:

- Hyperkeratosis
- Atrophic epidermis with loss of the normal rete ridge pattern
- Prominent homogenization of the papillary dermis with scattered melanophages present in the background

CASE PRESENTATION

- Histologic findings compatible with morphea were also noted:
 - Thick and hyalinized collagen bundles in the reticular dermis
 - Loss of peri-eccrine fat
 - Peri-adnexal, lymphoplasmacytic inflammatory infiltrate

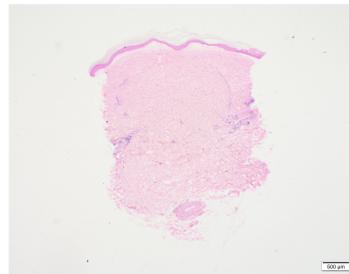


Figure A:
Low-power image highlighting the classic "square" biopsy (20x)

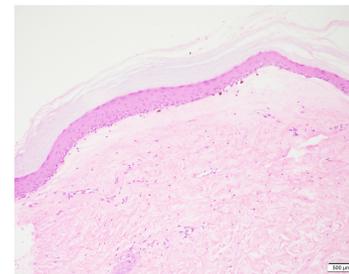


Figure B:
Epidermal atrophy and homogenization of the papillary dermal collagen (100x)

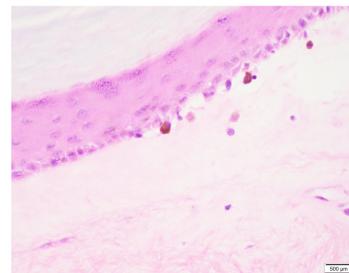


Figure C:
Rare foci of interface change with scattered melanophages in the papillary dermis (400x)

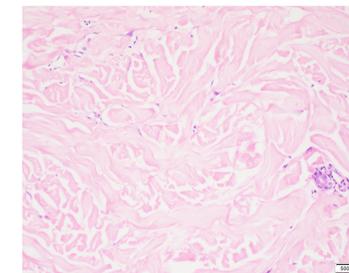


Figure D:
Hyalinized and thickened collagen bundles in the reticular dermis (200x)

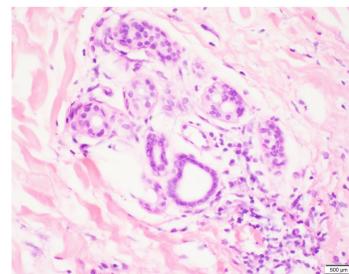


Figure E:
Loss of peri-eccrine fat with an associated lymphoplasmacytic inflammatory infiltrate (400x)

DISCUSSION

- Both lichen sclerosus and morphea are chronic inflammatory skin conditions which were long thought to be distinct disease processes. However, with more cases reporting the two conditions taking place simultaneously, it is now thought that the two conditions may exist on the same disease spectrum with a possible linked etiology.
- Coexisting lichen sclerosus and morphea may be linked to systemic autoimmune diseases. Several reports of lichen sclerosus occurring simultaneously with morphea have been described in patients with various autoimmune diseases, including autoimmune thyroid disease and insulin-dependent diabetes mellitus.
- Some reports have suggested that lichen sclerosus is significantly more common in patients with morphea than those in the general population. It has been reported that anywhere between 5.7 – 45% of those with plaque morphea have concomitant lichen sclerosus. Since there is a known risk for lichen sclerosus to evolve into squamous cell carcinoma, Lutz et al. advocated for the importance of screening for lichen sclerosus in patients with morphea.

CONCLUSION

- Coexisting lichen sclerosus and morphea is a rare skin condition that was disputed against for decades, with dermatopathologists claiming that the two conditions were distinct entities.
- Here we report a case of coexisting lichen sclerosus and morphea, providing further evidence that these two diseases can occur simultaneously and may exist along a spectrum.

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