Connective Tissue Diseases

Paul K. Shitabata, M.D.

Director of Dermatopathology Harbor-UCLA Dermatology

Pertinent histopathological features of LSEA include all except:

- Follicular plugging
- Interface dermatitis
- Increased elastic fibers within the papillary dermis
- Dermal sclerosis
- Mid-dermal inflammatory infiltrate

Increased elastic fibers

The hallmark of LSEA is decreased elastic fibers within the papillary dermis.

Associations with eosinophilic fasciitis include all except:

- Positive anti-nuclear antibody
- Peripheral blood eosinophilia
- Serotonin ingestion
- Acute onset
- Dermal sclerosis extending into subQ fat

Sertonin ingestion

Classically, the syndrome has been linked to l-Tryptophan ingestion leading to the similar eosinophila-myalgia syndrome

Characteristic histopathologic features of morphea and variants include all except:

- Less sclerosis than systemic scleroderma
- Histologically unremarkable skin
- Positive ANA
- Histopathologic overlap with LSEA
- Calcinosis cutis

Less sclerosis than systemic scleroderma

Morphea cannot be distinguished from systemic scleroderma by routine histopathology. There are increasing reports of overlap cases of LSEA and morphea, blurring the distinction between the two diseases.

Which is not a classic "5L"?

- Leprosy
- Lupus erythematosus
- Lymphocytic lymphoma
- Lymphocytic infiltrate of Jessner
- Polymorphous light eruption

Leprosy

The 5Ls can also include:

Lymphocytoma cutis

Lyme disease

Lues (secondary syphilis)

Histopathological cutaneous changes of SLE include all except:

- DLE changes
- Tumid changes
- Leukocytoclastic vasculitis
- Intra-epidermal bullous change
- Fibrinoid deposits

Intra-epidermal bullous change

Bullous LE is a sub-epidermal bullous disorder

Jessner's vs DLE

	Jessner's	DLE
DIF		
Interface changes		
T vs B-cell dermal		
phenotype		
S and D PV		
infiltrate		
Dermal mucinosis		

	Jessner's	DLE
DIF	_	+
Interface changes	_	+
T vs B-cell dermal phenotype	T cells	T cells
S and D PV infiltrate	+	+
Dermal mucinosis	-	+

Dermatomyositis Associated Ab

Disease Variant	Autoantibody
Pulmonary fibrosis	
Sclerodermatomyositis	
At DEJ on DIF	

Disease Variant	Autoantibody
Pulmonary fibrosis	Jo1
Sclerodermatomyositis	Ku
At DEJ on DIF	C5b-9
	(Membrane attack complex)

Poikilodermatous histopathology in all except:

- Rothmund-Thompson syndrome
- Congenital ichthyosiform erythroderma
- Dermatomyositis
- Bloom's syndrome
- Dyskeratosis congenita

Congenital ichthyosiform erythroderma
 This classically presents with epidermolytic hyperkeratosis