

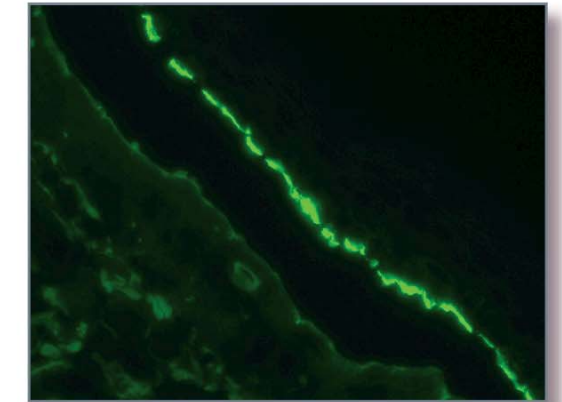
IMAGE	DISORDER	DIRECT IF FINDINGS
	<i>Dermatitis herpetiformis</i>	Granular or fibrillar IgA deposits at dermal-epidermal junction, with accentuation in superficial papillary dermis
	<i>Epidermolysis bullosa acquisita</i>	Linear IgA at basement membrane zone
	<i>Henoch-Schönlein purpura</i>	Granular deposits of predominantly IgA in superficial blood vessel walls
	<i>Linear IgA bullous dermatosis</i>	Linear IgA at basement membrane zone
	<i>Lupus</i>	Granular, fibrillar, or homogeneous IgM (and other immunoglobulins, along with C3) at dermal-epidermal junction zone ('lupus band')
	<i>Pemphigoid, bullous</i>	Linear C3 and/or IgG at basement membrane zone; 'salt-split' testing to distinguish EBA v. BP
	<i>Pemphigoid, cicatricial</i>	Linear C3 and IgG at basement membrane zone
	<i>Pemphigoid ('Herpes') gestationis</i>	Linear C3 at basement membrane zone; lower frequency of IgG
	<i>Pemphigus foliaceus and vulgaris</i>	'Chicken wire' pattern of IgG in epidermis and mucous membranes; in vulgaris, deposits present predominantly in lower portions of epidermis
	<i>Porphyrias</i>	Homogeneous, concentric deposits of predominantly IgG within walls of dilated vessels in papillary dermis and in basement membrane zone
	<i>Vasculitis</i>	Granular deposits of C3 and IgM in superficial blood vessel walls

## D

irect immunofluorescence studies, i.e., looking for the presence of deposits of immunoglobulin and complement in specific locations within a skin or mucosal biopsy, can play an important role in the identification of selected inflammatory skin disorders. For example: granular deposition of IgA at the dermal-epidermal junction is the hallmark of dermatitis herpetiformis. The presence of IgG deposits in a 'chicken wire' pattern within the epidermis is characteristic of the pemphigus family of disorders. Bullous pemphigoid is characterized by linear deposits of IgG and/or C3 along the dermal-epidermal junction. Indirect immunofluorescence, using the patient's serum, has a more limited role, but can also yield important information regarding the presence and titer of circulating antibodies to desmoglein proteins ('intercellular substance') characteristic of pemphigus.

## U

Using a 'salt-split skin' preparation, in which a biopsy of skin from a normal individual is artificially 'split' using a high salt solution, it is possible to distinguish between bullous pemphigoid (BP) and epidermolysis bullosa acquisita (EBA), which can yield identical direct immunofluorescence patterns of linear IgG deposits at the dermal-epidermal junction. In the 'salt-split skin' preparation, the hemidesmosomal protein identified by circulating antibodies in patients with BP is present on the roof of the induced separation, whereas the serum of EBA patients contains antibodies that react with proteins present in the floor of the separated skin preparation.



Salt-split skin preparation showing linear deposits of IgG on roof of blister, pointing to the diagnosis of bullous pemphigoid

## Spindle Cell Tumors of the Skin

Spindle cell tumors in the skin present a relatively restricted and distinctive differential diagnosis which includes atypical fibroxanthoma, dermatofibroma, dermatofibrosarcoma protuberans, leiomyoma/leiomyosarcoma, melanoma, and spindle cell squamous cell carcinoma. These tumors can be subclassified using a panel of antibodies that includes antibodies to p63 or cytokeratin 5, CD34, muscle actins, desmin, S100, and CD10.

	p63/Cytokeratin 5	CD34	Muscle actins	Desmin	S100	CD10
Spindle cell squamous cell						
Spindle cell/desmoplastic melanoma						
Leiomyoma/leiomyosarcoma						
Dermatofibroma						
Dermatofibrosarcoma protuberans						
Atypical fibroxanthoma						

Heatmap content available in the printed Pathology Reference Guide

Almost always positive	Usually positive	Not helpful	Usually negative	Almost always negative

See introduction to heat maps page 15