

## Paraneoplastic Skin Conditions, Part 2

William W. Huang, MD, MPH

Dr. Huang is Assistant Professor of Dermatology, Wake Forest University School of Medicine, Winston-Salem, North Carolina.

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Diagnosis	Commonly Associated Malignancies <sup>a</sup>	Common Sites and Presentations	Other
Dermatomyositis	Women more commonly affected than men ( <b>women: breast and ovarian carcinoma; men: gastric carcinoma and lymphoma</b> ); risk for malignancy returns back to normal 2–3 years after diagnosis; no increased risk for malignancy in pediatric dermatomyositis	Weakness of proximal muscle groups; many skin findings (ie, erythema and swelling of eyelids [heliotrope rash]; photosensitivity; violaceous flat-topped papules located over the dorsal aspect of the knuckles, wrists, elbows, and knees [Gottron papules]; symmetric, nonscaling, violaceous macules or plaques [Gottron sign]; fissured, scaly, hyperkeratotic and hyperpigmented hands [mechanic hands]; poikilodermatous macules distributed in a “shawl” pattern over the shoulders, arms, and upper back [shawl sign]; macular violaceous erythema of the lateral thighs and hips [holster sign]; calcinosis cutis; cuticular dystrophy; proximal nail fold telangiectases); interstitial lung disease in subset of patients	More commonly autoimmune than paraneoplastic (10%–25%); signs of muscle inflammation (ie, muscle enzymes, muscle biopsy, EMG, MRI); positive ANA common; multiple antibodies: anti-Mi2 associated with acute onset of classic dermatomyositis and favorable prognosis; anti-Jo1 associated with interstitial lung disease, Raynaud phenomenon, arthritis, and mechanic hands; anti-SRP associated with severe polymyositis; anti-Ku associated with overlapping features of myositis and scleroderma; anti-p140 and anti-p155 are cancer associated
Erythema gyratum repens	<b>Lung</b> , esophageal, breast, colorectal, and bladder cancers	Severe pruritus; “wood grain” patterned figurate erythema with trailing scale; rapid migration (up to 1 cm/d)	Symptoms improve with treatment of underlying malignancy
Hypertrichosis lanuginosa acquisita	<b>Lung</b> , colorectal, breast, uterine, and bladder cancers	Glossitis; rapid growth of fine lanugo hair	Can be associated with metabolic abnormalities and some medications

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Table. (continued)

<b>Diagnosis</b>	<b>Commonly Associated Malignancies<sup>a</sup></b>	<b>Common Sites and Presentations</b>	<b>Other</b>
Multicentric reticulohistiocytosis	No specific type; reported with melanoma, sarcomas, leukemias, and lymphomas, as well as breast, colon, cervical, gastric, and ovarian cancers	Nontender reddish brown nodules commonly found on hands and face; symmetric erosive arthritis that can progress to arthritis mutilans	Paraneoplastic in approximately 25% of cases
Necrolytic migratory erythema	<b>Alpha islet cell neuroendocrine pancreatic cancer</b>	Part of the glucagonoma syndrome; glossitis; erythema, vesicles, pustules, and erosions commonly in a periorificial, groin, buttock, and acral distribution; weight loss; diabetes mellitus; diarrhea, stomatitis	Often metastatic by time of diagnosis; elevated serum glucagon level; treatment includes tumor excision, somatostatin, and amino acids
Paraneoplastic pemphigus	<b>Non-Hodgkin lymphoma, chronic lymphocytic leukemia, thymoma, and sarcoma;</b> Castleman disease most common cause in children and adolescents	Intractable stomatitis and oral erosions; skin lesions variable with erythematous papules, lichenoid papules, targetoid lesions, bullae, and erosions	Antigens include Dsg3, Dsg1, plectin, desmoplakin 1, desmoplakin 2, BPAg1, envoplakin, and periplakin; IgG1 and IgG2 intercellular and linear at DEJ on DIF; IIF substrate: rat bladder
Leser-Trélat sign	Gastric, colon, breast, and lung cancers; 60% of cases are associated with adenocarcinoma of the gastrointestinal tract	Sudden and rapid presentation of numerous seborrheic keratoses; intense pruritus; can present along with tripe palms, acanthosis nigricans, and acquired ichthyosis	Eruptive seborrheic keratoses also can be seen after an erythroderma not in the setting of malignancy
Sweet syndrome (acute febrile neutrophilic dermatosis)	<b>Acute myelogenous leukemia, lymphoma, other hematologic malignancies, and solid organ malignancies (breast in women; gastrointestinal in men)</b>	Fever; erythematous and edematous tender dermal nodules on face, neck, upper trunk, and upper extremities that may progress to bullae; anemia, elevated ESR, and neutrophilia common; solitary, vesiculobullous, and necrotic lesions have an increased risk for malignancy	Paraneoplastic in approximately 20% of cases; can be associated with medications, infection, or inflammatory systemic disorders

Abbreviations: EMG, electromyogram; MRI, magnetic resonance imaging; ANA, antinuclear antibody; SRP, signal recognition particle; BPAg, bullous pemphigoid antigen; DEJ, dermoepidermal junction; DIF, direct immunofluorescence; IIF, indirect immunofluorescence; ESR, erythrocyte sedimentation rate.

<sup>a</sup>Most commonly associated malignancies appear in bold.

## Practice Questions

- 1. The most commonly used substrate for indirect immunofluorescence in diagnosing paraneoplastic pemphigus is:**
  - a. guinea pig esophagus
  - b. monkey bladder
  - c. monkey esophagus
  - d. rat bladder
  - e. rat esophagus
  
- 2. Which solid organ malignancy is most commonly associated with paraneoplastic Sweet syndrome in men?**
  - a. bladder
  - b. brain
  - c. gastrointestinal
  - d. lung
  - e. renal
  
- 3. A patient with necrolytic migratory erythema and glucagonoma syndrome can be expected to have the following abnormal laboratory results:**
  - a. elevated amino acid levels
  - b. elevated red blood cell count
  - c. elevated serum glucagon levels
  - d. elevated zinc level
  - e. low serum glucagon levels
  
- 4. Leser-Trélat sign has been reported to present with which of the following:**
  - a. acanthosis nigricans
  - b. acquired ichthyosis
  - c. severe pruritus
  - d. tripe palms
  - e. all of the above
  
- 5. Which of the following is most strongly associated with an increased risk for malignancy in dermatomyositis?**
  - a. anti-Mi2 autoantibody
  - b. anti-p155 autoantibody
  - c. elevated erythrocyte sedimentation rate
  - d. positive antinuclear antibody
  - e. positive double-stranded DNA

*Fact sheets and practice questions will be posted monthly. Answers are posted separately and require registration on [www.cutis.com](http://www.cutis.com).*