Photodermatoses: Diagnosis and Management

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Disclosure

• **Investigator:**
  – Estée Lauder
  – Ferndale
  – Allergan
Learning Objectives

• Recognize important and new development in photodermatoses
• Diagnose and manage patients with photodermatoses
Photodermatoses

- Polymorphous light eruption
- Actinic prurigo
- Hydroa vacciniforme
- Chronic actinic dermatitis
- Solar urticaria
- Drug-induced photosensitivity
- Erythropoietic protoporphyria

Photodermatoses

- Polymorphous light eruption
PMLE
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Kontos, A, Photodermatol Photoimmunol Photomed, 12/02; 18:303
PMLE

Kontos, A, Photodermatol Photoimmunol Photomed, 12/02; 18:303
Isedeh, P. J Drugs Dermatol. 2013
Juvenile Spring Eruption: a variant of PMLE

Chantron, Lim, Shwayder. JAAD 12/12; 67:1093 and 1113.

Courtesy of Tor Shwayder, MD
PMLE: Pathophysiology
(van de Pas, CB. JID 2/04; 122:295. London)

• PMLE patients are more resistant to UV-induced immunosuppression
PMLE: Treatment

- Seek shade
- Broad spectrum sunscreens
FDA new guidelines (12/12)

www.fda.gov/Drugs/Resources ForYou
PMLE: Treatment

- Seek shade
- Broad spectrum sunscreens
- NB-UVB
  - Start in spring
  - 50% MED-B, or 100-200 mJ/cm²
  - Increase by 5-10% per treatment
  - 2-3 x per week for 15 treatments
  - Weekly sun exposure (10-2PM); 20-30 min for rest of summer
PMLE: Treatment

- Seek shade
- Broad spectrum sunscreens
- NB-UVB
- Oral corticosteroid
  - Prednisone, 0.6-1 mg/kg/d
  - 5-7 days
PMLE: Treatment

- Seek shade
- Broad spectrum sunscreens
- NB-UVB
- Oral corticosteroid
- Hydroxychloroquine (200 mg bid)
- Azathioprine (2-2.5 mg/kg/d)
- ? Polypodium leucotomos (480-1200 mg/d)
Photodermatoses

- Polymorphous light eruption
- Actinic prurigo

Actinic Prurigo

• Familial (75%) American Indian
• Female: male = 3:1
• Photosensitivity usually to UVA
• Face, nose, ears
• Chelitis, conjunctivitis, photophobia
Actinic prurigo

Courtesy of Tor Shwayder, MD
Henry Ford Hospital, Detroit
Actinic Prurigo. 11 yo girl.
Greatgrandmother: Am Indian
Henry Ford Hosp., Detroit

Courtesy of Marta Valbuena, MD, Bogota, Colombia
Actinic Prurigo
Actinic Prurigo: Treatment

(Valbuena, M, Lim, HW. Clin Decision Support: Dermatology. 2012)

• Photoprotection
• Move to lower altitudes if possible
• Topical and oral corticosteroids
• NB-UVB, PUVA
• Thalidomide (100-200 mg/d)
• ? TNF-α inhibitor
Photodermatoses

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Hydroa Vacciniforme
(Chantron, R, Lim, HW, Shwayder, T. JAAD 12/12; 67:1093 and 1113. Detroit)

- Childhood onset
- Vesicles on sun exposed skin → scarring
- Improves in adolescence
- Rare: adult-onset; familial.
- ↓ MED-A, rarely ↓ MED-B
- Positive provocative test with UVA
- Tx: Hydroxychloroquine, UVB, PUVA
Hydroa Vacciniforme
Hydroa Vacciniforme

(Courtesy of Tor Shwayder, MD)
Hydroa Vacciniforme

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Hydroa Vacciniforme
Hydroa vacciniforme

Courtesy of Tor Shwayder, MD
Hydroa Vacciniforme
(G. Gupta, JAAD 2/2000; 42:208)

• Spontaneous clearing: 60% in 9 yrs (M: 11 yrs, F: 5 yrs)
• 53%: Abnormal response to UVA
• 40%: Positive provocative testing to UVA
• 3/5 pts: Benefited from NB-UVB
EBV-associated HV-like Eruption

(Ruiz-Maldonado, R, JAAD 1/95; 32:37; Mexico
Tanaka, C. BJD 2012 Jan;166(1):216-1. Japan
Sangueza, M, et al. JAAD 7/13; 69:112. Milwaukee and Bolivia

- Mexico, South America, Asia, Europe
- Edema of nose, lips, periorbital area; erythema, vesicles, ulceration, scars on sun-exposed and sun-protected areas
- Fever, wasting, hepatosplenomegaly, vasculitis, panniculitis
- May progress to lymphoma or leukemia
- Atypical lymphocytic infiltrate (CD8+) with angiotropism and angiocentricity.
EBV-associated HV-like Eruption
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Chronic Actinic Dermatitis

Diagnostic Criteria

• Chronic photodermatitis
• Low phototest results to UVA &/or to UVB &/or to visible light
• Histology:
  - dermal lymphohistiocytic infiltrate
  - +/- epidermal spongiosis
  - +/- atypical mononuclear cells
Chronic actinic dermatitis
Chronic actinic dermatitis
Chronic Actinic Dermatitis
Chronic Actinic Dermatitis
CAD: Natural History

(RS Dawe, Arch Derm 10/00. Dundee)

- 178 pts; 24 yr follow-up
- Probability of resolution
  - 10% (1 in 10) in 5 yrs
  - 20% (1 in 5) in 10 yrs
  - 50% (1 in 2) in 15 yrs
CAD: Treatment

- Sunscreen, sun protection
- Topical tacrolimus
- Mycophenolate mofetil
- Azathioprine
- Oral corticosteroids (for flares)
- Cyclosporine
- Hydroxychloroquine
- Hydroxyurea
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• Solar urticaria

Solar Urticaria

Driving x 1 hr (windshield, Solumbra shirt)
Solar Urticaria

Immediate

24 hrs
Solar Urticaria: Action Spectrum

- Visible
- UVA
- UVB
Solar Urticaria

(Beattie PE. Arch Dermatol 9/03; 139:1149. Dundee)

• Probability of resolution of those with SU alone:
  - 5 yrs: 15%
  - 10 yrs: 24%
  - 15 yrs: 46%
Solar Urticaria: Therapy

(Dawe RS, BJD 7/97; 137:144;
Calzavara-Pinton P. JAAD 2012 Jul;67(1):e5-9)

- Antihistamine
- **UVA (or UVA!)**
  - Start with 50-70% MUD, increase by 10-15%;
  - Exposed area only;
  - Daily for 15-20 tx;
  - 2-3 x wk for rest of sunny months
- **Cyclosporine** (3-5 mg/kg)
- **IVIg** (400 mg/kg/d x 5 d = 2 gm/kg)
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Voriconazole
(Cowen, EW. JAAD 1/10; 62:31. Bethesda, Durham, SF. Miller, DD. Arch Dermatol 3/10; 146:300. SF)

• Broad spectrum antifungal (Aspergillus, Candida, Fusarium, Scedosporium spp.)
• Inhibits cytochrome P450.
• Side effects: visual disturbances, ↑ LFTs, GI upset
• > 12 wks: photosensitivity (act spectr: UVA), pseudoporphyria, photoaging, lentigines, premature dermatohelioses
• > 12 months: Increased SCC, melanoma
22 yo man. SCC. Lentigines

22 yo man. Erythema. Lentigines

Cowen, EW. JAAD 1/10; 62:31
Vemurafenib
(Su, F. NEJM 1/19/12; 366:207. Nutley, NJ, USA)

- A selective inhibitor of oncogenic BRAF (BRAF V600E)
- Approved in US and Switzerland for metastatic melanoma
- Side effects: arthralgia, rash, fatigue, and photosensitivity
- 15-30% of patients develop SCC and keratoacanthoma
  - Associated with mutations in RAS
Vemurafenib – Phototesting

(Dummer, R. NEJM 2/2/12; 366:480-481. Zurich)

UVB (8-99 mJ/cm²)

UVA (10-40 J/cm²)

Action spectrum: UVA
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EPP
Afamelanotide for Erythropoietic Protoporphyria

Afamelanotide for EPP: EU & US


- Randomized, double-blind, placebo-controlled study
- EU (74 pts): 5 subcutaneous implants, every 60 days
- US (94 pts): 3 implants
- Duration of pain-free time following sun exposure was longer in the treatment gr.
- Improved quality of life
- Adverse effects: headache, nausea, nasopharyngitis, and back pain
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Take Home Message

• Key aspects of diagnosis include onset of eruption after sun exposure, morphology, and distribution of lesions.
• Phototesting is helpful.
• Management: photoprotection, hardening, immunosuppressants
Learning Objectives

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Pierre-Auguste Renoir
*Two Sisters.* 1881
Art Institute of Chicago