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AHC Media

The Photosensitive Patient

Why do patients with rashes come to the emergency department? I have asked myself this many times. Perhaps you have, too. For an acute rash that is especially itchy and keeps the patient from sleeping, relief is what the patient is seeking. That I understand. But for those rashes present for days to weeks, what makes it an emergency now? Those working the front lines know the answer: Many of our emergency department patients don't have a provider who can see them urgently, so they come to see us.

This issue discusses patients with photosensitive skin eruptions. This topic can be complex and confusing. Fortunately, principles useful for emergency physicians are simple: 1) recognize the rash is confined to sun-exposed areas; 2) know that drugs, underlying diseases, or genetic inheritance can make the patient photosensitive; and 3) begin initial treatment with medications to control symptoms.

— J. Stephan Stapczynski, MD, Editor

Figure 1. Erythematous, Annular Rash Over Arms, Neck, and Scalp

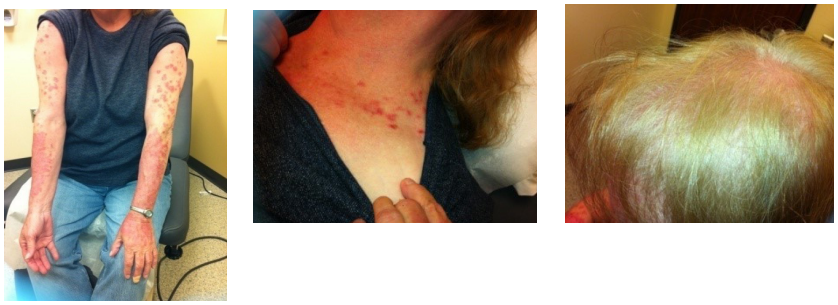
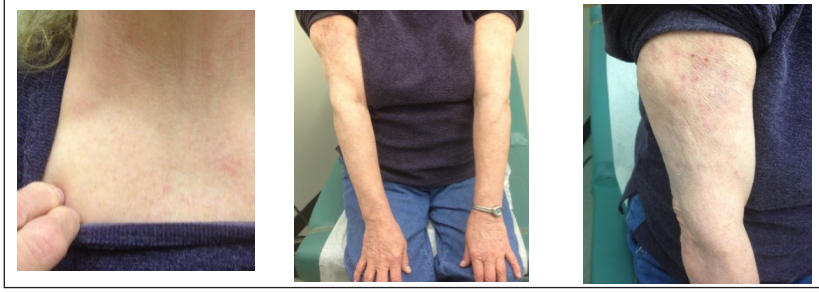


Figure 2. Rash Improvement Six Months Later



EXECUTIVE SUMMARY

- Suspect photosensitivity if the rash is confined to the classic sun-exposed areas of the dorsa of the arms and hands, the legs, the face, the ears, the posterior neck, and the anterior neck line, often "V" shaped.
- Photosensitivity is unlikely if the rash involves areas typically spared sun exposure, such as the skin folds of the neck, under the chin, nasolabial folds, behind the ears, and between the fingers.
- Reviewing current medications is useful, as many drugs can promote photosensitivity reactions.
- Two autoimmune disorders, lupus erythematosus and dermatomyositis, are associated with photo-exacerbated dermatoses.
- Initial treatment involves limiting sun exposure and using topical steroids to reduce inflammation.

Illustrative Case Presentation

A 65-year-old female presents to the emergency department with a chief complaint of a severe rash. The rash has been present for two weeks on her arms, neck, and scalp. It began while she was sitting on her porch one afternoon. She denies any associated fevers, chills, headache, or muscle ache. She denies any exposure to pets, new soaps, detergents, or lotions, and she denies recent travel.

Past medical history includes hypertension, type 2 diabetes mellitus, menorrhagia, hypercholesterolemia, and gout. Current medications include verapamil, progesterone, pravastatin, aspirin, lisinopril, allopurinol, metformin, and hydrochlorothiazide.

The review of systems is positive for arthralgia in the right lower extremity and back, as well as irregular menstrual bleeding. Social history is positive for remote tobacco use.

Her physical exam reveals a blood pressure of 142/90. Cutaneous examination reveals an erythematous, annular rash distributed over the extensor surfaces of the arms and forearms, the neck, and scalp. (*See Figure 1.*) Slight scale is also noted on the lesions. The trunk has a few scattered patches, but is otherwise clear.

The location of the rash on areas of potential sun exposure suggests a photosensitive reaction. An important step is assessing the patient's current medications, as many drugs can cause photosensitive reactions. Starting topical steroids is an appropriate response to this inflammatory rash.

On follow-up visit, several medications were held, including hydrochlorothiazide, allopurinol, metformin, and lisinopril. Topical steroids were continued and the patient was advised to avoid sunlight. Six months later, the rash had almost completely resolved with only two to three lesions on the right upper arm. (*See Figure 2.*) Post-inflammatory hyperpigmentation was present.

Photosensitive Disorders (Reactions)

Overview. Photosensitivity is an abnormal cutaneous reaction to visible or ultraviolet light. The photodermatoses often overlap clinically, and a broad understanding may help facilitate proper diagnosis, treatment, and/or referral. This discussion aims to address several of the different photodermatoses, their clinical entities, and a guide for evaluation of the photosensitive patient. They are described below based on the following six categories. (*See Table 1.*)

Pathophysiology

Ultraviolet radiation reaching the skin comes in three forms: UVA (320-400 nm), UVB (290-320 nm), and UVC (100-290 nm). In general, the longer the wavelength of light, the deeper the penetration into the skin and subcutaneous tissue.[1,2] Molecules in the skin called chromophores absorb this ultraviolet radiation.[2] DNA is one of these chromphores. The radiation energy absorbed is then passed on harmlessly or used to power unfavorable reactions, causing change anywhere on the spectrum of sunburn to carcinogenesis. Other molecules in the skin can absorb this energy, such as the protoporphyrins or typical photosensitizing medications (both ingested and topical), causing abnormal reactions known as the photodermatoses.[2] Not all individuals will experience an abnormal reaction to UV radiation.

Clinical Evaluation

There are some specific elements of history, such as age of onset, that can help to narrow down the differential when presented with a photosensitive patient. For example, chronic actinic dermatitis is mostly a disease of the elderly, whereas hydroa vacciniforme is found in adolescents.[3] Other history elements that are important to elicit include timing of lesions and duration of lesions once they appear. For example, the hives associated with solar urticaria come on within minutes of exposure to sunlight and will disappear within a few hours.[4] Family history and/or personal history of these lesions, or autoimmune or connective tissue disease should be included. Exposure to sunlight or other sources of light including tanning beds, seasonal variation, and effect of glass-filtered light on the skin are also important to elicit. Finally, inquiring about exposure to photosensitizers, including a review of medications, plant or food exposure, and topical products such as perfumes, lotions, and cosmetics, will complete a thorough evaluation of the patient's history.[1,3,5-8]

The physical examination should document the type of lesion seen and the distribution. Photodistribution tends to occur on the dorsa of the arms and hands, the legs, the face, the ears, the posterior neck, and anterior neck line, often "V" shaped. It may extend beyond these areas if exposed to scattered UV light from artificial sources or through lightly woven clothes and in the case of a severe reaction. Typical areas spared in a photosensitivity reaction include the skin folds of the neck, under the chin, nasolabial folds, submental area, behind the ears, and in the interdigital web spaces.[1,3,5-7]

Idiopathic Acquired Photodermatoses[1]

Polymorphous Light Eruption. Polymorphous light eruption (PMLE) is the most common idiopathic photodermatosis.[9] This chronic disease is one of many variations that may change over time, visually and clinically. PMLE is a disorder with seasonal exacerbation, mostly summertime. Although a number of different subtypes exist, patients typically develop the same type each year. A summary of the different subtypes is found in Table 2.[4] The lesions will typically start in a defined area, but will often spread beyond that area each subsequent summer. PMLE is associated with the phenomenon of hardening. Hardening is described as a decrease in light sensitivity with repeated sun exposure such that the eruption may diminish with each exposure. Therefore, the patient exposed to sunlight all year round will rarely be affected by an eruption. There is no age preference for PMLE.

The initial presentation of PMLE includes redness, itching, and burning. Systemic symptoms that may occur include headache, nausea, chills, and malaise. These generally subside within a couple hours of UV exposure. Diagnosis is often made by history, clinical presentation, and an otherwise negative workup. The differential includes atopic dermatitis if the papular subtype is presenting, and lupus in the plaque subtype. PMLE will be found in the typical photodistributed areas as described above, whereas atopic dermatitis presents in skin folds. PMLE will have a negative immunofluorescence staining on histology, whereas lupus will be positive for direct and indirect immunostaining.[4]

Treatment for acute eruptions includes topical steroids for a short period of time (3-14 days). Oral steroids can be used for patients experiencing extreme pruritus and extensive eruption. Treatment recommendations for these patients include avoidance of sunlight during greatest intensity (10 a.m.-2 p.m.) and proper protection with sunscreen. Phototherapy treatment directed by a dermatologist can be given in small dose increments for desensitization. Additional treatment using Psoralen+ UVA (PUVA) with two or three treatments a week for 1-3 months can induce remission. Antimalarial drugs can also be considered in recalcitrant disease for patients who are unresponsive to the above listed treatments and used only during summer months. Hydroxychloroquine can be dosed at 400 mg/day for the first month and 200 mg/day for subsequent months.[4] Hydroxychloroquine, if required, is not a benign medication and requires initial ophthalmologic examination and yearly monitoring due to ocular toxicity.

Table 1. Categories of Photosensitive Reactions[1,3,4,5-9]

Idiopathic Photodermatoses[1]
- Polymorphic light eruption
- Actinic prurigo
- Hydroa vacciniforme
- Hydroa aestivale
- Chronic actinic dermatitis
- Solar urticaria

Photodermatoses Secondary to Exogenous Agents[5]
- Phototoxicity
- Photoallergy

Photodermatoses Secondary to Endogenous Agents[3]
- Porphyria cutanea tarda
- Congenital erythropoietic porphyria
- Variegate porphyria
- Erythropoietic protoporphyria

Photoexacerbated Dermatoses[6]
- Lupus
- Dermatomyositis
- Pemphigus
- Bullous pemphigoid

Genodermatoses[7]
- Xeroderma pigmentosum
- Bloom syndrome
- Rothmund-Thomson syndrome
- Cockayne syndrome
- Kindler syndrome
- Trichothiodystrophy
- Hartnup disease
- Dyskeratosis congenita
- Oculocutaneous albinism

Nutritional Deficiency[8]
- Pellegra
- Pyridoxine deficiency

Actinic Prurigo (Hereditary PML Eruption). Actinic prurigo or hereditary PML eruption is mostly found within specific populations, namely the Inuit of North America and Native Americans of South America. Family history can help to identify this photodermopathy, as it is an autosomal dominant genetic disorder. The eruption appears most often on the face, but can be seen on all sun-exposed areas of the body from early spring to early fall. Associated symptoms and progression

Table 2. Clinical Subtypes of Polymorphous Light Eruption[4]

Subtype	Clinical Picture
Papular type	Most common form: Several small papules are found on a patchy erythematous base.
Plaque type	Second most common form: Plaques can range from smaller to larger coalescing types. They may be superficial or urticarial and at times can be eczematous.
Papulovesicular type	Begins as a plaque and progresses to groups of vesicles. It can be markedly itchy and almost always occurs in women.
Eczematous type	Almost always occurs in men; presents with papules, erythema, scale, and occasionally vesicles.
Erythema multiforme type	Targetoid lesions that typically begin as red macules.
Hemorrhagic type	Rare form that presents with purpura or hemorrhagic papules.
Source: 4, Chapter 19 pg.750-2	

Table 3. Common Medications Associated with Photoallergic Reactions

Topical sunscreens	Oxybenzone Cyclohexanol Benzophenones Salicylates Cinnamate Para-aminobenzoic acid (PABA): use has been eliminated due to high rate of allergic reactions
Topical antimicrobials	Chlorhexidine Hexachlorophene Dapsone
NSAIDs	Celecoxib Ketoprofen Etofenamate
Phenothiazines	Chlorpromazine Fluphenazine Perazine Thioridazine Perphenazine
Sulfonylureas	Glipizide Glyburide
Miscellaneous	Oral contraceptives Hydrochlorothiazides Itraconazole 5-fluorouracil

are associated with age of onset. Early onset (prior to age 21) actinic prurigo is associated with chelitis and conjunctivitis and more likely to improve over a five-year time span. Later onset (21 and older) is less likely to have associated symptoms but more likely to be persistent beyond five years. Clinical presentation and treatment follow that of PML.[4]

Juvenile Spring Eruption. Juvenile spring eruption occurs typically in adolescent or young adult males during the springtime. The lesions of this photodermopathy are found on the outer helices of the ear.[10] Initially, this individual presents with erythema, itching, and swelling of the helix, followed by progression to papules and vesicles that eventually crust over. The lesions heal with minimal to no scarring or dyschromia in 2-3 weeks.[10] This eruption tends to occur as small outbreaks within a localized population, such as military recruits, and may recur. Treatment is self-limiting, as these lesions resolve spontaneously.[10]

Hydroa Vacciniforme and Hydroa Aestivale. Hydroa vacciniforme (HV) and hydroa aestivale are rare photosensitive reactions, most commonly seen prior to puberty, most often around age 6. Itching and erythema begin to appear within the first two hours of exposure to sunlight. Hydroa aestivale presents as a papular eruption with occasional crust, most commonly in an acral, face, and ear distribution, with occasional involvement of non-sun-exposed areas such as the buttocks. The lesions of HV begin as papules that progress to vesicles on classic sun-exposed areas (ears, face, back of hands, and chest). The vesicles will umbilicate, become necrotic, and crust. The healed stage of the lesion can include scarring and hypopigmentation. It will typically clear after puberty. Patients should be educated on the use of sunscreen and avoidance of excessive sunlight. UVA light may exacerbate the disease, while UVB therapy can be effective for treatment. Other methods to control the disease include (Group V) topical steroids, cool compresses, and antimalarial medications.[4]

Solar Urticaria. Solar urticaria is defined as the development of hives within minutes of exposure to sun or

artificial light. The hives then disappear in an hour or less. Young adult females are more often affected. As a type of photoallergic reaction, this disorder may be mediated by an IgE-related mechanism. Each of the six different wavelengths of light corresponds to the six different types of this disease. Individuals may experience allergy to any one of the six different wavelengths of light, including waves in the visible light spectrum (greater than 400 nm). These individuals will experience hives even when exposed to light filtered through glass. Phototesting can help determine the specific wavelength responsible for the solar urticaria. Treatments include sunscreen, antihistamines, and gradual exposure to increasing amounts of light.[4]

Chronic Actinic Dermatitis. Chronic actinic dermatitis most often occurs in the elderly presenting as dermatitis on sun-exposed areas.[11] This photodermatosis can also commonly present in patients with increased susceptibility to hypersensitivity reactions to common exogenous/endogenous substances, triggering a concomitant contact dermatitis.[11] Patch testing may be necessary in order to avoid specific triggers. Treatment should include education on avoidance of sunlight and known allergens. Topical steroids are also appropriate.[11] If these measures are unsuccessful, oral steroids or immunosuppressants may be used as second line.[11]

Photosensitivity Reactions Caused by Exogenous Agents[5]

Photoallergy. A photoallergic reaction is an uncommon type IV hypersensitivity reaction that results in eczematous inflammatory eruption on sun-exposed areas. Ultraviolet light initiates a reaction between skin protein and a chemical or drug to form an antigen capable of inducing an immunologic response. Specifically, the photosensitizing agent creates unstable hapten free radicals, which covalently bind to macromolecules to form a complete antigen.[12] The antigen is then taken up by Langerhans cells and resident immune surveyor cells in the epidermis, and the inflammatory process ensues.[13] Only a small amount of the photosensitizing agent is needed to cause a photoallergic reaction. The patient is sensitized upon initial exposure to the agent, with a subsequent rash occurring after repeated exposures. The onset of duration is 24-72 hours after exposure. Similar to Rhus dermatitis, a photoallergic reaction manifests as pruritic, eczematous inflammation. Lichenoid eruptions have been reported, but the reaction usually resolves without significant post-inflammatory hyperpigmentation. The distribution of the skin findings is usually confined to sun-exposed areas but may involve unexposed areas. Some patients may experience a persistent light reaction. This is a continuous cycle of flares when exposed to sunlight without continued drug exposure.[4]

Pharmacologic agents known to cause photoallergic reactions are found in Table 3. In a retrospective review from 1993 to 2006, Victor et al[14] found that sunscreens and antimicrobial agents were the most frequent agents causing photoallergic reactions. Photoallergens, however, have changed over time: In the 1960s and 1970s, antimicrobial agents, such as salicylanilides, were prominent causes; in the 1970s and 1980s, fragrances such as musk ambrette and 6-methylcoumrain were prominent causes; and in the 1980s and 1990s, sunscreens and topical non-steroidal anti-inflammatory drugs (NSAIDs) were found to be causes.[14] Recent analysis has shown topical NSAIDs (ketoprofen, etofenamate) and sunscreens (octocrylene, benzophenone-3, butyl methoxydibenzoylmethane, and octyl dimethyl para-aminobenzoic acid [PABA]) to remain among the most common culprits in photoallergy.[15,16]

To confirm clinical diagnosis of photoallergy, photopatch testing can be done. In this test, known photosensitizing agents are applied to the skin and exposed to UVA. Eczematous inflammation at the site of a particular photosensitizing agent is considered positive. Management of a photoallergic reaction includes prompt identification and discontinuation of the photosensitizing agent. Sun exposure should be minimized. Cool, moist compresses, non-steroidal anti-inflammatory drugs, and topical glucocorticoids can provide symptomatic relief. Severe reactions may require systemic glucocorticoids. Immunosuppressive agents such as azathioprine, cyclophosphamide, cyclosporine, or mycophenolate mofetil may be used in patients with

Table 4. Medications Associated with Phototoxic Reactions

Medication Type	Medication
Anti-infectives	Fluoroquinolones: ciprofloxacin, levofloxacin Tetracyclines: doxycycline Sulfonamides: sulfamethoxazole Antimalarials: quinine, chloroquine, hydroxychloroquine
NSAIDs	Naproxen Piroxicam
Diabetic medications	Glyburide Chlorpropamide
Antihistamines	Diphenhydramine
Diuretics	Furosemide Hydrochlorothiazide
Topical medications	Isotretinoin Acitretin
Psychiatric	Phenothiazines: chlorpromazine Thioxanthenes: thiothixene Tricyclics: imipramine, desipramine

Table 5. Common Foods and Plants Associated with Phytophotodermatitis

Foods	Limes: especially Bergamot limes (*Citrus bergamia*) Celery (*Apium graveolens*) Wild carrot (*Daucus carota*) Figs (*Ficus carica*) Parsnips: both common parsnip (*Pastinaca sativa*) and cow parsnip (*Heracleum sphondylium*)
Plants	Queen Anne's lace or bishop's weed (*Ammi majus*) Giant hogweed or cartwheel flower (*Heracleum mantegazzianum*) Gas plant or burning bush of Moses (*Dictamnus albus*) Babchi or scurf pea (*Psoralea corylifolia*)

resistant light reaction who are unable to tolerate chronic systemic high-dose glucocorticoids.[12]

Phototoxicity

A phototoxic reaction is a non-immunologic cutaneous response to a topical or systemic agent after UV exposure. Unlike photoallergic reactions, phototoxic reactions have a higher incidence and can occur after a single exposure. If sufficient levels of a photosensitizing agent and UV light exposure occur, 100% of patients will experience a phototoxic reaction.[4] Many pathways are responsible for inducing phototoxic damage, and a single phototoxic agent may induce one or more of these pathways. The end result is a stable phototoxic product that induces an inflammatory response of the skin and apoptosis of host cells.[13] A large amount of agent is generally required to induce a phototoxic reaction. The onset of a phototoxic reaction is usually within minutes to hours of exposure, and the distribution is confined to only sun-exposed areas.[4] Pharmacologic agents known to cause phototoxic reactions are included in Table 4.

Phototoxic symptoms are dose- and UV-dependent, ranging from asymptomatic to an erythematous/edematous photodistributed rash with occasional vesicles and bullae, resembling an exaggerated sunburn. The areas may heal with post-inflammatory hyperpigmentation, which can take a year or longer to resolve.[13] Linear streaks are characteristic of topical exposure, in which the offending agent is drawn along the skin.

Phytophotodermatitis is caused by exposure to plants that contain light-sensitizing compounds. Common agents responsible for this type of phototoxic reaction are included in Table 5. These agents contain light-sensitizing compounds such as furocoumarin (psoralens: 8-MOP, 5-MOP) and can produce intense reactions.[4]

Diagnosis is often clinical. A photopatch test can be done to attempt to identify the photosensitizing agent.[12] Management of phototoxic reactions mirrors that of photoallergic reactions. In the most severe cases, systemic steroids can be used.

Photosensitivity Reactions Caused by Endogenous Agents[3]

Porphyria is a group of diseases caused by an accumulation of byproducts from the heme synthesis pathway. Each of the porphyrias is due to a specific enzymatic defect. Table 6 describes the specific enzymatic defects and clinical presentation associated with the most prevalent photosensitive porphyrias.

Testing for the porphyrias involves urine, fecal, and plasma porphyrins. The specific enzyme deficiency suspected should also be evaluated.[4,17] Treatment should include avoidance of sun exposure and education on protective sunscreens. Other treatment is specific to each porphyria and may include phlebotomy, chloroquine, and symptomatic therapies.[4,17] Referral to dermatology and gastroenterology is recommended.

Photo-exacerbated Dermatoses[6]

Lupus Erythematosus. Lupus erythematosus (LE) is a group of heterogeneous disorders in which the affected person develops autoantibodies to nucleic acids and their associated proteins, resulting in multisystem disease. LE is classified on a continuum with systemic manifestations in the acute LE subtype and predominantly cutaneous manifestations in chronic cutaneous LE subtype.[18] Women in their childbearing years account for more than 80% of cases of LE.[4] In 1997, the American College of Rheumatology defined 11 revised criteria for classification of systemic lupus erythematosus (SLE). The criteria can be found in Table 7. A person is defined as having SLE if any 4 or more of the 11 criteria are present.[19] Main types of cutaneous lupus erythematosus include discoid lupus erythematosus (DLE), subacute cutaneous lupus erythematosus (SCLE), and acute cutaneous lupus erythematosus (ACLE).

ACLE can be localized or generalized. When localized, it manifests as the classic butterfly or malar rash on the face, in which symmetric erythema and edema are present over the bridge of the nose and the malar eminences, with sparing of the nasolabial folds. Generalized ACLE can present as morbilliform or exanthematous eruptions on the extensor aspects of the arms and hands, but spares the knuckles.[18] Multisystem disease is usually present with ACLE.[4] This type of LE can take hours, days, or weeks to resolve, and scarring typically does not occur.[18] Common laboratory findings include positive anti-nuclear antibody (ANA), anti-double stranded DNA (anti-dsDNA), as well as others.

SCLE runs a milder course, with erythematous macules and/or papules developing into hyperkeratotic papulosquamous or annular plaques. These lesions occur in sun-exposed areas, such as the neck, shoulders, upper extremities, and trunk, but the malar region of the face is often spared. Anti-Ro/Sjögrens syndrome A (SS-A) antibodies

Table 6. Summary of Most Common Photosensitive Porphyrias[4]

Type of Porphyria	Mode of Inheritance	Enzymatic Defect and Confirmative Testing	Dermatologic Presentation	Other Clinical Symptoms
Porphyria cutanea tarda	Autosomal dominant	Uroporphyrinogen decarboxylase • different subtypes exist, recommend testing for urine, fecal, and plasma porphyrins, as well as uroporphyrinogen decarboxylase in RBCs	Bullae on sun-exposed skin that rupture, ulcerate, and heal with scarring and dyspigmentation. Hypertrichosis of the face. Neck and back may show thickened sclerotic skin.	Most common type. Liver disease may be present. Patients may have history of alcoholism, and all patients should be screened for hepatitis C.
Congenital erythropoietic porphyria	Autosomal recessive	Uroporphyrinogen III cosynthase • increased plasma uroporphyrin, coproporphyrin, and protoporphyrin with decreased uroporphyrinogen III co-synthase	Extreme burning with sun exposure causing the child to cry. Erythema, edema, and blistering will occur in sun-exposed areas. Hypertrichosis of the cheeks.	Found in neonates. Urine in diapers may be pink to red to purple in color. Erythrodontia. Growth retardation, anemia, bone fractures, gallstones.
Variegate porphyria	Autosomal dominant	Protoporphyrinogen oxidase • increased urine porphyrins • increased fecal coproporphyrin III/I ration (< 10) and protophorphyrin	Bullae and vesicles w/ erosion. Temporal hypertrichosis.	Common in South African ancestry. Abdominal symptoms such as colic, nausea, vomiting, diarrhea, constipation.
Erythropoietic protoporphyria	Autosomal dominant and autosomal recessive forms	Ferrochelatase • Increased free protoporphyrin in the plasma	Immediate burning of skin with sun exposure. Erythema, edema, and wheals can be seen. Skin becomes leathery with repeated exposures.	Elevated transaminases, gallstones. Cirrhosis is possible.
(4; Ch. 19 pg.754-760., 24, Ch. 26)				

support, but are not required, for diagnosis. SCLE presents with telangiectasias and dyspigmentation without follicular involvement and does not typically cause scarring upon resolution in most individuals.[18] Common lab findings include positive antinuclear and anticytoplasmic antibodies. Anti-dsDNA may be found in 30% of patients.[4]

Classic DLE presents as red-purple macules, papules, or plaques and quickly develops hyperkeratosis. These lesions are sharply demarcated, coin-shaped, erythematous plaques covered by an adherent scale.[18] This scale is described as "carpet tack" and is created by keratin plugs penetrating the hair follicle.[4] These plaques leave behind characteristic atrophic central scarring, telangiectasia, and hypopigmentation. DLE causes scarring alopecia when plaques are present on hair-bearing skin. Lesions of DLE are most commonly present on the face, scalp, ears, neck, and extensor aspects of the arms, with the most common site being the conchal bowl. DLE is referred to as localized when lesions occur only on the head and/or neck and generalized when lesions occur both above and below the neck.[18] Ninety-five percent of cases are limited to the skin. Patients typically present with an elevated erythrocyte sedimentation rate (ESR), +/- ANA, and leukopenia.

Drug-induced lupus can take on all three forms discussed above. Symptoms occur more commonly after months to years of being on the medications.[4] It most often effects the older population. Drug-induced SLE (DI-SLE) usually present with milder symptoms of acute SLE. Arthralgia, myalgia, pleurisy, and pericarditis may be present.[4] Important laboratory findings in DI-SLE include a positive ANA and anti-histone antibodies; p-ANCA has also been described. A positive antibody to single-stranded DNA is fairly specific. Drug-induced SCLE (DI-SCLE) presents with a rash in the typical photo-distributed areas and comes in two variants: papulosquamous and annular polycyclic.[4] Important laboratory findings associated with DI-SCLE include a positive ANA, positive SS-A antibody, and +/- anti-histone. Drug-induced discoid or chronic lupus is rare. All variants are treated with cessation of the causative medication. Table 8 summarizes common medications associated with these conditions.

Diagnosis of LE includes clinical and laboratory evaluation. Treatment for all forms of LE should involve patient education about sun protection. Photosensitizing drugs should be avoided. The first-line treatment for all forms of cutaneous LE is topical steroids. Intralesional corticosteroids can be used for DLE lesions that are resistant to topical corticosteroids. Antimalarial agents, such as hydrochloroquine, are a mainstay of treatment due to its safety and effectiveness in all forms of cutaneous LE. Oral corticosteroids are occasionally utilized for

Table 7. American College of Rheumatology Criteria for Diagnosis of SLE

1. Malar rash
2. Discoid rash
3. Photosensitivity
4. Oral ulcers
5. Non-erosive arthritis
6. Pleuritis or pericarditis (include photosensitivity here)
7. Renal disorder (persistent proteinuria or cellular casts) (proteinuria > 0.5 grams)
8. Neurologic disorder (seizures or psychosis)
9. Hematologic disorder (hemolytic anemia, leukopenia, lymphopenia, or thrombocytopenia)
10. Immunologic disorder (anti-DNA antibody, anti-Sm antibody, or positive finding of antiphospholipid antibodies)
11. Positive antinuclear antibody (ANA)

Table 8. Medications Associated with Drug-induced Lupus[4]

Drug-induced ACLE

- Procainamide
- Isoniazid
- Methyldopa
- Hydralazine
- Chlorpromazine
- Quinidine
- Anticonvulsants
- Beta-blockers
- Penicillamine
- Sulfasalazine
- Lithium
- OCPs
- Estrogens
- Gold salts
- Penicillin
- Phenylbutazone
- Minocycline
- Valproate
- Interferon
- IL-2
- Interferon-alpha
- Paclitaxel
- COL-3 (anti-angiogenesis)
- Sulfonamides

Drug-induced SCLE

- Hydrochlorothiazide
- Calcium channel blockers
- Nitrendipine
- Clizapril
- Acebutolol
- Efalizumab
- Adalimumab
- Terbinafine
- Griseofulvin
- Piroxicam
- Naproxen
- Bupropion
- Lansoprazole
- Pantoprazole
- Ticlopidine
- Statins
- Tamoxifen
- Interferon beta
- Leflunomide
- Docetaxel
- Glyburide
- NSAIDs
- Spironolactone
- Beta blockers

Drug-induced DLE

- Fluorouracil agents (tegafur, uracil tegafur)
- TNF-α antagonists (infliximab, etanercept)
- NSAIDs

patients who do not respond to topical corticosteroids.[4] Referral to a rheumatologist is warranted whenever LE is suspected.

Dermatomyositis. Dermatomyositis (DM) is an autoimmune inflammatory disease associated with cutaneous and musculoskeletal manifestations. Polymyositis (PM) is a similar disease process in which cutaneous findings are absent. Conversely, amyopathic dermatomyositis are patients with cutaneous findings only, patients with subsequent muscle findings, or those with myositis demonstrated on biopsy or electromyography with normal muscle enzymes. The distribution of DM is bimodal, with the highest incidence in children and adults older than 40 years of age. Adult DM is a multisystem disease with the following proposed diagnostic criteria: proximal symmetric muscle weakness, compatible muscle biopsy, compatible dermatologic features, myopathy or inflammatory myositis, elevated skeletal muscle enzymes (CPK, aldolase, SGOT), and exclusion of other disorders causing myopathy. The diagnosis is possible when one criterion is met, probable when two criteria are met, and definite when three or four criteria are met, given that the patient has compatible dermatologic features. The most common presenting feature is proximal muscle weakness.[4]

Cutaneous manifestations are an initial presenting sign in about 40% of patients with DM. Cutaneous manifestations of DM include a violaceous erythema of the eyelids, known as a heliotrope rash. A pathognomonic sign of DM is Gottron's papules, which are smooth, violaceous-to-red, flat-topped papules occurring over bony prominences such as the knuckles and the sides of the fingers. Gottron's papules are present in about 60% to 80% of patients with DM at some point during the disease. Periungal erythema, telangiectasia of the proximal nail, and ragged cuticles (Samitz sign) can also be seen. Another cutaneous finding may be localized or generalized violaceous erythema with or without scaling. When distributed in specific areas, it is recognized as the V-sign (anterior neck), shawl sign (located over the back, shoulders), or on the lateral thighs

(holster sign). These lesions generally involve the knuckles and spare the skin over the phalanges, which is opposite of the findings in SLE. Poikiloderma atrophicans vasculare is a characteristic violaceous erythema with telangiectasia, dyschromia, and possible atrophy, most commonly over V-neck area and trunk.

DM myopathy involves symmetrical proximal muscle groups with difficulty during activities such as rising from a chair or climbing stairs. Additionally, DM is associated with malignancy, most commonly ovarian, but others may also occur. Malignancy may precede, occur simultaneously, or follow the diagnosis of DM. Some pharmacologic agents are known to cause DM, including most commonly: statins, hydroxyurea, NSAIDs, quinidine, D-penicillamine, and tumor necrosis factor (TNF) antagonists.[4] Overlap syndromes refer to the concomitant presence of inflammatory myopathies with connective tissue disorders. Dermatomyositis may overlap with systemic sclerosis or mixed connective tissue disease.[20]

The diagnosis of DM includes skin biopsy, tricep muscle biopsy with serum muscles enzymes including transaminases, creatine kinase, aldolase, lactate dehydrogenase, and myoglobin. Creatine kinase is the most sensitive enzyme for DM, which can be elevated as much as fifty-fold.[20] Creatine kinase and lactate dehydrogenase are used to follow response to therapy. Additionally, electromyography (EMG) or magnetic resonance imaging (MRI) can be performed to assess muscle activity. A comprehensive examination including chest X-ray or CT scan, pulmonary function studies, pulmonary diffusion studies, esophageal motility studies, occult malignancy evaluation, and electrocardiogram can assess the degree of systemic involvement. Female DM patients should be followed at 6 and 12-month intervals for the first two years following diagnosis, as the incidence of ovarian malignancy increases.[4] Treatment involves oral corticosteroids, with the use of immunosuppressive drugs in patients who do not respond to oral corticosteroids alone. Approximately 25% of patients will be unresponsive or experience side effects to systemic corticosteroid treatment. Steroid-sparing agents may be effective in these patients, including the use of methotrexate, mycophenolate mofetil, azathioprine, cyclophosphamide, chlorambucil, or cyclosporine. Skin lesions can occasionally be treated with anti-malarials, such a hydroxychloroquine. All patients with DM should begin physical therapy in an attempt to prevent joint contractures and muscle atrophy. Cutaneous manifestations should be treated with topical steroids. All patients should be educated on sun protection.[4] Patients with malignancy-associated DM will be less responsive to steroid treatment.

Pemphigus. Pemphigus is a group of autoimmune blistering disorders. Autoimmune attack of desmoglein, a desmosomal structural protein, causes weakened keratinocyte adhesion in the epidermis, triggering bullae formation. Clinically, the patient presents with flaccid blisters that easily rupture, ulcerate, and eventually crust over. Lesions will heal with hyperpigmentation and usually without scarring. There are several variants of pemphigus that are beyond the scope of this paper. Ultraviolet radiation is very harmful to these patients and should be avoided, as it is known to induce and exacerbate the skin lesions of pemhigus. Referral to dermatology is necessary for ongoing treatment and monitoring.

Bullous Pemphigoid. Bullous pemphigoid (BP) is an entity not caused by photosensitivity but exacerbated by ultraviolet radiation. BP is an autoimmune disease, mostly of the elderly, most commonly induced by medications. BP presents with large blistering lesions that begin as erythematous, pruritic, sometimes urticarial plaques. The plaques will swell and progress from target type lesions to the vesicles and bullae typical of the disease. Unlike the bullae of pemphigus, these are tense and not easily ruptured. Treatment should include avoiding triggers such as trauma, scratching, and UV light. Topical steroids are appropriate for all stages of the disease and have been shown to be superior to oral steroid therapy in extensive disease.[4] Tetracycline antibiotics, erythromycin, and nicotinamide have been shown to give excellent results for this maintenance of BP. Dapsone, oral steroids, methotrexate, mycophenolate mofetil, and azathioprine, among others, may be used in patients unresponsive to initial treatment.[4] Referral to dermatology is warranted for continued monitoring and ongoing treatment.

Genodermatoses[7]

Genodermatoses are dermatologic disorders that have a specific underlying genetic defect. A thorough family history is paramount in making these diagnoses and, although rare, most likely will initially present to the primary care provider.

Photosensitive Nutritional Deficiencies[8]

Pellagra. Pellagra is a disorder characterized by dermatitis, diarrhea, dementia, and death. These manifestations are caused by niacin deficiency (vitamin B3), an important cofactor in multiple reactions, including oxidation-reduction and ceramide biosynthesis, which explains its characteristic systemic effects. Niacin deficiency can be caused by dietary factors (alcoholism, malabsorption, etc.), drugs (isoniazid, 5-fluorouracil, azathioprine), and systemic conditions with increased tryptophan metabolism such as in carcinoid syndrome. Classic cutaneous findings include symmetric erythema in sun-exposed areas with pruritic patches involving the upper chest and neck in a "V" pattern referred to as "Casal's necklace." Treatment is with oral nicotinamide replacement.[21,22]

Pyridoxine Deficiency. Pyridoxine (vitamin B6) deficiency can be found in patients with systemic comorbidities including cirrhosis, uremia, and celiac disease, among other causes. Another common cause of pyridoxine deficiency can be drug-induced by such medications like isoniazid, penicillamine, hydralazine, oral contraceptives, phenelzine, and cycloserine. Cutaneous manifestations of pyridoxine deficiency resemble seborrheic dermatitis with associated glossitis, stomatitis, conjunctivitis, and inflammation accentuated in the body folds.[17,23] In addition, patients can have hematologic abnormalities, anorexia, and peripheral neuropathy.[23] Treatment is replacement of the vitamin with recommended dosing at 50 mg/day.[23]

Conclusion

Overall, many disease entities and/or causative factors can induce photosensitive changes in our patients. It is important to recognize the photosensitive patient and elicit the proper history, including onset and duration of symptoms, exacerbating and alleviating factors, time of year, exposure to photosensitizing agents, and a complete medication list. Past medical history and family history are also important in evaluating the photosensitive patient. This paper serves to offer a reliable resource for the emergency department physician when addressing the photosensitive patient and help expedite appropriate diagnosis and treatment. Some take home points include reducing sun exposure and proper sun protection in photosensitive patients until the proper diagnosis is achieved. Topical steroids can be used to calm the inflammatory rash until that time. If complicated or recalcitrant cases arise, referral to dermatology or other appropriate specialists is a reasonable approach to care.

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CME Questions

1. Which of the following symptoms is the most common complaint in systemic lupus erythematosus?
   A. mouth sores
   B. rash
   C. joint pain
   D. seizures

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CME Objectives

Upon completion of this educational activity, participants should be able to:

- recognize specific conditions in patients presenting to the emergency department;
- apply state-of-the-art diagnostic and therapeutic techniques to patients with the particular medical problems discussed in the publication;
- discuss the differential diagnosis of the particular medical problems discussed in the publication;
- explain both the likely and rare complications that may be associated with the particular medical problems discussed in the publication.

2. A 23-year-old male presents to the ED complaining of a rash on the backs of his arms and hands. There are several bullae noted on the extensor surfaces of both hands and forearms. There are also ulcerated and scabbing lesions present. Some have healed, showing scarring and pigment changes. Hypertrichosis of the face is noted. Past medical history is positive for hepatitis C. Which of the following corresponds to the mode of inheritance of this disease?
   A. autosomal dominant
   B. autosomal recessive
   C. not inherited
   D. random DNA defect
3. A 65-year-old male presents complaining of diarrhea and forgetfulness. He has a known history of alcohol abuse. On exam, an erythematous glossy rash is present on the backs of both hands. What is the most likely diagnosis?
   A. polymorphous light eruption
   B. porphyria cutanea tarda
   C. pellagra
   D. pyridoxine deficiency
4. A 30-year-old female presents with the chief complaint of hives. She states that the hives come on while driving in her car and are located only on her left arm. She states that the hives resolve within 60 minutes of exiting her car and do not occur every day. What is the most likely diagnosis?
   A. polymorphous light eruption
   B. solar urticaria
   C. photoallergy
   D. phototoxicity
5. Which category of UV radiation penetrates deeper into the skin and subcutaneous tissue?
   A. UVA (wavelength 320 to 400 nm)
   B. UVB (wavelength 290 to 320 nm)
   C. UVC (wavelength 100 to 190 nm)
   D. They all penetrate equally.
6. Which of the following body parts is *not* considered a classic sun-exposed area?
   A. back of the hands
   B. lower legs
   C. nasolabial folds
   D. anterior neck
7. Which of the following is most characteristic of a polymorphous light eruption?
   A. Patients exposed to sunlight year round can have a persistent eruption.
   B. The eruption typically exacerbates in the summer.
   C. Eruptions are always papular.
   D. Systemic symptoms are absent.
8. Which of the following statements is *not* true of solar urticaria?
   A. The hives develop within minutes of sun exposure.
   B. Younger males are more affected than younger females.
   C. Patients may develop hives in response to a specific wavelength of UV or visible light.
   D. Patients may develop hives when exposed to light filtered through glass.
9. Currently, the substances most commonly implicated as photoallergens are:
   A. topical NSAIDs and sunscreens
   B. perfumes
   C. antibiotics
   D. neuroleptics
10. Which of the following is *not* characteristic of a phototoxic reaction?
   A. It is a non-immunologic reaction.
   B. The incidence increases with amount of the photosensitizing agent and UV light exposure.
   C. It is less common than a photoallergic reaction.
   D. It occurs within minutes to a few hours of UV exposure.

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The Photosensitive Patient

Categories of Photosensitive Reactions

Idiopathic Photodermatoses[1]
- Polymorphic light eruption
- Actinic prurigo
- Hydroa vacciniforme
- Hydroa aestivale
- Chronic actinic dermatitis
- Solar urticaria

Photodermatoses Secondary to Exogenous Agents[5]
- Phototoxicity
- Photoallergy

Photodermatoses Secondary to Endogenous Agents[3]
- Porphyria cutanea tarda
- Congenital erythropoietic porphyria
- Variegate porphyria
- Erythropoietic protoporphyria

Photoexacerbated Dermatoses[6]
- Lupus
- Dermatomyositis
- Pemphigus
- Bullous pemphigoid

Genodermatoses[7]
- Xeroderma pigmentosum
- Bloom syndrome
- Rothmund-Thomson syndrome
- Cockayne syndrome
- Kindler syndrome
- Trichothiodystrophy
- Hartnup disease
- Dyskeratosis congenita
- Oculocutaneous albinism

Nutritional Deficiency[8]
- Pellegra
- Pyridoxine deficiency

Clinical Subtypes of Polymorphous Light Eruption

Subtype	Clinical Picture
Papular type	Most common form: Several small papules are found on a patchy erythematous base.
Plaque type	Second most common form: Plaques can range from smaller to larger coalescing types. They may be superficial or urticarial and at times can be eczematous.
Papulovesicular type	Begins as a plaque and progresses to groups of vesicles. It can be markedly itchy and almost always occurs in women.
Eczematous type	Almost always occurs in men; presents with papules, erythema, scale, and occasionally vesicles.
Erythema multiforme type	Targetoid lesions that typically begin as red macules.
Hemorrhagic type	Rare form that presents with purpura or hemorrhagic papules.
Source: 4, Chapter 19 pg.750-2	

Common Foods and Plants Associated with Phytophotodermatitis

Foods	Limes: especially Bergamot limes (*Citrus bergamia*) Celery (*Apium graveolens*) Wild carrot (*Daucus carota*) Figs (*Ficus carica*) Parsnips: both common parsnip (*Pastinaca sativa*) and cow parsnip (*Heracleum sphondylium*)
Plants	Queen Anne's lace or bishop's weed (*Ammi majus*) Giant hogweed or cartwheel flower (*Heracleum mantegazzianum*) Gas plant or burning bush of Moses (*Dictamnus albus*) Babchi or scurf pea (*Psoralea corylifolia*)

Common Medications Associated with Photoallergic Reactions

Topical sunscreens	Oxybenzone Cyclohexanol Benzophenones Salicylates Cinnamate Para-aminobenzoic acid (PABA): use has been eliminated due to high rate of allergic reactions
Topical antimicrobials	Chlorhexidine Hexachlorophene Dapsone
NSAIDs	Celecoxib Ketoprofen Etofenamate
Phenothiazines	Chlorpromazine Fluphenazine Perazine Thioridazine Perphenazine
Sulfonylureas	Glipizide Glyburide
Miscellaneous	Oral contraceptives Hydrochlorothiazides Itraconazole 5-fluorouracil

Medications Associated with Phototoxic Reactions

Medication Type	Medication
Anti-infectives	Fluoroquinolones: ciprofloxacin, levofloxacin Tetracyclines: doxycycline Sulfonamides: sulfamethoxazole Antimalarials: quinine, chloroquine, hydroxychloroquine
NSAIDs	Naproxen Piroxicam
Diabetic medications	Glyburide Chlorpropamide
Antihistamines	Diphenhydramine
Diuretics	Furosemide Hydrochlorothiazide
Topical medications	Isotretinoin Acitretin
Psychiatric	Phenothiazines: chlorpromazine Thioxanthenes: thiothixene Tricyclics: imipramine, desipramine

Summary of Most Common Photosensitive Porphyrias

Type of Porphyria	Mode of Inheritance	Enzymatic Defect and Confirmative Testing	Dermatologic Presentation	Other Clinical Symptoms
Porphyria cutanea tarda	Autosomal dominant	Uroporphyrinogen decarboxylase • different subtypes exist, recommend testing for urine, fecal, and plasma porphyrins, as well as uroporphyrinogen decarboxylase in RBCs	Bullae on sun-exposed skin that rupture, ulcerate, and heal with scarring and dyspigmentation. Hypertrichosis of the face. Neck and back may show thickened sclerotic skin.	Most common type. Liver disease may be present. Patients may have history of alcoholism, and all patients should be screened for hepatitis C.
Congenital erythropoietic porphyria	Autosomal recessive	Uroporphyrinogen III cosynthase • increased plasma uroporphyrin, coproporphyrin, and protoporphyrin with decreased uroporphyrinogen III co-synthase	Extreme burning with sun exposure causing the child to cry. Erythema, edema, and blistering will occur in sun-exposed areas. Hypertrichosis of the cheeks.	Found in neonates. Urine in diapers may be pink to red to purple in color. Erythrodontia. Growth retardation, anemia, bone fractures, gallstones.
Variegate porphyria	Autosomal dominant	Protoporphyrinogen oxidase • increased urine porphyrins • increased fecal coproporphyrin III/I ration (< 10) and protophorphyrin	Bullae and vesicles w/ erosion. Temporal hypertrichosis.	Common in South African ancestry. Abdominal symptoms such as colic, nausea, vomiting, diarrhea, constipation.
Erythropoietic protoporphyria	Autosomal dominant and autosomal recessive forms	Ferrochelatase • Increased free protoporphyrin in the plasma	Immediate burning of skin with sun exposure. Erythema, edema, and wheals can be seen. Skin becomes leathery with repeated exposures.	Elevated transaminases, gallstones. Cirrhosis is possible.
(4; Ch. 19 pg.754-760., 24, Ch. 26)				

American College of Rheumatology Criteria for Diagnosis of SLE

1. Malar rash
2. Discoid rash
3. Photosensitivity
4. Oral ulcers
5. Non-erosive arthritis
6. Pleuritis or pericarditis (include photosensitivity here)
7. Renal disorder (persistent proteinuria or cellular casts) (proteinuria > 0.5 grams)
8. Neurologic disorder (seizures or psychosis)
9. Hematologic disorder (hemolytic anemia, leukopenia, lymphopenia, or thrombocytopenia)
10. Immunologic disorder (anti-DNA antibody, anti-Sm antibody, or positive finding of antiphospholipid antibodies)
11. Positive antinuclear antibody (ANA)

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