Photosensitivity in children: An approach to diagnosis and management

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ABSTRACT

Photosensitivity disorders in children include a wide array of conditions, many of which are unique to this age group. Prompt diagnosis of these disorders becomes difficult at times because of the overlapping clinical pictures. Genodermatoses and metabolic disorders may have associated systemic involvement, which may lead to these children presenting to pediatricians who may overlook the photosensitivity. A dermatologist’s consultation is essential in such cases for specific instructions and counseling of parents regarding photoprotection in these children. This intervention may improve the quality of life by reducing the morbidity and chances of early mortality. This review includes a comprehensive discussion of the distinguishing clinical features of childhood photodermatoses along with general guidelines regarding their investigation and treatment.

Key Words: Photosensitivity, Genodermatoses, Idiopathic photodermatoses

INTRODUCTION

Photosensitivity is defined as an abnormal response to “ordinary” light exposure.[1] The prevalent skin types in India are Fitzpatrick skin types IV and V. The incidence of photosensitivity disorders in the pediatric age group is much lower than in adults. In contrast to adults, the bulk of photosensitive children are included under the group of metabolic and genetic disorders[1] and pose a diagnostic challenge to the treating physician. The causes of photosensitivity in children are summarized in Table 1.

DIAGNOSTIC APPROACH

Elicitation of adequate history is vital. A systematic approach with a detailed history of the age of onset, the chronological order of appearance of symptoms associated with clinical examination and investigations are helpful in such cases.

HISTORY

What was the age of onset?
The age of onset of photosensitivity and related skin lesions helps in diagnosing different disorders (Table 2). Most of the genodermatoses and a few rare types of porphyrias manifest during infancy. Idiopathic photodermatoses usually affect older children. Solar urticaria (SU) is uncommon in children and affects adolescents or adults.

Is there any seasonal variation?
Patients with Hartnup disease show an exacerbation...
Lesions of polymorphous light eruption (PLE) and SU also aggravate in the spring and early summer but gradually improve through the rest of the summer (hardening effect). Actinic prurigo worsens in summer and persists through winter. Hydroa vacciniforme (HV), a very rare condition, occurs mostly during summer. What is the course of the disease? An acute single episode disease is indicative of idiopathic photodermatoses like PLE, SU, actinic prurigo (AP) and HV may also have an intermittent course with complete clearing between episodes. A chronic course with acute exacerbations and a tendency for skin changes to persist between exacerbations is suggestive of genodermatoses and metabolic disorders. Conditions like collagen vascular disorders are aggravated by exposure to light. In patients with Hartnup disease the photosensitivity and the dermatitis become milder with age. HV resolves or improves in adolescence. Are any other family members affected? A family history of photosensitivity is available in cases of photodermatoses with a genetic basis. A family history is positive in 50% cases of actinic prurigo. Clinical evidence of any collagen vascular disease is seen in nearly 40% of mothers of children with neonatal lupus erythematosus (LE). Sixty per cent of them suffer from Sjögren’s syndrome, systemic lupus erythematosus (SLE) or rheumatoid arthritis. However, the mother’s disease is subclinical in 60% cases at the time of childbirth. If such cases are followed up, a majority of these mothers develop collagen vascular disorders subsequently. Multiple affected family members are seen in albinism and phenylketonuria (PKU). Is there any history suggestive of systemic involvement? Idiopathic photodermatoses like PLE, SU, AP and HV usually present with only cutaneous lesions. Occasionally, patients with SU may develop systemic features like headache, nausea, bronchospasm and syncope if exposed inadvertently to sunlight for a prolonged period. Various acute or chronic systemic features involving different organ systems accompany the photosensitivity in genodermatoses, and in metabolic and nutritional disorders. Is there any history of exposure to photosensitizing agents? Exposure to photosensitisers is unusual in very young children. Citrus fruits, mango and extracts of a few common weeds containing furocoumarins are common sensitizing agents in school-going children. Lime juice (which contains ten times the oil of bergamot as other citrus fruits) is the commonest offending agent. Handling flowers of the Compositae group of plants (e.g.
chrysanthemum, marigold, dahlia and sunflower), which contain oleoresins, may give rise to photosensitivity. Photoallergic reactions are relatively rare in children. However, several therapeutic agents may give rise to both phototoxic and photoallergic reactions. Topical antimicrobials included in soaps, cosmetics and medicaments like halogenated salicylanilides, clioquinol and sulfonamide derivatives are common photosensitizers and may go unnoticed. Some systemically administered drugs such as sulfonamides, nalidixic acid, chlorpromazine, ceftazidime, griseofulvin, ibuprofen, furosemide and dapsone can cause photosensitivity.

CLINICAL FEATURES

Intolerance to sunlight of varying degrees is the presenting complaint in all the conditions. Infants and children with porphyrias experience burning and stinging pain on sun exposure and present with incessant crying even at night. A characteristic feature of ataxia telangiectasia (AT) is a high sensitivity to ionizing radiation (such patients are frequently subjected to ionizing radiation for associated systemic malignancies) which may alert the clinician to consider this diagnosis (Table 3). Children with idiopathic photodermatoses, drug-induced photosensitivity and phytophotodermatitis may not have overt and significant sensitivity to sunlight. Following symptoms and signs may be present in several of the disorders

Butterfly erythema: This is the presenting feature in Bloom’s syndrome (BS), Rothmund Thomson syndrome (RTS), Cockayne syndrome (CS), SLE, pellagra and Hartnup disease (HD). The edema and violaceous discoloration involving the periorbital and malar area of the face in juvenile diabetes mellitus (DM) is accentuated by sun exposure. In longstanding disease, there is a persistent edematous plaque associated with scaling in the involved area.

Blisters: Vesiculo-bullous lesions on photo-exposed areas that heal by scarring and pigmentation resembling discoid LE are seen in RTS, BS (Figure 1) and porphyrias. In addition, porphyrias are associated with milia formation and mutilation of the affected parts.

Dermatitis: A sharply demarcated, erythematous, dry, scaly dermatitis over the face, neck and other photo-exposed areas is seen in pellagra and Hartnup disease (Figure 2). A well-defined eruption on the front of the neck resembling sunburn (Casal’s necklace) is characteristic of pellagra. Covered body parts are spared in these conditions. However, vulvitis or scrotal dermatitis may be associated with pellagra. Flexural scaly, lichenified lesions simulating atop dermatitis

Table 3: Distinguishing clinical features among photosensitive genodermatoses

| Clinical features                      | BS   | CS   | RTS  | XP   | AT                      |
|---------------------------------------|------|------|------|------|-------------------------|
| Photosensitivity                      | +    | +    | +    | +    | + High sensitivity to ionizing radiation |
| Telangiectasia                        | +    | +    | +    | +    | +                      |
| Poikilodermia                         | -    | +    | -    | -    | -                      |
| Mottled pigmentation                  | -    | +    | +    | +    | +                      |
| Freckles                              | -    | -    | +Pigmented | macules on conjunctiva |
| Premature graying of hair             | -    | +    | -    | -    | +                      |
| Skin atrophy                          | -    | +    | +    | -    | -                      |
| Loss of subcut fat                    | -    | +    | -    | -    | -                      |
| Involvement of covered body parts     | -    | +    | -    | -    | -                      |
| Facies                                | Narrow, keel-shaped, dolicocephaly | Mickey mouse-like | Bird-like | - | - |
| Recurrent infections                  | ++   | -    | +    | -    | ++                      |
| Mental retardation                    | -    | +    | -    | +    | -                      |
| Deafness                              | -    | +    | -    | +    | -                      |
| Neurological abnormality              | -    | +    | -    | +    | ++                      |
| Cutaneous malignancy                  | +    | -    | +    | +    | +                      |
| Systemic malignancy                   | +    | -    | +    | -    | +                      |

Key: BS – Bloom’s syndrome, CS – Cockayne syndrome, RTS – Rothmund Thomson syndrome, XP – Xeroderma pigmentosum, AT – Ataxia telangiectasia
are seen in phenylketonuria.\textsuperscript{[2]} An eczematous eruption over the photo-exposed parts may be found in patients with AT. A scaly, erythematous annular eruption involving the periorbital area (spectacles-like distribution) and trunk is the presenting cutaneous feature of neonatal LE.\textsuperscript{[6]} Sometimes patients with AP present with facial dermatitis, particularly involving the nose.\textsuperscript{[13]}

**Telangiectasia:** This is the prominent feature in genetic disorders like RTS, BS, xeroderma pigmentosum (XP) and CS. Prominent conjunctival telangiectases may also be seen in these patients.\textsuperscript{[11,14]} Involvement of the bulbar conjunctiva with telangiectases is the initial presenting feature of AT.\textsuperscript{[15]} Thereafter lesions appear over other photo-exposed areas. Children with SLE and juvenile DM may show prominent facial telangiectases.\textsuperscript{[12]}

**Poikiloderma:** Marked poikiloderma of the photo-

![Figure 1: Dolicocephaly and narrow keel-shaped face in Bloom’s syndrome with crusted lesions and telangiectasia over central face](image1)

![Figure 2: Sharply demarcated dermatitis involving the butterfly area in Hartnup disease](image2)

![Figure 3: Poikiloderma over photoexposed areas in RTS](image3)

![Figure 4: Childhood PLE](image4)
exposed skin (Figure 3) is the predominant clinical feature in older children with RTS. Longstanding patients with juvenile DM also present with poikilodermatous changes.

**Pigment dilution:** Pigment dilution involving the skin, hair and eyes is observed in children with albinism, giving rise to pink skin, white hair and light colored iris. In dark-complexioned children with PKU, the color dilution of the skin and hair may not be readily appreciable as the resultant skin pigmentation is darker than in average white children.

**Hypertrichosis:** Lanugo-like hair over the extremities and coarse facial hair are seen in porphyrias, especially congenital erythropoietic porphyria (CEP). Juvenile DM is characterized by patchy areas of hypertrichosis.

**Associated systemic features:** Many disorders that manifest with photosensitivity are associated with systemic involvement whose recognition helps in the diagnosis.

Cerebellar ataxia and psychiatric disturbances follow the skin lesions in patients with Hartnup disease. Prominent cerebellar ataxia and nystagmus are also features of AT. Epilepsy and extrapyramidal disorders are seen in association with phenylketonuria. Mental retardation is seen in HD, PKU, AT, CS and XP. Subsequently, frank disorientation and neurological symptoms develop. Seizure and psychosis are seen in 50% of cases of childhood SLE.

Growth retardation, both intra- and extraterine, characterizes BS. Retarded physical growth is also a feature of PKU, porphyrias, AT, CS and XP. A musty odor resulting from excreted amino acids in urine and sweat is characteristic of PKU. Dark-colored urine, present since birth, is seen in HEP. Pink to brown staining of the diapers is seen in infants with CEP. Variable features of immunodeficiency are seen in BS, RTS, AT and PKU. Early onset malignancies are characteristic of BS, XP and RTS.

Ophthalmic involvement in the form of scarring of eyelids, loss of eyelashes and prominent conjunctival telangiectases is seen in XP, BS and RTS. Patients with CS may develop cataract, retinal degeneration and optic atrophy. Watery eyes and photophobia resulting from conjunctivitis are seen in AP. Photophobia is also observed in children with albinism, PKU and CEP. A red ocular reflex is seen in patients with albinism.

**Idiopathic photodermatoses**

PLE, SU, AP and HV have overlapping clinical pictures but can be distinguished easily. PLE is the commonest photodermatosis in childhood. PLE lesions usually appear 2 hours to 3 days following sun exposure. They are commonly seen over the face, the ‘V’ area of the chest, the back of the neck and the dorsolateral aspects of the forearms and persist for several days to weeks. Grouped papules, vesicles and eczematous plaques are the commonest morphological patterns observed. A particular type observed in 5-12 year-old boys is juvenile spring eruption. Here, recurrent episodes of an itchy papulo-vesicular eruption occur over the helices of the ears and adjacent areas, followed by crusting and healing without scarring.

Solar urticaria is a rare condition particularly in young children. Typical urticarial wheals appear within seconds to minutes following sun exposure. They generally resolve within 1-2 hours and almost always within 24 hours of avoidance of sun exposure. The distribution of lesions is similar to that of PLE, except that facial lesions are commoner in SU. Sunlight-induced urticaria may be a symptom of erythropoietic protoporphyrinaemia (EPP), but the latter starts at an earlier age, family history is often positive and the skin lesions are painful. The heat following sun exposure may be the precipitating factor for cholinergic urticaria but the lesions are commoner over covered body parts where the temperature is higher.

Actinic prurigo is commoner in girls and manifests by 9-10 years of age as pruritic excoriated papules and nodules on exposed body parts. Associated conjunctivitis and actinic cheilitis are characteristic.

Hydroa vacciniforme is a very rare condition seen among school-going boys. Recurrent crops of deep-seated tense vesicles with surrounding erythema appear on exposed body parts within 1-2 days of sun exposure and heal with pock-like scars.
Phytophotodermatitis is not uncommon in school-going children. Lesions are localized to the hands, lower legs and around the lips. Bizarre streaks of dermatitis may be observed over the trunk secondary to dripping of the juice of the fruit. Dermatitis bullosa striata pratensis is characterized by a linear vesicular eruption of acute onset followed by hyperpigmentation. It occurs following contact with certain plant products and subsequent exposure to sunlight.\(^9\)

**INVESTIGATIONS**

Phototesting and photo-patch testing are not commonly performed in children. Phototesting is indicated when idiopathic acquired photodermatoses are suspected.\(^1\) If history and physical examination suggest the influence of a photoallergen, photo-patch testing is helpful. These tests are not helpful in diagnosing genodermatoses, porphyrias and nutritional disorders.\(^1\) Photo-patch testing is not helpful in diagnosing PLE and SU and is contraindicated for evaluation of phototoxic reactions.\(^1\) Both these tests need patient motivation and cooperation, and are therefore difficult to conduct in children.

All photosensitive children without definitive clinical diagnosis should be evaluated for antinuclear (ANA), anti-Ro (SSA), and anti-La (SSB) antibodies and porphyrin levels.\(^19\) Wood’s lamp examination helps in demonstrating erythrodontia in CEP and HEP.\(^19\) It is also a helpful tool in detecting the reddish pink fluorescence of serum, erythrocytes, urine and stool in patients with different types of porphyrias.\(^2,19\)

Urinary ferric chloride test is indicated in children with suspected aminoaciduria.\(^2,19\) Since this test may be negative in the first few months of life in patients with PKU, screening of suspected neonates by a blood test (serum level of phenylalanine > 20 mg/dl) or Guthrie test is advised.\(^19\) Urine chromatography for detection of amino acids is useful in children with PKU and HD.\(^2\)

Patients with childhood SLE and juvenile DM need a thorough evaluation for systemic involvement. Muscle biopsy, muscle enzyme estimation and electromyographic studies are performed in the presence of proximal myopathy and muscle tenderness.\(^12\) ECG and echocardiography should be performed in all suspected cases of neonatal LE to rule out congenital heart block or other anomalies.\(^12\)

Skin biopsy is helpful in idiopathic photodermatoses. and porphyrias (deposition of PAS positive porphyrins in a perivascular distribution is characteristic).\(^2\) Direct immunofluorescence study of the skin biopsy specimen is helpful in diagnosing SLE (complement and immune deposits at the dermoepidermal junction)

Rarely, chromosomal or genetic studies are needed for the specific diagnosis of genodermatoses.

**MANAGEMENT**

Dermatologists play a major role in diagnosing and treating photosensitive children. Their duty also encompasses counseling the parents regarding specific light avoidance, photoprotection and sometimes, change of lifestyle.\(^9\) Outdoor activities of such children should be curtailed as much as possible. Environment (tropical countries) and socioeconomic status are prohibitive factors for complete sun avoidance. Whenever feasible, use of tightly woven, dark colored, full-sleeved clothing and a hat provide enough photoprotection.\(^9\) Window glass filters out most of the UVB in sunlight, but transmits UVA and visible light readily.\(^1\) Hence, children with photosensitivity disorders precipitated by these spectra of the sunlight should be provided with protection during their indoor stay as well. Strict sun avoidance is a must for children with porphyrias, BS, XP and RTS. Shades or filters can provide protection from fluorescent lights.\(^9\)

Habitual use of sunscreens on all exposed areas is helpful in all such cases. Topical agents, especially conventional absorbent sunscreens for UVA and UVB, are ineffective in patients with porphyrias.\(^2\) Preparations containing titanium dioxide are more effective, since these patients are sensitive to the visible range.\(^2\) Commercial preparations of zinc oxide available in different skin tones are cosmetically acceptable.\(^9\) Beta carotene improves light tolerance in patients with porphyrias. A daily dosage of 50-200 mg/day to achieve a serum level of 500 µg/dl is effective and benefit is observed by 1-3 months.\(^2\) Cysteine (500 mg twice daily) prevents photosensitivity associated with EPP.\(^2\)
In addition to sunscreens, topical steroids and antihistamines are used in idiopathic photodermatoses. Thalidomide has been found to be effective in AP affecting some races. Prophylactic use of low-dose UVA/PUVA/UVB is helpful in preventing recurrence in this group of disorders.

Regular surveillance of children with albinism and genodermatoses like BS, XP and RTS is essential for the early diagnosis of cutaneous and systemic malignancies. In some XP patients, oral isotretinoin (1 mg/kg/day) decreases the incidence of cutaneous malignancies significantly.

A low-phenylalanine diet continued lifelong is the mainstay of therapy in patients with PKU. Skin color, photosensitivity, foul odor and eczema are reversible with such treatment. Children with HD and pellagra need supplementation with nicotinamide.

CONCLUSION

The accurate diagnosis of photosensitivity disorders in children is difficult. Rare disorders like BS, RTS, HD and AP can be confused with commoner ones like pellagra and PLE. On such occasions, the clinician must be able to identify the subtle symptoms and signs of each disorder to ensure the appropriate management.

There are certain primary dermatological conditions that are exacerbated by sun exposure. These include atopic dermatitis, pemphigus erythematosus, herpes labialis, erythema multiforme, actinic lichen nitidus, actinic lichen planus, viral exanthem and photosensitive psoriasis. Children suffering from these disorders deserve counseling and photo-protection in addition to the specific treatment.

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