Photodermatoses: classification, approach and management

Speaker: Dr. Yiu-Hoi Chan
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Photodermatoses are a group of heterogeneous disorders with diverse clinical manifestations and complex aetiology. They can be classified into five general categories including (1) photodermatoses which are secondary to exogenous agents, such as phototoxic and photoallergic reactions, (2) photoexacerbated dermatoses, (3) photodermatoses which are idiopathic such as polymorphic light eruption (PMLE), actinic prurigo (AP), hydroa vacciniforme, chronic actinic dermatitis (CAD), and solar urticaria (SU), (4) photodermatoses which are secondary to endogenous agents mainly the porphyrias, and (5) genodermatoses. The initial step in evaluating a photosensitive patient is a detailed and directed personal and family history as well as physical examination. History should include time of onset of symptoms, duration, drug or herbs intake, travel history, exposure to cosmetics and aroma therapy. Physical examination should focus on distribution of the lesion, morphology, pattern and sign of underlying causes such as lupus erythematosus. Investigations will be directed by history and physical examination. Skin biopsies, autoimmune markers, porphyrin profiles, phototesting and photopatch tests may be required to confirm the diagnosis.

The three primary sources of exogenous photosensitivity are industrial, cosmetic and therapeutic. Drug-induced photosensitivity is classified into either a phototoxic response to both oral and topical agents, which is more common, or a photoallergic response. Common drugs that induce photosensitivity include cephalosporins, quinolones, tetracyclines, non-steroidal anti-inflammatory drugs, diuretics, amiodarone, anticonvulsants, sulphonylurea and anti-hyperlipidemics. Treatment is avoidance of the culprit agent and restriction of ultraviolet light exposure until this is done.

Examples of photoexacerbated dermatoses are dermatomyositis, rosacea, lichen planus, erythema multiforme, lupus erythematosus, seborrhoeic dermatitis, Darier's disease, herpes simplex infection and pemphigus erythematosus. Ultraviolet B irradiation seems to be responsible but the mechanism is unclear. Usually the diagnosis can be made in consideration of the clinical features of the underlying dermatosis.

For idiopathic photodermatoses including PMLE, AP, hydroa vacciniforme, CAD and SU, diagnosis may be established on the basis of clinical symptoms, location of lesions, time course and
relationship with sun exposure, histology and phototesting. Basic principles of treatment for this group of photodermatoses are restriction of ultraviolet light exposure, use of broad-spectrum sunscreen and phototherapy to induce a hardening phenomenon. Hydroxychloroquine may be used in patients in PMLE. Low dose thalidomide is effective for severe cases of AP and hydroa vacciniforme. Treatment for CAD consists of avoidance on sunlight and allergen, topical steroid, systemic prednisolone and azathioprine for resistant cases. Non-sedating antihistamines are effective for SU. For intractable cases of SU, plasmapheresis is occasionally used to eliminate the photoallergen.

Porphyrias are a peculiar group of diseases resulting from hereditary or acquired partial deficiencies of different enzymes involved the biosynthetic pathway of heme. There are seven basic clinical forms of porphyrias related to a deficiency of each of the involved enzymes. Porphyria cutanea tarda (PCT) presents with increased susceptibility of exposed skin to minor trauma with erosions, blisters, crusting and later scarring and milia. Hypertrichosis and pigmentation are common. Approximately 80% of patients have sporadic or type I PCT and the uroporphyrinogen decarboxylase enzyme deficiency being restricted to the liver. Alcohol abstinence and avoidance of other hepatotoxins are essential and repeated venesection may be needed for clinical resolution.

Finally, the speaker highlighted on the tips and misconceptions about sun avoidance and use of sunscreen. Sufficient amount of sunscreen should be applied up to the dosage of 2 mg/cm² and do not miss areas such as ears, perioral area and medial canthus.

**Learning points:**
Photodermatoses can be classified into five general categories. Diagnosis of photosensitive dermatoses relies on detailed personalised history and physical examination followed by relevant investigations such as skin biopsies, phototesting and porphyrin studies. Mainstay of treatment includes restriction of ultraviolet light exposure, avoidance of culprit chemicals or drugs, use of sunscreen, phototherapy and other topical or systemic drugs. Sufficient amount of sunscreen should be used for effective protection.

**Skin disorders among HIV-infected patients**
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Skin disorders are commonly encountered in human immunodeficiency virus (HIV)-infected patients. Up to 90% of HIV-infected persons suffer from skin diseases during their course of illness and, in many cases, skin disease may be the first manifestation of HIV infection. In general, a declining immunity is associated with more extensive skin disease, a more protracted course and greater resistance to treatment. The immunosuppression and immune dysregulation caused by HIV infection also tend to alter the clinical features and natural history of skin disease. The stage of HIV disease, as indicated by CD4 count, often determines the type of skin disease encountered.

Based on the pathogenic mechanism, the skin diseases can be classified as follows: (1) skin infections due to viruses, bacteria, fungi and infestations (2) cutaneous neoplasm such as Kaposi’s sarcoma (3) inflammatory conditions such as seborrhoeic dermatitis, psoriasis and eosinophilic folliculitis (4) miscellaneous disorders such as lipodystrophy and cutaneous drug eruption.
Molluscum contagiosum is caused by a pox virus which selectively infects human epidermal cells. The typical lesions are umbilicated pearly papules with a diameter of 3-5 mm. Molluscum contagiosum infection is a clinical marker of the degree of immunodeficiency. The prevalence and the severity are inversely related to CD4 count. Extensive mollusca are almost always associated with CD4 count less than 200/mm³. Unlike immunocompetent host, molluscum contagiosum in HIV-infected person tends to persist. Those present in advanced HIV disease are particularly resistant to therapies. Various modalities have been tried such as surgical removal, curettage, electrocoagulation, cryotherapy, shave excision and carbon dioxide laser. Topical agents used in this condition include podophyllin, tretinoin, cantharidin, imiquimod and cidofovir.

Bacillary angiomatosis is an infectious disease of the skin and viscera characterised by angiomatosus lesions. The causative agent is either *B. quintana* or *B. henselae*. Among HIV-infected individuals, bacillary angiomatosis is most commonly seen in advanced HIV disease status with CD4 count less than 50/mm³. The morphology of the lesions can present with the following features: pyogenic granuloma-like lesion, hyperpigmented indurated plaques, subcutaneous nodules and dusky-red color nodule. The lesions have a predilection to eyelids. It may also affect the oral mucosa and internal organs, especially the liver and the spleen. Erythromycin for 8-12 weeks can be used in this condition. Other options include doxycycline and minocycline. A Jarisch-Herxheimer reaction is not uncommon after initiation of therapy. Although untreated bacillary angiomatosis is a fatal disease, most patients respond well to antibiotic. In the event of recurrence, it may be necessary to retreat for a further 16 weeks or even indefinitely.

*Penicillium marneffei* is a dimorphic fungus that can cause systemic mycosis in human. It is most frequently found in immunocompromised hosts, as in advanced HIV-infected patients (CD4 <50/mm³). Disseminated *Penicillium marneffei* infection is classified as an AIDS-defining illness in Hong Kong and indeed contributing 10% of the primary AIDS-defining illness. Characteristic lesions are umbilicated papules with or without central necrosis (molluscum contagiosum-like), but maculopapules, ulcers, nodules, acneiform lesions and folliculitis have been reported. The skin lesions are mainly distributed on the upper half of the body with 70% being on the face, scalp, upper extremities and trunk. Umbilicated papules are also commonly present on the oral mucosa in disseminated disease. Disseminated *Penicillium marneffei* is a fatal disease. Amphotericin B for two weeks followed by 10 weeks of oral itraconazole is necessary. Since relapse is common in the absence of chronic suppressive therapy, all patients should be given oral itraconazole for secondary prophylaxis.

Eosinophilic folliculitis is a chronic skin condition that occurs in the more advanced stage of HIV infection, usually when CD4 count is less than 200/mm³. It is a clinicopathological entity of uncertain pathogenesis, though it has been postulated that the normally innocuous follicular micro-organisms become antigenic in a Th2-dominant milieu. *Pityrosporum yeasts, Demodex mites, or Corynebacterium* have been implicated. It mainly affects adult HIV-infected man who has sex with man. All cases have chronic intense intractable pruritus. This condition often presents with erythematous perifollicular papules and pustules occasionally. Trunk is involved in almost all the cases. Peripheral eosinophilia has been reported in 25-50% of patients. Serum IgE levels have been raised in a high proportion of patients. CD4 counts are universally decreased and generally less than 250/mm³. Eosionophilic folliculitis is notoriously resistant to treatment. Oral antihistamines and potent topical steroid are used for symptomatic relief. Antimicrobials such as metronidazole, itraconazole and permethrin are also used. Other modalities include isotretinoin that inhibits sebum secretion, phototherapy and highly active antiretroviral treatment (HAART).
In the epidemic HIV-associated Kaposi's sarcoma (KS), it is now believed that human herpesvirus type 8 is a necessary, but not a sufficient aetiological factor. Other factors such as immunosuppression play a major role. KS occurs almost exclusively in men having sex with men, bisexual men and, to a lesser degree, in women who acquire HIV infection from heterosexual contact. The initial lesions frequently develop on the face and on the trunk where the lesions follow the lines of cleavage. The oral mucosa is the initial site of disease in 10-15% of patients, usually on the hard palate. Gastrointestinal tract involvement is found in 80% of AIDS patients, especially in those with extensive cutaneous involvement. Lung involvement can be fatal. There is no curative treatment. The aim is to treat disfiguring KS, to alleviate symptoms and to improve functional impairment. Local treatment includes cryotherapy, radiotherapy, intralesional vinblastine, surgical excision, laser (carbon dioxide or pulsed-dye laser) and alitretinoin gel. Systemic therapy is indicated for aggressive disease and systemic involvement. The risk to benefit ratio should be given careful consideration as the responses are generally partial and short term and HIV replication may be enhanced by the systemic therapy (doxorubicin, vincristine, bleomycin). Many authorities recommend observing the clinical response to HAART before considering systemic therapy.

Clinical features of cutaneous melanoma: A promotion for early detection
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In considering the risk factors for cutaneous melanoma affecting local Hong Kong population, age is a very important risk factor, whereas it was much less apparent in Caucasians where sun exposure behaviour plays an important role. The risk of age was confounded by the birth cohort effect. As the local population is aging, the incidence of cutaneous melanoma in Hong Kong is predicted to increase in the coming 10 to 20 years.

The incidence and mortality rates of cutaneous melanoma have increased for decades in many western countries. Although the local rates are low as compared to the Caucasian populations, they are higher than other Asian populations. Moreover, the case fatality ratio is 38% in Hong Kong but only 10-14% in Australia.

Surgical excision at its early stage remains the only treatment that can provide survival benefits. Since the prognosis of cutaneous melanoma is mostly dependent on its staging, early detection and timely management is the most important solution to the high case fatality ratio. Promotion for early detection and timely management is needed to achieve a lower case fatality ratio in our locality. Health care professions familiar with the clinical features of cutaneous melanoma will help to detect the condition in its early stage without causing unnecessary anxiety.

Clinical categorisation of cutaneous melanoma is traditionally based on Clark's classification and is divided into four types. These are superficial

Learning points:
The frequent occurrence of both infectious and non-infectious skin diseases in HIV infected patients is thought to be due to impairment of immunity. HAART reduces the incidence and severity of skin disorders. A success in management relies on accurate diagnosis, appropriate treatment and restoration of immunity by HAART.
spreading melanoma, nodular melanoma, acral lentiginous melanoma and lentigo maligna melanoma. The practical usage of the American ABCD mnemonic as well as the Glasgow seven-point checklist has been re-iterated.

Based on a local case series with 32 Social Hygiene Service dermatology clinics, the clinical features including the subtypes and duration of the lesions, the predisposing site and size of the tumours and other signs and symptoms were reviewed in patients with cutaneous melanoma. Lastly, some other common pigmented skin lesions which might enter into the differential list of cutaneous melanoma were discussed. These include freckles, lentigo simplex, solar lentigo, nevomelanocytic naevus, atypical naevus and seborrhoeic keratosis.

Around 20 patients in Hong Kong died from cutaneous melanoma every year. Early detection and timely treatment are needed to improve survival.

**Learning points:**
Although the local incidence rate of cutaneous melanoma is low, the case fatality ratio is high. Early detection is an important measure to reduce mortality.