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Symptoms and signs may include livedo reticularis, subcutaneous nodules, palpable purpura, ulcer or digital gangrene, hypertension and internal organ involvement in systemic PAN.

Small-vessel vasculitis includes Churg-Strauss syndrome which is characterised by prodromal allergic rhinitis and asthma, peripheral eosinophilia and systemic vasculitis. Anti-cytoplasmic neutrophilic antibody (ANCA) is positive in 80% of cases of Wegener's granulomatosis (predominantly PR3-cANCA) and 90% of microscopic angiitis (mostly MPO-pANCA). Henoch-Schönlein purpura is characterised by IgA deposits on direct immunofluorescence.

Vascular disease in paediatric dermatology

Speaker: Prof. John Su

Adjunct Clinical Associate Professor, Eastern Health Clinical School, Faculty of Medicine Nursing and Health Sciences, Monash University, Australia

Vasculitis is common in paediatrics. It is classified into large-vessel, medium-sized vessel and small-vessel vasculitis.

Large-vessel vasculitis includes Takayasu's vasculitis which affects teenagers. Symptoms include high blood pressure, headache, fever, dyspnoea, weight loss and abdominal pain. Involvement of the aorta may cause pulse deficit, claudication or blood pressure discrepancy.

Medium-sized vessel vasculitis such as Kawasaki disease and polyarteritis nodosa (PAN) were discussed. Kawasaki disease is characterised by fever, strawberry tongue, cervical lymphadenopathy, erythema or oedema of palms and soles, and periungual desquamation in the subacute phase. Polyarteritis nodosa may be systemic or cutaneous.

Differential diagnoses include haemorrhagic oedema of infancy, idiopathic thrombocytopenic purpura, disseminated intravascular coagulation, haemolytic uraemic syndrome, infections, perniosis and chilblain lupus.

Treatments include supportive treatment, systemic steroids, immunosuppressants and biologicals.

Learning points:

Vasculitis may present with potentially serious systemic sequelae. It is classified into large, medium and small vessel vasculitis associated with drugs, infections, malignancies and autoimmunity.

The latest development in syphilis serology

Speaker: Dr. Edman Lam

Acting Consultant Medical Microbiologist, Public Health Laboratory Centre, Department of Health, Hong Kong

The diagnosis of syphilis remains very dependent on the conventional serological treponemal and non-treponemal tests. However, the diagnosis may not be straightforward in cases with discordant results. Previous local studies showed that line immunoassay (LIA) provided good sensitivity and specificity which was even better than that of Western blot test. Further local study done recently showed LIA may not agree with the consensus results drawn from the other conventional serological tests with a kappa coefficient of 0.55. Therefore, these newer methods should be more thoroughly studied to determine its clinical role.

Learning points:

Use of line immunoassay may provide high sensitivity and specificity in syphilis diagnosis, but because of its fair agreement with other treponemal tests, its clinical use still needs further study.

Syphilis in Guangdong Province, China

Speaker: Dr. Li-gang Yang

Consultant Dermatologist, Head of STI Control Department, Guangdong Provincial Center for STI & Skin Diseases Control, China

"Guangdong boils" is a commonly used term for syphilis infection in Southern China because it suffered from substantial syphilis epidemics around the early 16th century. With the

development and initiation of the national sexually transmitted disease (STD) control program, syphilis was almost eliminated in the 1960s. However, there was resurgence of syphilis in China in late 1970s, the disease burden disproportionately increased from 1.78 per 100,000 population in 1995 to 50.85 per 100,000 population in 2012. Guangdong reported 53,043 syphilis cases in 2012, and syphilis became the third most common infectious diseases after tuberculosis and hepatitis B. In some areas of Guangdong, the sero-prevalence of syphilis reached 30% among men who have sex with men (MSM) and low-tier female sex workers (FSWs).

Despite the measures that have been initiated, there are still many barriers to effectively control syphilis in Guangdong province. The barriers include poor access to the hard-to-reach populations who have a high prevalence of syphilis, such as MSM and FSWs, inadequate standards of STD clinics, poor coverage of syphilis screening e.g. pregnant women, affordability to STD medical care, social stigma and concerns on confidentiality, etc. The Chinese Ministry of Health released the national 10-year plan for the prevention and control of syphilis in China. The strategies included increasing access to STD testing, case detection and surveillance, improving quality of STD services and preventing mother-to-child transmission of syphilis.

Learning points:

Syphilis has been resurgent in China such as Guangdong province with a rapidly increasing trend. Efforts have been made to address the epidemic but many challenges remain, particularly for the high-risk groups. Government commitment and innovative strategies are needed.

Vascular anomalies: an overview and update

Speaker: Dr. Francis Ip
Medical and Health Officer, Social Hygiene Service,
Department of Health, Hong Kong

The most common congenital abnormality seen in the paediatric group is vascular anomalies. Vascular anomalies are regarded as birthmarks, most of them are harmless and no intervention is needed. However active intervention needs to be considered when complication, functional disturbance or cosmetic concern arises. The rarely associated systemic complications and syndromes should be borne in mind.

There has been confusion in the classification of vascular anomalies for many years. Currently, a comprehensive classification, adopted by the International Society for Study of Vascular Anomalies (ISSVA), which is based on clinical and histopathological features is widely used. Vascular anomalies are broadly divided into haemangiomas and vascular malformations. Advances in the use of immunochemistry and molecular methods improve our understanding in the pathogenesis and characteristics of the vascular anomalies. Infantile haemangioma was used as an example for discussion. Propranolol has become one of the mainstay treatments for complicated infantile haemangioma, although large randomised control trials are still ongoing.

Learning points:

Vascular anomalies are common. We need to keep up-to-date on the information and treatment about them.

Suction blister epidermal grafting for treating stable vitiligo

Speaker: Dr. William Tang
Dermatologist, Private Practice, Hong Kong

Various surgical methods aiming at replenishing lost melanocytes in vitiligo have gained popularity since last century. Among them, suction blister epidermal grafting (SBEG) was widely used.

In 1964, Kiistala firstly reported in-vivo separation of epidermis by the production of suction blisters. Later in 1971, Falabella described the suction blister technique for repigmentation of vitiligo. The technique is rather straightforward and suitable for small localised stable vitiligo. Stable vitiligo is defined as the absence of new lesions, absence of progression of existing lesions and absence of the Koebner's phenomenon in the previous one year. Minigraft test before the procedure can be used to ensure the stability of such lesion. The selected patient also has to be co-operative and motivated but not with unrealistic expectation.

The first step of SBEG is to create a suction blister by applying the suction cup on the donor site which is usually the inner arm, abdomen, thigh or buttock. The suction pressure is kept at around 300 mmHg (40 kPa) for 1-2 hours. The second step is to remove the recipient site epidermis by dermabrasion. The final step is to harvest the epidermal graft from the donor site and place it over the recipient site.

Studies showed that SBEG can achieve more than 75% repigmentation in 85% of vitiligo patients receiving this treatment.

Learning points:

SBEG is one of the surgical repigmentation methods that is suitable for small localised stable vitiligo and can be carried out in general dermatology clinics.

Treatment of vitiligo: review on the medical treatment and phototherapy

Speaker: Dr. Elsa Chu

Medical and Health Officer, Social Hygiene Service, Centre for Health Protection, Department of Health, Hong Kong

Topical corticosteroids are commonly used as a first-line treatment of vitiligo. Their efficacy is attributed to the modulation of the immune response. Based on comparative studies, topical corticosteroid is the most clinically effective choice for topical therapy. However, possible side-effects including epidermal atrophy prohibit its long-term use. Topical calcineurin inhibitors provide similar to slightly inferior results compared to topical corticosteroids. The study done by Dr. Chu showed the potential of topical tacrolimus in inducing repigmentation in vitiligo patients who fail to respond to topical steroid. Calcipotriene is a topical vitamin D3 analogue which when combined with corticosteroids, can increase repigmentation rates and shorten the delay in the onset of repigmentation of vitiligo.

The second-line treatments of vitiligo include phototherapy and laser. Studies have proven that narrowband UVB is superior to psoralen-UVA in producing disease stability and repigmentation. The mechanism of action of laser therapy for treatment of vitiligo is similar to that of phototherapy, but it allows for targeted treatment with less body irradiation, and less impact on the healthy skin.

Finally, camouflage should be recommended to patients at all stages of treatment. Depigmentation may also be considered in patients with extensive disease or who cannot attain cosmetically acceptable outcome with other modalities of treatment.

Learning points:

The aims of treatment for vitiligo are to minimise disease progression, attain repigmentation and achieve cosmetically pleasing result. Treatment should start with less aggressive, cost-effective modalities, reserving more invasive and expensive options for those who fail first-line therapies.

Cutaneous manifestations in renal transplant recipients in a regional hospital in Hong Kong

Speaker: Dr. Lai-ping Wong

Medical and Health Officer, Social Hygiene Service, Department of Health, Hong Kong

The findings of a study on cutaneous manifestations in renal transplant recipients (RTR) in a regional hospital in Hong Kong were presented. When compared with matched age and sex controls, RTR had a higher prevalence of skin diseases. More RTR developed skin infections and non-infective, non-malignant drug-related cutaneous manifestations than controls. Renal transplant recipients with transplantation less than 12 months were more prone to acneiform eruption; RTR with transplantation more than 12 months were more prone to skin infections. Older RTR were more prone to develop warts, younger RTR were more prone to have acneiform eruption and Cushingoid features. Male RTR were more prone to have dermatophytoses, folliculitis, acneiform eruption and sebaceous hyperplasia while female RTR were more prone to have hypertrichosis and gingival hyperplasia.

Learning points:

Renal transplant recipients in Hong Kong are prone to skin infections and drug-related cutaneous manifestations. Patient education, skin examination by physicians during follow-up and early referral to dermatologist, if necessary, can improve the skin condition of renal transplant recipients in Hong Kong.

Neutrophilic dermatoses in children

Speaker: Prof. John Su

Adjunct Clinical Associate Professor, Eastern Health Clinical School, Faculty of Medicine Nursing and Health Sciences, Monash University, Australia

Neutrophilic dermatoses can be classified by histology and associated systemic diseases. Histologically, it can be divided into non-angiocentric entities like pustular psoriasis and

acne fulminans, and angiocentric entities which can be subdivided into vessel wall-affected types like leucocytoclastic vasculitis, polyarteritis nodosa or vessel wall-spared types like Sweet's syndrome and pyoderma gangrenosum.

Auto-inflammatory syndromes are characterised by recurrent fever, joint pain and skin rashes. Examples include familial Mediterranean fever, Muckle-Wells syndrome and PAPA (syndrome of pyogenic arthritis, pyoderma gangrenosum and acne). They share a common feature with the autoimmune diseases in that body tissues are attacked by its own immune system. Most of the auto-inflammatory syndromes are hereditary with chronic activation of the innate immune system in the absence of pathogen or pathogenic antibodies.

Cases illustrated included a case with severe acne and pyoderma gangrenosum. The differential diagnoses include PASH (syndrome of pyoderma gangrenosum, acne and suppurative hidradenitis: a new entity that requires prompt treatment to prevent complications) and other auto-inflammatory syndromes like PAPA, paediatric pyoderma gangrenosum (PG) which accounts for 5% of PG cases and is associated with conditions such as Takayasu's arteritis and immunodeficiency. Treatments include systemic steroid, cyclosporine and dapsone.

Learning points:

Neutrophilic dermatoses are rare in real life. Associated conditions other than skin manifestations like joint pain and systemic diseases should be looked for in order to arrive at a correct diagnosis and treatment plans.

Giant congenital melanocytic naevi

Speaker: Dr. David Orchard
Director, Dermatology Department, Royal Children's Hospital, Melbourne, Australia

Large to giant congenital melanocytic naevi (CMN) (>20 cm) are uncommon but can be complicated by melanoma and neurocutaneous melanocytosis,

cosmetic or psychological issues. For the risk of melanoma, the absolute risk (AR) for large/giant CMN was 2.5-5% compared to 0-3% for smaller CMN. The relative risk (RR) for large/giant CMN was 50- to 1000-fold compared to 4-to 21-fold for smaller CMN. Melanoma develops within 10 years in >70% of subjects with large/giant CMN compared to >20 years in smaller CMN. Risk factors of melanoma development include very large size (>50 cm) and variegated colour. Melanoma develops in the large CMN usually before puberty, frequently starts in the lesion and can develop in other places but not in the satellite naevi. Neurocutaneous melanocytosis develop in the embryonic stage. The excess number of melanocytes can proliferate like naevus in the skin, and melanoma may arise. The risk of CNS melanoma is <1%. Large CMN occurs more on the trunk than limbs. Lesions near the spine or >20 satellite naevi have a higher risk of developing melanoma (50-fold). Magnetic resonance imaging may be considered in children with large/giant CMN (20/40-60 cm), multiple satellite lesions and any symptomatic patients. Initial negative scan would be reassuring though a few cases developed melanoma after a normal scan.

Learning points:

For giant CMN, regular surveillance for malignant change is warranted.

Skin grafting for vitiligo patients, the National Skin Centre's experience

Speaker: Dr. Steven Thng
Senior Consultant, National Skin Centre, Singapore

Skin grafting is indicated for vitiligo patients who do not respond to conservative treatment strategies.

Proper selection of patients is essential for successful repigmentation in skin grafting. The selected cases should have no new lesions in the past six months and the old lesions are not spreading. Segmental vitiligo is usually stable and thus very suitable for skin grafting. For vitiligo vulgaris, test graft is required to ensure the stability of the vitiligo. Other selection criteria include no

tendency of keloid formation and absence of Koebner's phenomenon previously.

The technique of non-cultured epidermal cellular grafting is to obtain an epidermal sample from the donor site such as the hip by superficial shaving, then treat the sample with trypsin, followed by trypsin inhibitor and centrifugation to extract epidermal cells and prepare a cellular suspension containing melanocytes. Before transplanting this cellular suspension onto the recipient site, de-epithelisation of the recipient site is done by dermabrasion or CO₂ laser ablation. The advantage of cellular grafting over tissue grafting is that it can treat recipient skin up to 20-fold the size of the donor skin. The whole procedure can be completed in two to three hours. The success rate (more than 75% repigmentation six months post-grafting) is above 90% for segmental vitiligo, and about 70% for vitiligo vulgaris.

Learning points:

Skin grafting is a good alternative for patients with stable vitiligo who have failed conservative treatments.

The use of immunosuppressive therapy in atopic eczema

Speaker: Dr. David Orchard

Director, Dermatology Department, Royal Children's Hospital, Melbourne, Australia

For atopic eczema (AE), the rationale of using immunomodulatory therapy is to stop scratching, restore the skin barrier and allow sleep. All of us are aware of the side-effects of topical corticosteroid (TCs), but in the speaker's experience, 15 grams of mometasone or methylprednisolone cream per week for six months is considered to be 'child-safe'. TCs prescribed should be potent enough and given in sufficient quantities to clear the bursts. Topical calcineurin inhibitor can be used for periorbital dermatitis with an efficacy similar to weak- to medium-strength topical steroid but it can cause irritation.

Antihistamines are considered not effective in controlling pruritus in AE but effective for symptomatic dermographism as demonstrated in a Cochrane review. For systemic immunosuppressants, options include phototherapy, prednisolone, cyclosporine, azathioprine, mycophenolate etc. Prednisolone is used for short-term as a kick-start for severe eczema and special occasions, e.g. school camp, at a dose of 1 mg/kg/day for three days followed by tapering over 10 days. Cyclosporine is available in syrup and has a fast onset of action (1-2 weeks). Dose is up to 5 mg/kg/day in divided doses for 3-9 months with regular blood test monitoring at baseline and then every 1-3 months. Side-effects include hypertension, nephrotoxicity, immunosuppression and hyperlipidaemia. Patients would expect a flare after weaning. It is advisable in children with a severe, protracted flare of eczema. Azathioprine is advised for chronic eczema with normal IgE level. The target dose is 2.5 mg/kg/day, starting at 1/4-1/3 of the full dose then gradually increasing to the full dose over 6-8 weeks with blood test monitoring and pre-treatment thiopurine S-methyltransferase (TPMT) level measurement (in Australia, ~1% population have TPMT deficiency and are prone to myelotoxicity). No cumulative myelotoxicity was observed in children, though various degrees of lymphopaenia were most commonly observed. For the suggested maintenance dose of 1-1.5 mg/kg/day, disease control can be maintained. Patients were able to wean with control maintained from months to one year. Serum IgE does not seem to improve upon therapy. Methotrexate is relatively safer but not as powerful and can be considered in recalcitrant eczema not improved by or unsuitable for cyclosporine or azathioprine. Side-effects include cumulative hepatotoxicity. Other options include mycophenolate mofetil (not funded in Australia), gamma-globulin and interferon gamma.

Learning points:

Atopic eczema should be controlled with more liberal use of topical corticosteroids. Systemic agents may be used if required.

Drug reaction with eosinophilia and systemic symptoms (DRESS): from bedside to the bench

Speaker: Dr. Johnny Chan

Clinical Assistant Professor, Division of Dermatology, Department of Medicine, Li Ka Shing Faculty of Medicine, The University of Hong Kong, Hong Kong

Drug reaction with eosinophilia and systemic symptoms (DRESS) is an acute distinct idiosyncratic drug related hypersensitivity reaction. It has a significant mortality of 10%-20% and is classified under a category termed severe cutaneous adverse reaction (SCAR). Fulminant hepatitis or myocarditis is the most common cause of death. Drug reaction with eosinophilia and systemic symptoms is associated with long term autoimmune complications including diabetes, thyroiditis and systemic lupus erythematosus. It has a characteristic delayed onset of two to six weeks after drug administration and a classic triad of rash, fever and visceral involvement. Common cutaneous manifestations include sterile pustules over the head and neck regions, exanthematous eruptions and facial oedema. All internal organs can be affected.

A complex interaction of impaired endogenous drug metabolism, genetic predisposition and viral infection was postulated in the pathophysiology. Several drug-specific phenotypic patterns were identified. An example would be the longer incubation period and higher rate of renal impairment in allopurinol-induced DRESS. In a local single centre study, cotrimoxazole was found to be associated with a long-term severe psoriasiform eruptions and DRESS caused by aromatic anti-convulsants tended to result in more severe liver parenchymal enzymes impairment.

In the last decade, there has been some academic researches on the genetic basis of individual susceptibility to severe drug reactions. For example, the HLA-B 58:01 and HLA-B 15:02 alleles have been identified as being associated with SCAR caused by allopurinol and carbamazepine respectively. A large, local study was being conducted with an objective to define the efficacy and cost-effectiveness of HLA screening in the prevention of allopurinol-induced

SCAR. Meanwhile, other laboratory tests like the lymphocyte transformation test have emerged in recent years to assist in the management of DRESS.

Learning points:

DRESS is an acute drug-related hypersensitivity reaction. It has diverse systemic and cutaneous manifestations. Physicians should have a high index of suspicion of this disease as it has a significant mortality.

Bullous pemphigoid: a review of patients managed in hospital and its risk factors for poor prognosis and early mortality

Speaker: Dr. Mimi Chang

Resident Specialist, Department of Medicine and Therapeutics, Prince of Wales Hospital, Hong Kong

Bullous pemphigoid (BP) is the most common autoimmune blistering disease. Local data on the treatment and prognosis are lacking. This study was performed to evaluate the disease characteristics, inpatient treatment, morbidity and to analyse the risk factors for flare-up and early mortality in Chinese patients that were hospitalised for BP. This was a retrospective cohort of patients with newly diagnosed BP managed in Prince of Wales Hospital from 1st January 2002 to 31st December 2011. Patients with newly-diagnosed histologically confirmed BP with compatible immunofluorescence (IMF) findings were studied. Patients with possible BP without biopsy, subepidermal blistering disease of other causes or were old cases with flare-up during the study period were excluded. A total of 121 patients were analysed. The mean age of onset was 79.9 years. Eighty-one percent of the patients had generalised BP. Most patients had poor premorbid status and multiple medical comorbidities with diabetes, hypertension and cerebrovascular disease being the most common. First-line treatment with systemic prednisolone 0.5 mg/kg was given in 91% of

the patients. Adjuvant immunosuppressants were given in 13.2% of cases. Treatment-related infective complications occurred in 66% of the patients. Late flare-up occurred in 42% of the patients and was associated with delayed initial remission, use of immunosuppressants and rapid tapering of steroid early in the course of treatment. The average duration of survival after diagnosis was 18 months. First year mortality was 41.5%. Risk factors associated with early mortality after hospitalisation were bedbound status, presence of malignancy, anaemia and hypoalbuminaemia. However, patients with the use of immunosuppressants or had a history of flare-up seemed to be fare better against early mortality.

Learning points:

Patients with BP admitted to hospital represented those at the more severe spectrum of the disease, and had poor physical status, multiple comorbidities, generalised involvement and a tendency for flare-up. There was significant morbidity and mortality especially in the first year. Patients with poor prognosis (anaemic, hypoalbuminaemia, bedbound status, and malignancy) should be monitored closely.

As mitosis is the basis of cell proliferation, anti-mitotic agents have been tried in the management of hypertrophic and keloidal scars with promising results. 5-fluorouracil (5-FU) was shown to inhibit fibroblast and cell proliferation in tissue culture. It was also found to be effective in treating keloidal scars in several uncontrolled and non-randomised studies.

In this small scale randomised controlled study, 26 patients were recruited. Both intralesional steroid therapy and intralesional 5-FU were found to cause significant volume reduction and symptomatic improvement. However, no statistically significant difference was shown between two groups. 5-fluorouracil was well-tolerated with no systemic side-effects except hyperpigmentation.

Learning points:

The management of keloids remains a clinical challenge. Both intralesional steroid and intralesional 5-FU were shown to cause a significant volume reduction and symptomatic improvement in this local study.

Use of intra-lesional 5-fluorouracil in the treatment of keloids

Speaker: Dr. Mildred Wat

Medical and Health Officer, Social Hygiene Service,
Department of Health, Hong Kong

Keloid scars are abnormal proliferations of fibrous tissue after dermal injuries. They are not only cosmetically disturbing, but also lead to pruritus, pain, restriction of motion and deformity. However, a universally accepted treatment for keloids is lacking and the management of keloids is a clinical challenge. Several treatment options (e.g. intralesional steroid injection, cryotherapy, surgical excision, silicone gel sheet and radiotherapy) had been tried with variable efficacies and complications.