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Adult T-cell leukaemia/lymphoma

Speaker: Dr. Yu Ho Tak, John Timothy
Medical and Health Officer, Social Hygiene Service, Department of Health, Hong Kong

A 58-year-old man presented to Tuen Mun Social Hygiene Clinic in July 2005 with a one year history of itchy rash on his trunk and limbs and weight loss. On examination, there were erythematous scaly patches and plaques on his trunk and limbs with lichenification. Enlarged cervical lymph nodes and hepatomegaly were noted. Blood test showed the presence of human T-cell lymphotropic virus type 1 (HTLV-1) antibody and borderline hypercalcaemia. Incisional skin biopsy and excisional lymph node biopsy were performed and they showed perivascular infiltrate of pleomorphic T-lymphocytes. Polymerase chain reaction for T-cell receptor gene rearrangement showed Jγ gene rearrangement and a diagnosis of adult T-cell leukaemia/lymphoma (ATLL) was made.

ATLL is a lymphoproliferative disorder aetiologically associated with HTLV-1 infection and was first identified in Japan as a distinct clinical entity in 1977. HTLV-1 is endemic in Southern Japan, Caribbean, South America and Central Africa. However, only a minor proportion of seropositive patients eventually develop ATLL. Clinical features include lymphadenopathy, organomegaly, hypercalcaemia and diverse skin lesions such as erythematous patches, plaques and papules. Histologically, diffuse infiltration of pleomorphic T-cells, which often display marked epidermotropism similar to mycosis fungoides, is seen. So far, there is no consensus on the best available therapy for ATLL and the prognosis is poor.

Learning points:
ATLL is an aggressive lymphoproliferative disorder associated with HTLV-1 infection. Its clinical and histological features are similar to mycosis fungoides. Hypercalcaemia is common in patients with ATLL but not in mycosis fungoides.

Indolent tender pigmented mass with progressive enlargement: an unusual case of dermatofibroma

Speaker: Dr. Chan Yee Ki
Medical and Health Officer, Social Hygiene Service, Department of Health, Hong Kong

Dermatofibroma is a common benign dermal tumour composed of fibrohistiocytic cells. Despite the fact that dermatofibroma is usually not difficult to diagnose, the deceptive cellular polymorphism and varying architectural pattern can sometimes be quite perplexing. Confusion may arise with respect to the biological behaviour of the lesion.

A 31-year-old Chinese woman presented with a pigmented mass on her left upper arm. Physical examination revealed a well circumscribed solitary nodule measuring 2.3 by 2 centimetres. It was bluish brown, scaly and fluctuant. No ulceration was noticed. There was no lymphadenopathy. Histopathological examination showed features consistent with fibrous histiocytoma (dermatofibroma).

The classical presentation of dermatofibroma is brownish firm papule (usually less than 5 mm in diameter) with positive dimple sign. Our case is a good illustration of how tricky the lesion can be as it is not a classical presentation of fibrous histio-cytoma (dermatofibroma). The differential diagnoses include benign and malignant lesions. Benign lesions include haemangioma, blue naevus and spitz naevus; malignant lesions include cutaneous B cell lymphoma, dermatofibrosarcoma protuberans, Kaposi's sarcoma and nodular melanoma. The highly variable clinical appearances could sometimes give rise to significant difficulty in diagnosis.

Learning points:
Dermatofibroma can sometimes have unusual presentation. Clinico-pathological correlation is essential and a high index of suspicion is required for diagnosing atypical cases of dermatofibroma.

Pathology of cutaneous vasculitis

Speaker: Dr. Lee Kam Cheong
Chief of Service, Department of Pathology, Princess Margaret Hospital, Hong Kong

Vasculitis can be defined as inflammation and destruction of blood vessels. The histological approach to evaluation of vasculitis is to determine whether vascular damage is present or not. Then, it is essential to identify the size and type of the vessels involved (Table 1) and the dominant inflammatory response preferably with a deep incisional skin biopsy. The most common vasculitic pattern seen in skin biopsy is leucocytoclastic vasculitis. Sometimes, the infiltrate may consist of lymphocytes, eosinophils and granulomas and their identification can lead to more specific diagnoses. However, biopsy features need to be interpreted together with the clinical history, physical and laboratory findings. In addition, the age of the cutaneous lesion biopsied and the possible effect of prior treatment must also be taken into consideration. It is preferable to biopsy cutaneous vasculitic lesions within 24 to 48 hours of onset. The speaker illustrated the pathology of various cutaneous vasculitis during the presentation.

Table 1. Chapel Hill consensus classification of vasculitis

Large-vessel vasculitis
- Giant cell arteritis
- Takayasu's arteritis

Medium-vessel vasculitis
- Classic polyarteritis nodosa
- Kawasaki disease

Small-vessel vasculitis
- Wegener's granulomatosis
- Churg-Strauss syndrome
- Microscopic polyangiitis
- Henoch-Schonlein purpura
- Essential cryoglobulinaemia
- Cutaneous leucocytoclastic vasculitis

Learning points:
It is essential to identify the size and types of vessels involved in vasculitis by obtaining a deep incisional skin biopsy. Other features such as the component of inflammatory cellular infiltrate, the presence of specific histological features and clinico-pathological correlation are useful for diagnosis of cutaneous vasculitis.

Redefining treatment expectation in moderate-to-severe psoriasis

Speaker: Professor Kristian Reich
Private Dermatologist, Dermatologikum, Hamburg, Germany

In Germany in 2005, a study had shown that 37% of patients attending dermatology outpatient clinic had a Dermatology Life Quality Index (DLQI) of greater than 10, i.e. psoriasis had a very large impact on a substantial proportion of patents. Conventional systemic treatments such as cyclosporine, methotrexate, retinoids and phototherapy have their own side effect profiles. The development of biologics such as alefacept, efalizumab, infliximab, etanercept and adalimumab provide new second line treatment options for recalcitrant moderate to severe psoriasis. In general, the followings can be considered in choosing psoriatic therapy: psoriasis area and severity index (PASI), nail or joint symptoms, age, sex, comorbidity or co-medication, previous response to therapy, side effect profiles and cost.

Infliximab, a tumour necrosis factor alpha inhibitor, given at 5 mg/kg intravenous infusion at week 0, 2 and 6 will give a 75% reduction in PASI score at week 10. This is accompanied by an improvement in DLQI as well. Patients with psoriatic arthropathy can also be potentially benefited from the use of infliximab. Subsequently, the speaker suggested that infliximab should be given at 5 mg/kg every 8 weeks as maintenance. Low dose methotrexate (7.5 mg/week) could be added to reduce the development of antibodies which occur in around 10% of patient on infliximab. Up till now, there is no head to head comparison between different biologics in terms of improvement in PASI.

Important side effects of infliximab include infection, infusion reaction, precipitation of heart failure and multiple sclerosis. Patient should have tuberculosis screening before infliximab and infusion reaction could be managed by decreasing the rate or stopping the infusion together with oral paracetamol and antihistamine.

Learning points:

Recalcitrant psoriasis can be treated with infliximab swiftly and effectively. Infliximab is effective in psoriatic arthropathy.

Successful outcomes in the treatment of melasma in Asians

Speaker: Dr. Chan Hin Lee, Henry
Honorary Clinical Associate Professor, Department of Medicine, University of Hong Kong, Hong Kong

Melasma can be classified into epidermal, dermal and mixed types. Increased number of melanocyte or increased melanogenesis is the postulated pathophysiological mechanism for melasma. Topical therapy remains the first line management. Triple combination including topical steroid, hydroquinone and topical retinoid can be used. This triple combination has less irritative side effects as compared with hydroquinone alone. The use of sunblock is also strongly advised. The degree of improvement is assessed by Melasma Area and Severity Index (MASI) which measures lesional area, darkness and homogenicity.

In one study comprising patients mainly of type IV skin, Tri-luma has been shown to be useful. Laser and intense pulse light are reserved for those resistant to topical therapy for more than 3 months. Post inflammatory hyper-pigmentation may occur in 10% of cases. Other options include fractional photothermolysis. It can be divided into scanning and stamping mode. Scanning mode is reliable, even and

faster. The disadvantages are staining, the need of a separate cooling device and only a single platform. Stamping mode has the advantages of single platform with multi-purposes, more effective cooling and hence less painful. However, up till now, there is no good meta-analysis of the treatment efficacy and long term adverse effect of this mode of treatment.

Learning points:

Melasma can be classified into epidermal, dermal and mixed types. Topical therapy remains the first line management for melasma and the use of sunblock is strongly advised. Non-pharmacological measures (laser and intense pulse light) are reserved for resistant cases.