

## Dermato-venereological Quiz

MLS Chiu 趙麗珊 and PCL Choi 蔡祥龍

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A 28 year old man presented with chronic bilateral ulceration of axillae with discharge (Figure 1) and acneiform eruptions on the scalp (Figure 2) for 3 years. Wound swab for bacteria and fungal culture

were negative. Skin biopsy was taken from the axillae. Immunohistochemical stains for CD1a (Figure 3) and S100 protein were performed (Figure 4).



**Figure 1.**



**Figure 2.**

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**Department of Medicine and Therapeutics, Prince of Wales Hospital, Hong Kong**

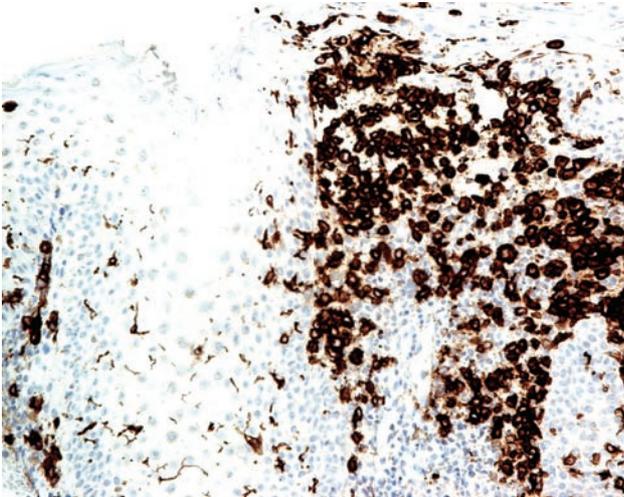
MLS Chiu, MBChB, MRCP(UK)

**Department of Pathology, Prince of Wales Hospital, Hong Kong**

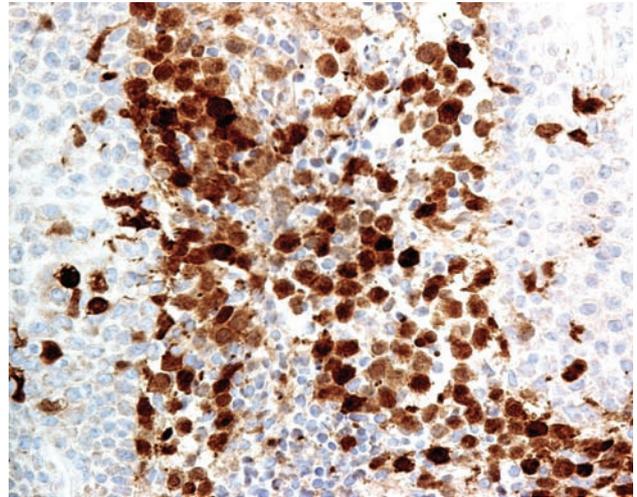
PCL Choi, FHKAM(Pathology)

Correspondence to: Dr. MLS Chiu

Department of Medicine and Therapeutics, Prince of Wales Hospital, 30-32 Ngan Shing Street, Shatin, N.T., Hong Kong



**Figure 3.** CD1a stain.



**Figure 4.** S100 stain.

## Questions

1. What are the clinical differential diagnoses?
2. The CD1a stain and S100 stain are both positive. What is the definitive diagnosis?
3. What is the characteristic electron microscopic feature?
4. Does he need further investigations?
5. What is the prognosis?
6. What is the treatment?

(Answers on page 60)

### **Answers to Dermato-venereological Quiz on pages 50-51**

1. The clinical differential diagnoses include seborrhoeic dermatitis, hidradenitis suppurativa, Darier disease, Hailey Hailey disease, Langerhans cell histiocytosis, and intertriginous psoriasis.
2. The diagnosis is Langerhans cell histiocytosis (LCH). The usual presentation is maculopapular eruptions followed by ulcerations with serous discharge at flexural areas. Sometimes, it may present as acneiform eruption. Another common presentation is diffuse erythematous scaly patches on the scalp which may or may not be accompanied by alopecia. It may be misdiagnosed as seborrhoeic dermatitis or scalp eczema, which are more common skin diseases. The presence of severe and haemorrhagic lesions may prompt one to think of the diagnosis.
3. Birbeck granules in the cytoplasm of lesional Langerhans cells.
4. Skin involvement occurs in 50% of patients. 68.6% of patients have multiple systems involvement. These include bone, lymph node, posterior pituitary (presenting as diabetes insipidus), ear, gum, lung and gastrointestinal tract in descending order of frequency. Therefore, further investigations are needed to rule out involvement of other organs.
5. Prognosis is excellent for single organ disease with a 100% five year survival rate and 91.7% even for those with multiple organ system involvement.
6. Various treatment modalities have been tried for this disease. PUVA and radiotherapy have been reported to be effective. Drug treatment which includes immunosuppressants such as methotrexate, azathioprine, and cyclosporine A, and etoposide based chemotherapy has also been tried with variable results. Systemic steroid and thalidomide have been reported to be useful as well. However, there is not yet any conclusion which treatment modality is better than the others.