

Case Report

Langerhans cell histiocytosis in a Chinese male patient misdiagnosed as hidradenitis suppurativa

男性華人患者朗罕氏細胞組織細胞增多症誤診為化膿性汗腺炎

MLS Chiu 趙麗珊 and PCL Choi 蔡祥龍

A 28-year-old Chinese gentleman presented with chronic bilateral axillary ulcers and discharge was diagnosed to have Langerhans cell histiocytosis. The clinical presentation mimics that of hidradenitis suppurativa. The definitive diagnosis can be made by distinctive histological findings.

28歲男性華人呈慢性雙側腋下潰瘍伴滲出，診斷為朗罕氏細胞組織細胞增多症。其臨床表現類似化膿性汗腺炎。其特徵性組織學表現可確立診斷。

Keywords: Hidradenitis suppurativa, Langerhans cell histiocytosis

關鍵詞：化膿性汗腺炎，朗罕氏細胞組織細胞增多症

Case report

A 28-year-old gentleman enjoyed good past health presented with chronic bilateral axillae ulcerations with discharge (Figure 1) for 3 years. He was treated as hidradenitis suppurativa with multiple courses of antibiotics. Roaccutane (Isotretinoin) was also given for a few months but was not very effective. His symptoms worsened

and pain limited his arm movement. Physical examination showed tender macerations, sinus



Figure 1. Serous discharge from the axillae that mimics hidradenitis suppurativa.

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

tracts formation with purulent yellowish discharge in both axillae. Extensive acneform eruption covered with scale and yellowish crust was noted on the scalp and pre-auricular area (Figure 2). A biopsy from the macerated lesion on the axilla showed mononuclear cell infiltrate involving both epidermis and upper dermis. Some of the mononuclear cells had reniform nuclei (Figure 3). These mononuclear cells also stained positively



Figure 2. Acneform eruption on the scalp the resembles scalp eczema.

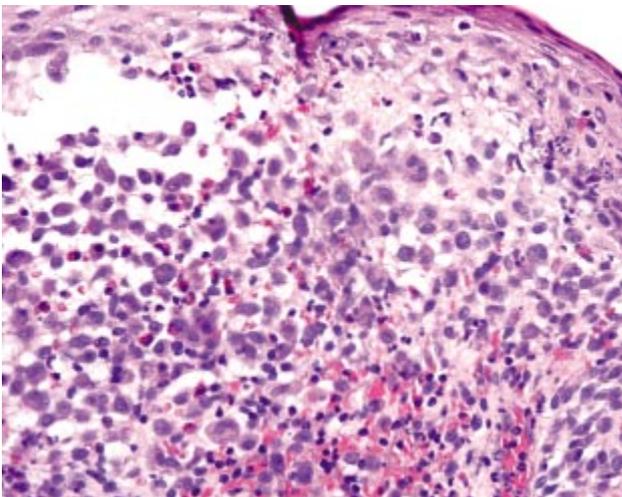


Figure 3. Medium power showed mononuclear cell infiltrate involving both epidermis and upper dermis. Some of the mononuclear cells had reniform nuclei. Noted also the scattered eosinophils. (H&E Original magnification x 20)

with CD1a and S-100 protein (Figures 4 & 5). Clinical and histopathological findings confirmed the diagnosis of Langerhans cell Histiocytosis. Wound swabs for bacteria and fungal culture were negative. The patient was referred to the oncology department. Systemic work up which includes blood test, skeletal survey, bone marrow and CT scan of abdomen were done and showed no abnormality. The patient was treated with

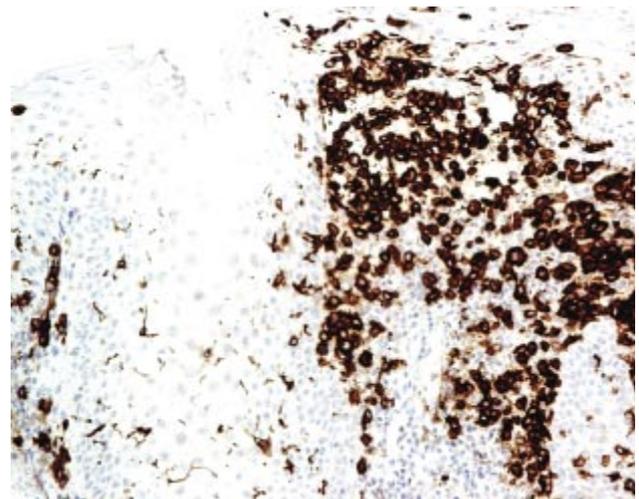


Figure 4. CD1a stain for Langerhan cells showed positive stain of the mononuclear cells. Noted also the normal dendritic form of Langerhan cells as positive control. (CD1a stain)

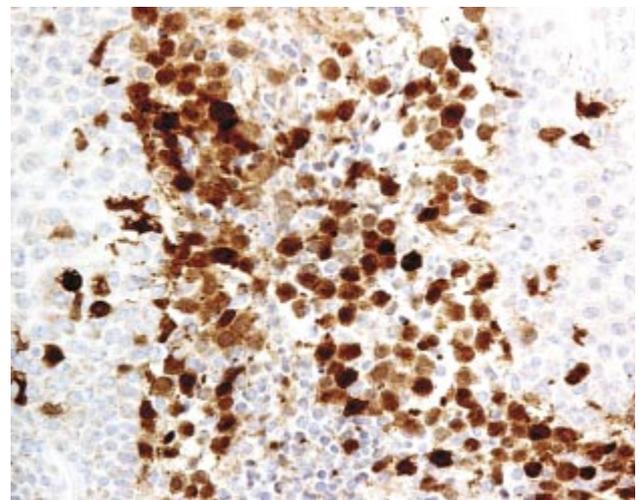


Figure 5. S100 protein stain showed both nuclear and cytoplasmic stains of the Langerhan cells. (S100 stain)

prednisolone 40 mg daily for 4 days and thalidomine 100 mg daily for one month with satisfactory response.

Discussion

Langerhans cell histiocytosis (LCH) is an accumulation or proliferation of a clonal population of cells bearing the phenotype of a Langerhans cell that has been arrested at an early stage of activation and is functionally deficient.¹ It is a rare disease. The incidence in children is 4-5 per millions. The incidence in adult is only 30% of that of children.² Skin involvement occurs in 50% of patients with LCH.² 68.6% of patients have multiple systemic involvement³ including bone, lymph node, posterior pituitary (presenting as diabetes insipidus), ear, gum, lung and gastrointestinal tract in descending order of frequency.⁴

The usual presentation is maculopapular eruptions followed by ulcerations with serous discharge at flexural areas but can be generalised. Sometimes, it may present as acneform eruption. Hidradenitis suppurativa (HS) is a more common disease which can present in the same way. It is difficult to distinguish between the two diseases without skin biopsy. Clinical suspicion was raised in our patient as he did not respond to HS treatment modalities. LCH can also present as diffuse erythematous scaly patches on the scalp with or without alopecia which may be misdiagnosed as seborrhoeic dermatitis or scalp eczema. The chronic discharge from flexural area mimics intertrigo. The discharge should be cultured to rule out bacterial or fungal infection. Other differential diagnosis includes Hailey Hailey disease and Darier disease.

The diagnosis of Langerhans cell histiocytosis can be confirmed by typical histological findings in skin biopsy. A definite diagnosis is characterised by (1) two or more positive stains for ATP-ase,

S-100, Alpha-D-mannosidase or Peanut lectin plus (2) Birbeck granules in the lesional cell under electron microscopy and or (3) Positive staining for CD1a on the lesional wall.⁴ Involvement of other organs should be ruled out. Prognosis is excellent for single organ disease with a 100% five year survival rate and 91.7% even for those with multiple organ involvement.² PUVA and radiotherapy have been reported to be effective treatment for treating LCH.⁵ Drug treatment which includes methotrexate, azathioprine, cyclosporine A, and etoposide have been tried with variable outcomes.⁵ Systemic steroid and thalidomide have been reported to be useful and were effective in our patient. This treatment regime for Langerhans cell Histiocytosis involving the skin in the future can be considered for future cases.

In summary, our patient presented with chronic bilateral axillary ulcers with discharge on the axillae and acneform eruptions on the scalp was diagnosed to have Langerhans cell histiocytosis. This condition can mimic a few common skin conditions such as seborrhoeic dermatitis, eczema and hidradenitis suppurativa. Clinicians should be aware of an diagnosis in recalcitrant cases of common skin diseases not responding to usual type of medications.

References

1. Chu T, D'Angio GJ, Favara BE, Ladisch S, Nesbit M, Pritchard J. Histiocytosis syndromes in children. *Lancet* 1987;2:41-2.
2. Arico M, Girschikofsky M, Genereau T, Klersy C, McClain K, Grois N, et al. Langerhans cell histiocytosis in adults. Report from the International Registry of the Histiocyte Society. *Eur J Cancer* 2003;39:2341-8.
3. Singh A, Prieto VG, Czelusta A, McClain KL, Duvic M. Adult Langerhans cell histiocytosis limited to the skin. *Dermatology* 2003;207:157-61.
4. Chu T. Langerhans cell histiocytosis. *Australas J Dermatol* 2001;42:237-42.
5. Munn S, Chu AC. Langerhans cell histiocytosis of the skin. *Hematol Oncol Clin North Am* 1998;12:269-86.