

Dermato-venereological Quiz

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A 5-month-old boy presented with generalized itchy blisters and hives since 6 weeks after birth. The lesions were found on the trunk, limbs and scalp and some were provoked by scratching. The mucosa and nails were not affected. Clinical examination showed multiple bullae and vesicles over the back. Erosions, wheals, and yellowish to tan coloured plaques were also seen (Figure 1). His parents noticed frequent flushing of the face, otherwise there was no other systemic symptom. His growth and development was normal. Skin biopsy was performed, which showed mononuclear cells with strongly positive chloroacetate esterase and c-kit (CD 117) stains (Figures 2 and 3).



Figure 1. Multiple vesicles, erosions, tan to yellowish plaques and wheals over the back.

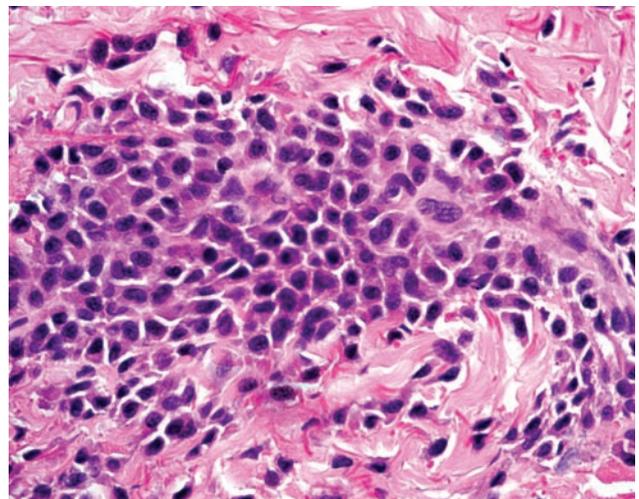


Figure 2. Mononuclear cells having a basophilic granular cytoplasm, some with round nuclei and others with carrot shaped nuclei (H&E stain, Original magnification x 600).

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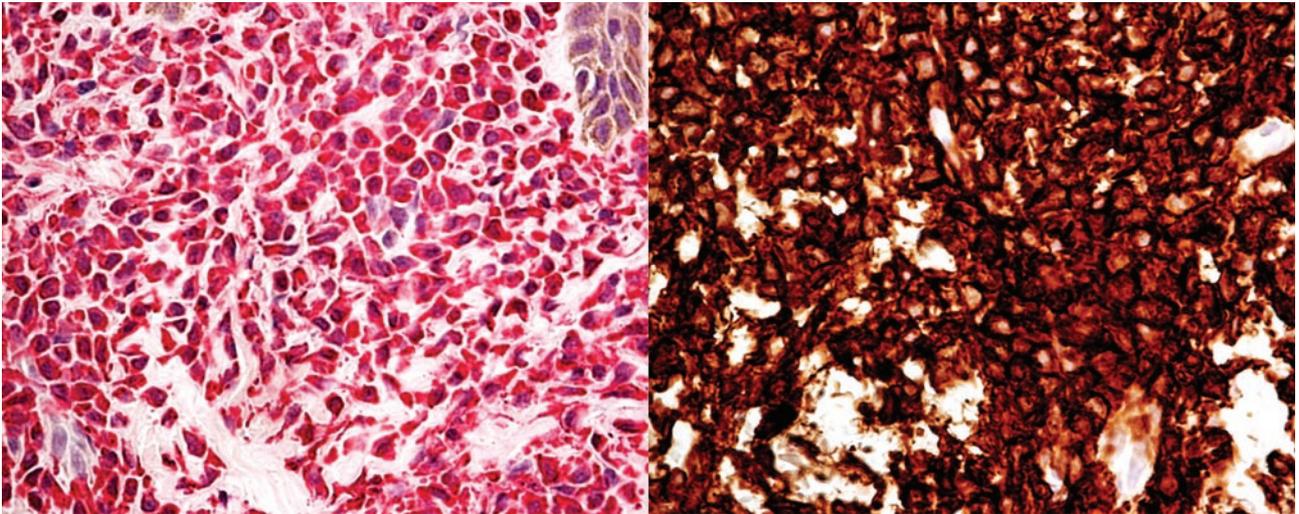


Figure 3. Chloroacetate esterase (left) and c-kit (CD117) (right) stains are both strongly positive.

Questions

- 1) What are the clinical differential diagnoses?
- 2) What are the histology findings?
- 3) What is the most likely diagnosis?
- 4) What important clinical sign can be demonstrated in patients suffering from this disease and how reliable is this clinical feature?
- 5) Is this disease related to any other serious and life-threatening conditions?
- 6) What are the treatment options?

(Answers on page 63)



Web sites of Dermatology & Venereology in Hong Kong

The homepage of The Hong Kong Society of Dermatology & Venereology
<http://www.medicine.org.hk/hksdv/>

Hong Kong Journal of Dermatology & Venereology
 (Official Publication of The Hong Kong Society of Dermatology & Venereology)
<http://www.medicine.org.hk/hksdv/bulletin.htm>

The homepage of The Asian Dermatological Association
<http://www.medicine.org.hk/ada/>

Answers to Dermato-venereological Quiz on pages xxx-xxx

- 1) The differential diagnoses include infiltrative diseases like mastocytosis, congenital diseases like epidermolysis bullosa, infective diseases like recurrent bullous impetigo, autoimmune bullous diseases such as linear IgA dermatosis, bullous pemphigoid, bullous lupus erythematosus and iatrogenic causes such as bullous drug eruption, and generalized contact allergic dermatitis.
- 2) The mononuclear cell infiltrates are mast cells that stain positive with chloroacetate esterase and c-kit (CD 117).
- 3) The diagnosis is mastocytosis. There are different types of cutaneous presentations. Our patient is pertaining to a rare subtype – diffuse cutaneous mastocytosis. Other more common presentations in paediatric patients are urticaria pigmentosa and mastocytoma. Telangiectasia macularis eruptiva perstans (TMEP) is mainly found in adults.
- 4) Darier's sign may be positive in patients with mastocytosis. The skin will be urticated after gentle stroking of the lesion. It is, however, not always present, particularly in adult patients with TMEP.
- 5) It is estimated that around 49% of adult and 9% of children may suffer from anaphylaxis. Systemic involvement such as bone marrow infiltration, hepatosplenomegaly, lymphadenopathy, abdominal pain and diarrhoea are more common in adult patients. Mast cell leukaemia and other haematological diseases such as myelodysplastic syndromes and myeloproliferative disorders are possible associations.
- 6) Patients and their parents should be educated to avoid possible triggers like drugs (non-steroidal anti-inflammatory drugs, aspirin, and narcotics), alcohol, heat and friction. First line treatment includes H₁ and H₂ histamine antagonists. Sodium cromoglycate, topical and systemic steroids, psoralen with UVA phototherapy have been reported to be effective.