Hong Kong J. Dermatol. Venereol. (2010) **18**, 164-165

Dermato-venereological Quiz

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A 31-year-old HIV-positive man on highly active antiretroviral therapy (HAART) was admitted for the treatment of cryptococcal meningitis and septicaemia. He was treated with intravenous amphotericin B and his CD4 count was 96 cells/μL. He presented with a small asymptomatic purplish papule on the right thigh for 2 months. There was no history of trauma, insect bite or contact with topical irritant. Apart from fever and headache, the patient did not have any systemic complaints. Clinically it was a well-defined purplish non-blanchable papule measuring 1 x 0.5 cm with scaling. There was neither satellite lesion nor sporotrichoid spread. The mucosal, genital areas and the rest of body were uninvolved. Baseline investigations including haemoglobin, platelet count, clotting profile, liver and renal functions were all within normal ranges. Excisional biopsy with wide margin was done.

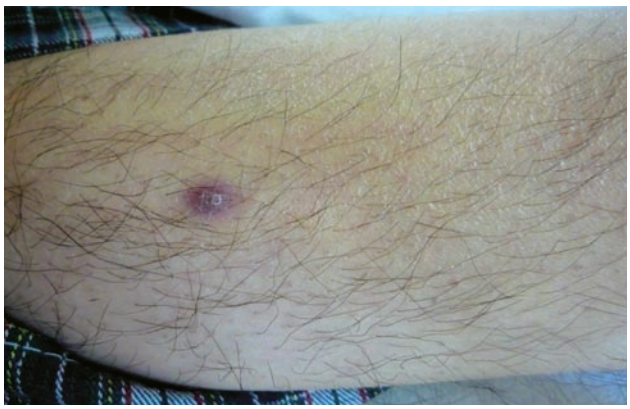

Figure 1. Well-defined purplish non-blanchable papule measuring 1 x 0.5 cm with scaling on the right thigh.

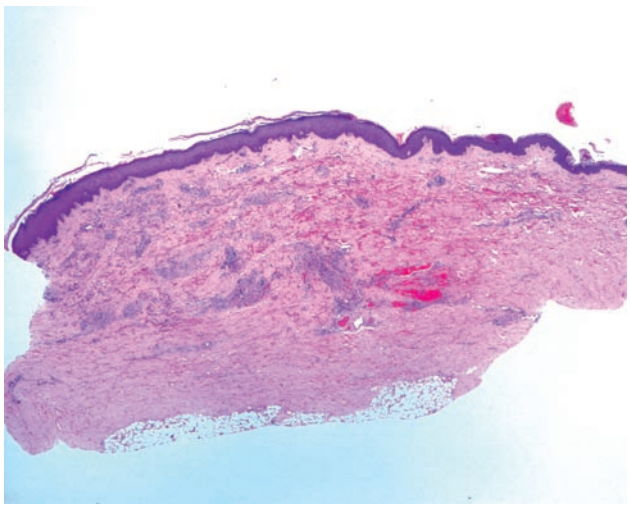

Figure 2. Histology showing dermal perivascular proliferation of spindle cells and extravasated red blood cells (H & E, original magnification x 20).

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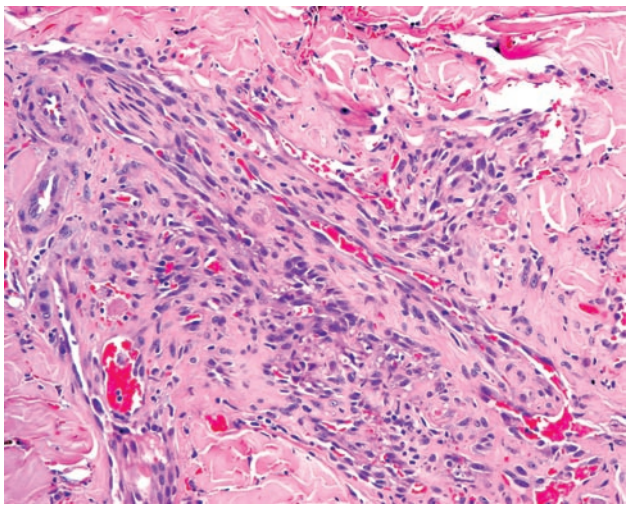

Figure 3. Increased vascular channels and protrusion into newly formed blood vessels lined by bland endothelial cells were seen (promontory sign) (H & E, original magnification x 200).

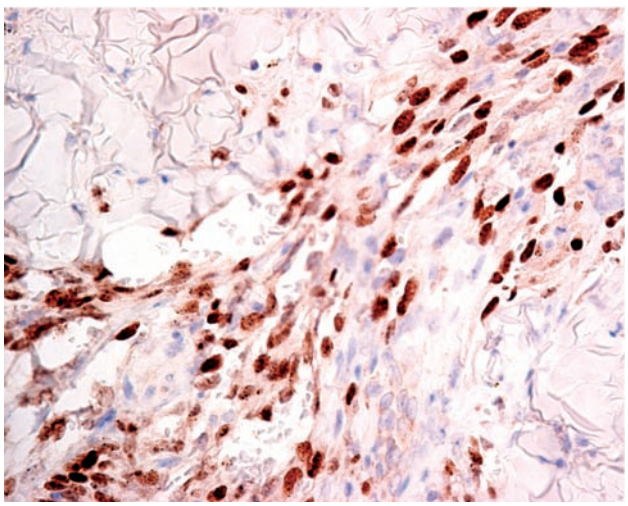

Figure 4. Human herpesvirus 8 immunohistochemical stain (original magnification x 400).

Questions

1) What are the differential diagnoses and likely clinical diagnosis?
2) What are the skin biopsy findings?
3) What other investigations should be done?
4) What are the treatment options?

(Answers on page 174)

Answers to Dermato-venereological Quiz on pages 164-165

1) The differential diagnoses include Kaposi's sarcoma, disseminated cutaneous cryptococcaemia, other deep fungal infection, bacillary angiomatosis, mycobacterial and atypical mycobacterial infection, and benign causes including prurigo nodularis, pyogenic granuloma or keloid. The likely clinical diagnosis is isolated cutaneous Kaposi's sarcoma associated with AIDS, given its purplish color, single lesion, lack of symptoms and minimal epidermal changes.

2) Histologically, the epidermis showed focal hyperkeratosis and focal acanthosis. There was a dermal proliferation of perivascular spindle cells with promontory sign, some containing hyaline globules, in a background of extravasated red blood cells, haemosiderin deposits and mixed inflammatory cells. Periodic acid Schiff stain did not demonstrate any fungal organism. Human herpesvirus 8 stain was positive in the nuclei of tumour cells. The histology was diagnostic of Kaposi's sarcoma.

3) Other staging investigations for the tumour include extensive oral mucosal examination, palpation for lymphadenopathy, chest X-ray, fecal occult blood and gastrointestinal endoscopic evaluation, which were negative in our patient.

4) Treatment of Kaposi's sarcoma depends on the staging and symptoms of disease.

   For localized isolated cutaneous lesion, local destructive therapy is preferred. Options include surgical excision, cryotherapy, topical 9-cis retinoic acid (alitretinoin gel 4 to 8 weeks), intralesional chemotherapy (vinblastin, vincristine or bleomycin), radiation therapy or photodynamic therapy.

   In patients with immunosuppression, local therapy has to be supported by immune-restoration through HAART in HIV-positive individuals or reduction in immunosuppression therapy in transplant-related Kaposi's sarcoma.

   For disseminated Kaposi's sarcoma, systemic therapy with liposomal anthracyclines has been shown to be most effective and well-tolerated. Other systemic chemotherapies include paclitaxel, bleomycin with vincristine and doxorubicin and interferon-alpha. Emerging treatments, for example, thalidomide, vascular endothelial growth factor and matrix metalloproteinase inhibitors have also been reported.