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Dermato-venereological Quiz

TW Tse 謝梓華 and LY Tse 謝麟英

A 82 years old Chinese male was admitted to orthopaedic ward for gouty attack. There was an incidental finding of hyperkeratotic lichenified erythematous lesion over the penile shaft and scrotal region which has been slowly growing over the past 5 years without major symptoms (Figure 1).

Questions

1. What are the clinical differential diagnoses?
2. Biopsy (Figure 2) showed parakeratotic skin with full thickness infiltration of the epidermis by large pale tumour cells with large nuclei, prominent nucleoli and cytoplasmic mucin (confirmed by PAS stain with diastase digestion). What other ancillary studies can be performed to confirm the diagnosis?
3. What further investigations are justified?
4. What should be the management of this condition?

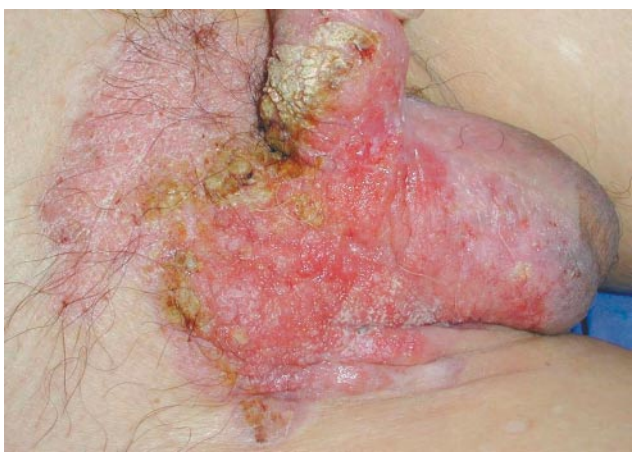

Figure 1.

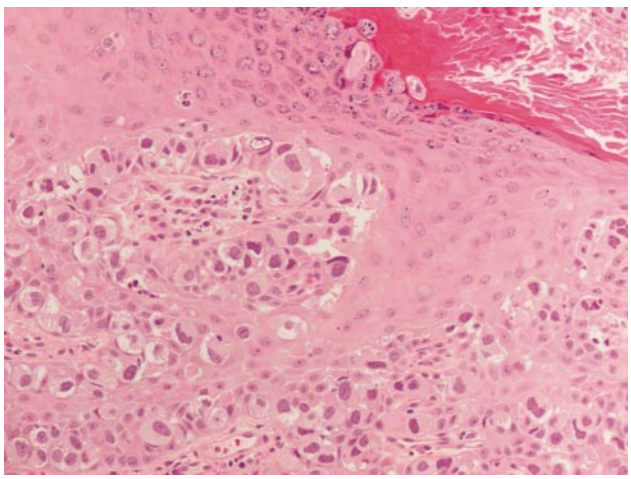

Figure 2. H & E stain. Pagetoid infiltration of epidermis by large carcinoma cells with pleomorphic hyperchromatic nuclei and prominent nucleoli.

Private Practice, Hong Kong

TW Tse, MRCP(UK), MSc Clin Derm(Lond)

Department of Pathology, Queen Elizabeth Hospital, Hong Kong

LY Tse, FRCPA, FHKAM(Path)

Correspondence to: Dr. TW Tse

Room 1502, Hang Lung Centre, 2-20 Paterson Street, Causeway Bay, Hong Kong

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1. The clinical differential diagnoses include squamous cell carcinoma, Bowen's disease, Paget's disease, condylomata acuminate, psoriasis, contact dermatitis, and mycosis fungoides.

2. Immunohistochemical studies can be performed. The tumour cells express monoclonal carcinoembryonic antigen and low molecular-weight cytokeratin CAM 5.2 but not S100. In Figure 3, the carcinoma cells were highlighted by immunostaining for CAM 5.2 against a negative background of squamous cells in the epidermis. This confirms the diagnosis of Penoscrotal Extramammary Paget's Disease (EMPD).

   EMPD is a rare intraepidermal neoplasm primarily found in areas of skin bearing apocrine glands. Patients characteristically have undergone all sorts of topical therapies without success until the diagnosis is made histologically.

3. Among the penoscrotal EMPDs that were associated with an internal malignancy, 92% of the underlying tumours were genitourinary tract tumours including prostate adenocarcinoma,[1] transitional cell carcinoma[2] and renal cell carcinoma. Therefore, further investigation for genitourinary cancer is justified.[3,4] In this patient, urine for cytology, intravenous urogram, ultrasound of kidneys and prostate surface antigen were all negative.

4. EMPD should be treated with wide local excision. The large skin defects were immediately reconstructed with split-thickness skin graft or local skin flap.[3] Prognosis of primary EMPD confined to the epidermis is excellent but those with dermal invasion will depend on the depth of the lesion.[5] For secondary EMPD, the prognosis depends on the underlying neoplasm. Other treatment options for EMPD include radiotherapy, systemic chemotherapy, laser and photodynamic therapy.

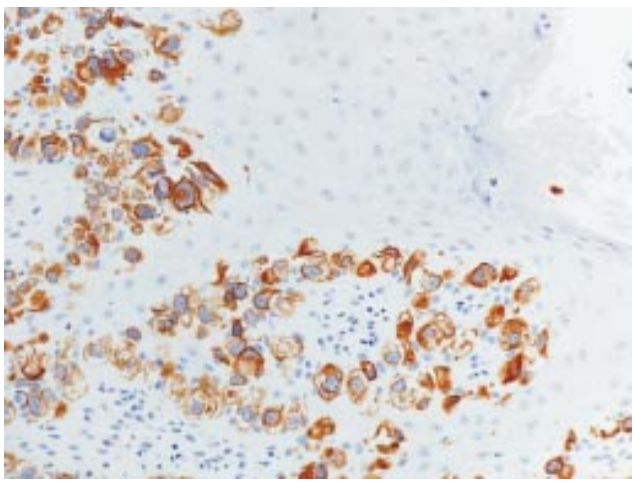

Figure 3. Immunohistochemical studies. The tumour cells express CAM 5.2.

References

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