A 76-year-old lady presented with an erythematous nodule over her right forearm for five months. It was neither painful nor itchy. Physical examination showed a non-tender 2 x 3 cm erythematous nodule over the right forearm (Figure 1).

She was afebrile and did not have any systemic symptom. Incisional skin biopsy was performed with histological findings as shown in Figures 2 and 3.
Questions

1) What are the possible clinical differential diagnoses?
2) What is the diagnosis?
3) What are the histological differential diagnoses?
4) What are the risk factors for this condition?
5) What are the treatment options and prognosis?

Figure 3. CK-20 stain showed paranuclear dot-like positivity.

(Answers on page 233)
Answers to Dermato-venereological Quiz on pages 221-222

1) Differential diagnoses include squamous cell carcinoma, metastatic small cell lung carcinoma, lymphoma, amelanotic melanoma, basal cell carcinoma, keratoacanthoma, merkel cell carcinoma.

2) With the clinical presentation, histology of small blue round cell tumour positive for CK-20 and negative finding when searched for carcinoma of the lung, the diagnosis is merkel cell carcinoma. It is a rare neuroendocrine malignancy of the skin.

3) The pathology of merkel cell carcinoma falls into the category of small blue round cells tumours. It is difficult to differentiate it from other metastatic neuroendocrine tumours especially small cell lung carcinoma (SCLC). Immunohistochemical stains are helpful to confirm the diagnosis. CK-20 is a very sensitive marker for merkel cell carcinoma but it can also be positive in SCLC. Its use, in combination with thyroid transcription factor 1 (TTF-1) which stains positive in SCLC but is absent in merkel cell carcinoma, has made the histopathological diagnosis of merkel cell carcinoma more accurate.

4) Those who have treatment with psoralens and ultraviolet A phototherapy, or those who have received organ transplant, or HIV patients have higher risk of developing merkel cell carcinoma. Recent research showed that polyomavirus may play a role in the pathogenesis based on the finding that the virus is found in most of the merkel cell carcinoma.

5) Wide local excision with margin of 2-3 cm is the recommended treatment. Radiotherapy and chemotherapy can be considered as adjuvant treatment in advanced disease. The local recurrence rate is high which is estimated to be around 44%. The overall 5 year survival rate is around 50-68%.