

## Reddish Tumour with Scar on the Right Thigh: A Quiz

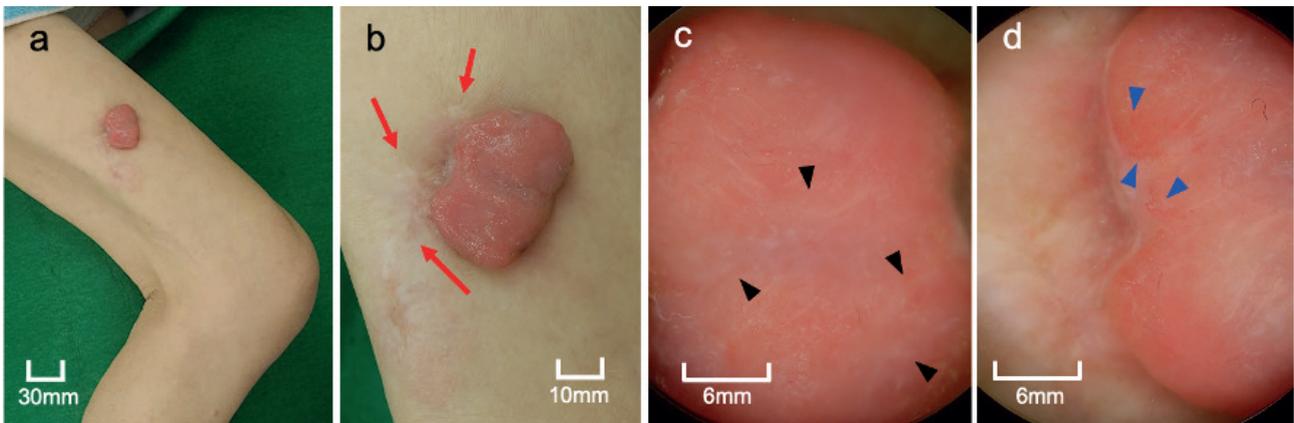
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An 85-year-old female patient was referred to our Department of Dermatology, Shimane University Hospital, Izumo, Japan, for the presence of a tumour 30 mm in diameter on her right thigh. She had noticed 2 rice grain-sized asymptomatic nodules 8 years previously, which had gradually enlarged. The 2 nodules subsequently became a single tumour. She was found to have a lung shadow at a check-up 4 months previously; therefore, she underwent further examination with positron emission tomography-computed tomography (PET-CT). The findings revealed accumulation of 18F-fluorodeoxyglucose in the tumour site of the right thigh, and the patient was treated with topical drug, Gentamicin Sulfate Ointment at a local internal medicine clinic.

She had visited a local dermatology clinic because the mass in her right thigh had been increasing in size for 1 month, and she was referred to our department with a suspected diagnosis of squamous cell carcinoma. A coin-sized reddish tumour with scarring was found on the lateral side of her right thigh (Fig. 1a, 1b), and a bean-sized lymph node was palpable in her right groin. Dermoscopy revealed a generally unstructured and homogeneous salmon-pink coloured finding (Fig. 1c) with an indistinct white linear structure (Fig. 1d). In the marginal region, there was a slight linear red structure that appeared to correspond to the serpentine linear vessels.

*What is your diagnosis? See next page for answer.*



**Fig. 1.** (a) Reddish tumour 30 mm in diameter on the lateral side of the right thigh. (b) The tumour was partially surrounded by scars (→). (c) Dermoscopy findings showing a generally unstructured and homogeneous salmon-pink coloured finding with an indistinct white linear structure (▶). (d) In the marginal region, a slight linear red structure was seen, which appeared to correspond to the serpentine linear vessels (▶).

## ANSWERS TO QUIZ

## Reddish Tumour with Scar on the Right Thigh: A Commentary

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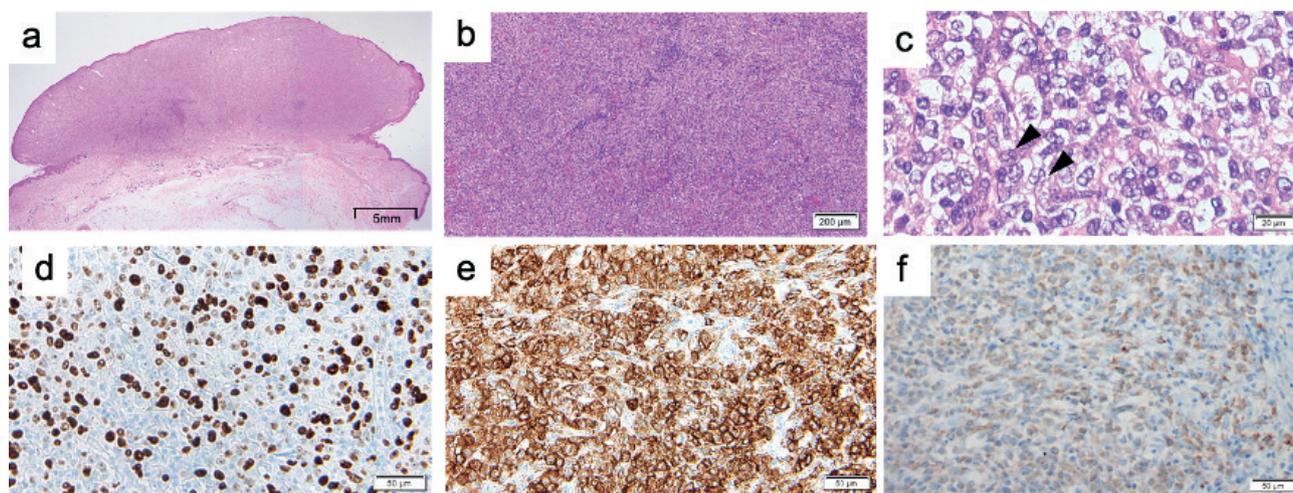
### Diagnosis: Langerhans cell sarcoma

Total excisional biopsy revealed that the lesion was located in the dermis, and the epidermis was compressed and thinned by the lesion (Fig. 2a). In the lesion, atypical cells ranging from round to short fusiform had diffusely and densely proliferated (Fig. 2b, c). The atypical cells had bright cytoplasm, the connectivity between the atypical cells and the boundary between cells was unclear, and the nuclear fission has been observed (Fig. 2c). Tumor cells with prominent nucleoli and coffee-bean-like grooves were found intermingled. The Ki-67 index was approximately 50% positive (Fig. 2d). Immunostaining was negative for S-100 protein, but positive for CD1a (Fig. 2e) and CD207 (Fig. 2f). Based on these findings, the tumour was diagnosed as a Langerhans cell sarcoma (LCS). Extended resection of the tumour, with a 2-cm margin, and right inguinal and external iliac lymph node dissection were performed. The primary lesion had been removed, however 13 of 17 right inguinal lymph nodes were positive for metastasis, and 3 of 4 external iliac lymph nodes were positive. We are under observation without additional treatment at the patient's request.

Dermoscopy is used mainly for diagnosis of pigmented lesions. However, it has recently been applied to diseases other than pigmented lesions (1–5). To the best of our knowledge, this is the first case in which dermoscopy findings of LCS have been demonstrated. LCS is an extremely rare malignant tumour derived from Langerhans cells, characterized by positive immunostaining for CD1a, S-100 protein,

and CD207 (6). The tumour cells are characterized by a high Ki-67 index, together with malignant findings including strong atypia and mitotic figures. When the tumour originates in the skin, it shows reddish protuberant plaques, red nodules, and papules and is often accompanied by erosion and ulcer (7, 8).

The lesion in this case was clinically suspected to be squamous cell carcinoma (SCC) or eccrine porocarcinoma or malignant lymphoma. Recently, haemorrhage, keratinization, and vascular features (glomerular, hairpin, and linear irregular morphologies) on dermoscopy have been reported to be useful for diagnosing nodular SCC (1). Rosendahl et al. (2) reported that coiled vessels are a strong indication of SCC compared with basal cell carcinoma, but are not helpful when the differential diagnosis is actinic keratosis and Bowen's disease. Lallas et al. (3) tried to specify the dermoscopic criteria that enable distinguishing highly and poorly differentiated SCCs in vivo. Poorly differentiated SCCs showed predominantly red colour, attributed to the absence of scaling and keratin and the presence of bleeding and ulcerations or dense vascularity. The quantity and calibre of vessels correlated significantly with the grade of differentiation. If vessels covered more than 50% of the lesion and small-calibre vessels dominate, then the lesion was more likely a poorly differentiated SCC. The presence of keratin, white circles, and white structureless were in favour of moderately or highly differentiated SCCs. Edamitsu et al. (4) reported that 7 of 8 cases with an eccrine porocarcinoma exhibited a polymorphous vascular pattern, consisting mainly of hairpin, linear-irregular, and dotted vessels. A combination of round-to-oval pink-white structureless areas and white-to-pink halo was observed in 5 of 8 cases, with 1 case showing the white-to-pink halo alone. Sławińska et al. reported that the structures most commonly observed in classical mycosis fungoides were fine short linear vessels/linear vessels, spermatozoa-like vessels,



**Fig. 2.** (a) Skin excisional biopsy from the right thigh revealed an elevated tumour approximately 3 cm in size, with the lesion being present in the dermis (haematoxylin and eosin (H&E) staining; original magnification:  $\times 1$ ). (b) The tumour is composed of dense, diffuse proliferation of atypical cells ranging from round to short fusiform (H&E staining; original magnification:  $\times 40$ ). (c) Atypical cells have bright cytoplasm, and the connectivity between atypical cells and the boundaries between cells is unclear; and the nuclear fission has been observed. Tumor cells with prominent nucleoli and coffee-bean-like grooves were found intermingled (▶) (H&E staining; original magnification:  $\times 400$ ). (d) The Ki-67 index was approximately 50% positive (original magnification:  $\times 200$ ). The tumour cells were diffusely and highly positive for CD1a (e) and CD207 (f) (original magnification:  $\times 200$ ).

and orange-yellow patchy areas (5). Primary cutaneous marginal zone B-cell lymphoma and primary cutaneous follicle centre lymphoma most commonly appeared against a salmon-coloured background and with fine short/linear irregular/serpentine vessels. In the current case, dermoscopy revealed poorly keratinized, poorly vascularized, generally unstructured, homogeneous, salmon-pink findings with an indistinct white linear structure. This suggests that dermoscopy findings could be used to differentiate LCS from SCC, eccrine porocarcinoma, and lymphoma. Proliferation of tumour cells in LCS may compress all of the surrounding structures, including blood vessels, resulting in uniform, non-structural dermoscopic findings.

Satolli et al. (9) reported that yellow colour and homogeneous distribution, as observed on dermoscopy, can facilitate the complex diagnosis in histiocytic sarcoma (HS). They described the dermoscopy image of HS as a “yellow submarine”. HS is an extremely rare, non-Langerhans cell tumour and is usually diagnosed at an already advanced clinical stage. It is associated with a high mortality rate even today. LCS, like HS, is extremely rare and is usually diagnosed at an already advanced clinical stage, and it is also associated with a high mortality rate (10).

In conclusion, we report here a case of LCS that showed characteristic findings of homogeneous distribution and salmon-pink coloration on dermoscopy. To date, there is very little information about the dermoscopy findings of LCS, and hence these results may be useful for the early detection and diagnosis of LCS.

*The authors have no conflicts of interest to declare.*

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