Title: A superficial leiomyosarcoma histopathologically mimicking dermatofibroma: pitfall in the diagnosis

Short title: Dificulty in diagnosis of superficial leiomyosarcoma

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Dear Editor,

A 21-year-old Japanese woman was referred to our department for evaluation of a nodule on her leg. She had been aware of the lesion for 1 month. Physical examination revealed a reddish-brown nodule, 11×8 mm in diameter, located on her left lower leg (Fig. 1a). Dermoscopic examination showed peripheral brown pigmentation with yellowish-white keratotic structures on the central surface (Fig. 1b). The first biopsy was incisional and performed on the periphery of the lesion. Histopathological examination revealed elongation of rete ridges and diffuse proliferation of spindle cells in the whole dermis with collagen trapping in the periphery (Fig. 1c). At this point, a diagnosis of dermatofibroma was made. However, when she came for operation after four months, ulceration was seen on its surface (Fig. 1d). Histopathologically, it showed massive proliferation of atypical spindle-shaped cells with elongated, cigar-shaped nuclei from the upper dermis to the subcutaneous tissue and dilation of capillary vessels and hemorrhage in the peripherally superficial dermis (Fig. 1e). Immunohistochemically, the spindle tumor cells were positive for anti-alpha smooth muscle actin (α -SMA) (Fig. 1f) and calponin (not shown) but negative for S-100 protein and CD34. Finally, a diagnosis of superficial leiomyosarcoma (LMS) was made.

LMS accounts for 23.9% of all soft tissue sarcomas.[1] Superficial LMS arising from the

dermis is a rare tumor that accounts for 2-3% of all soft tissue sarcomas. Superficial LMS is classified in two subtypes: dermal LMS originating from the arrector pili muscles of the hair follicles and subcutaneous LMS arising from vascular smooth muscle of cutaneous adipose tissue.[1, 2] Clinically, the tumor presents as an indolent exophytic or subcutaneous, firm, violaceous or purpule-black small nodule on the extensor of the extremities. The overlying epidermis sometimes shows erosion or ulceration. Histopathologically, the tumor presents with a poorly circumscribed uncoated lesion composed of a fascicular proliferation of spindle cells with elongated or cigar-shaped nuclei.[2, 3] Immunohistological staining with α-SMA or calponin is useful for its diagnosis.[4,5] In our case, clinical features and histopathological and immunohistochemical findings of the resected specimen were typical for dermal LMS.

As far as we know, there has been only one reported case of dermoscopic findings for cutaneous metastasis of LMS. Chernoff et al. reported that a vascular pattern was seen in a nonpigmented metastatic lesion of LMS.[5] However, there has been no report on the dermoscopic features of primary superficial LMS.

In our case, dermoscopic examination showed a peripheral brown pigmentation, which corresponded to dilation of capillary vessels and hemorrhage in the tumor. We considered that the yellowish-white keratotic structures on its central surface were

derived from regenerating epithelium or crusts.

Based on our findings, we consider that it is difficult to make a correct diagnosis of superficial LMS by dermoscopy and even by incisional biopsy. We emphasize that it is important for clinicians to include superficial LMS in differential diagnosis of a nonspecific ulcerative nodule histopathologically composed of spindle cells.

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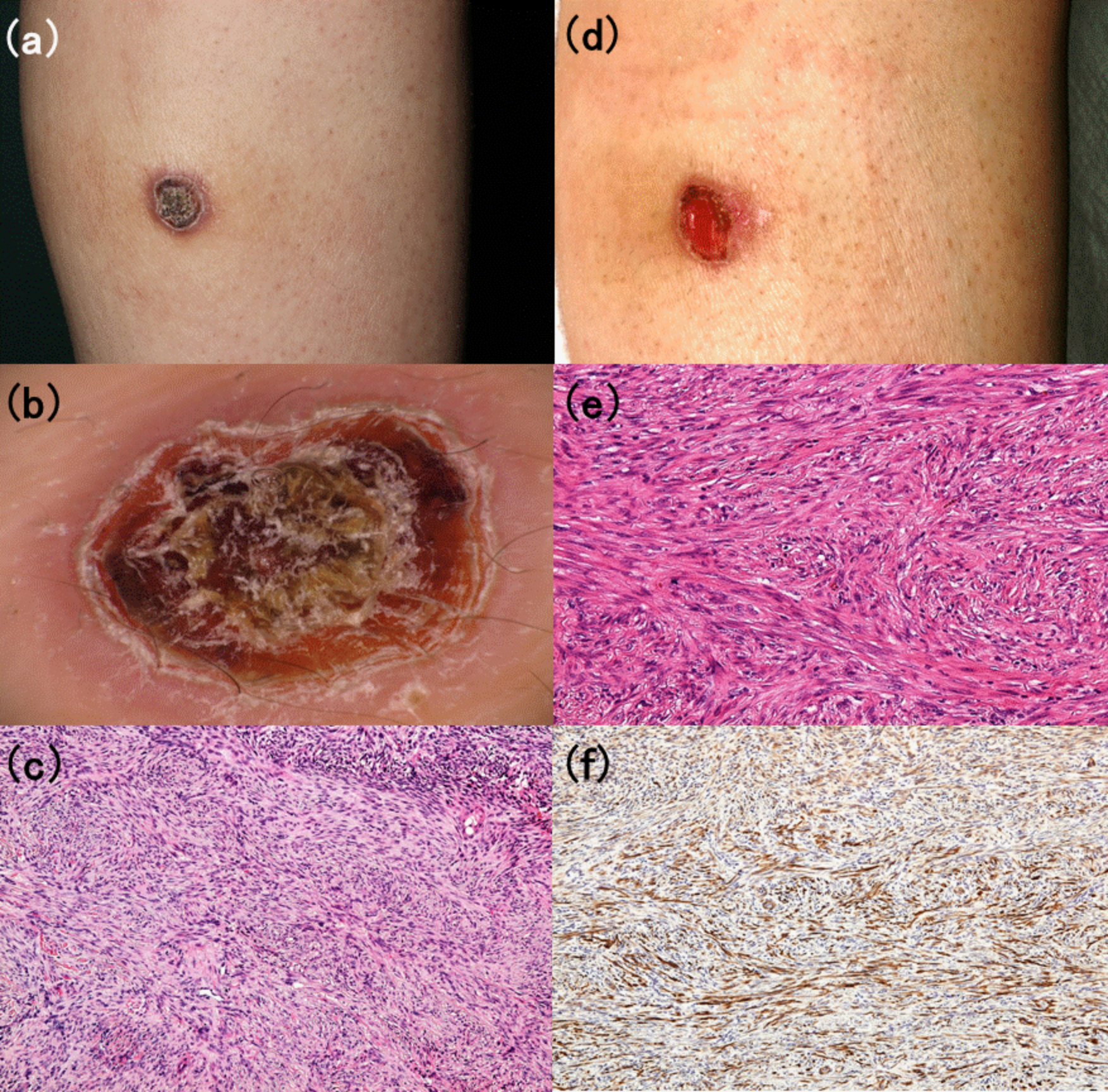
Figure Legends

Figure 1 (a) A brownish-red nodule on the left lower leg. **(b)** The lesion showed brown-red pigmentation with yellowish-white keratotic structures on its central surface by dermoscopy. **(c)** Diffuse proliferation of fibroblast-like spindle cells in the whole dermis (haematoxylin-eosin [HE], original magnification × 100). **(d)** Ulceration was seen on its surface after four months. **(e)** Massive proliferation of atypical spindle-shaped cells with cigar-shaped nuclei from the upper dermis to the subcutaneous tissue (HE, original magnification × 200). **(f)** Positive immunostaining for α-smooth muscle actin (SMA) (original magnification × 100).


(a)
(d)
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