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Dermatoscopic findings of atrophic dermatofibrosarcoma protuberans

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ABSTRACT Dermatofibrosarcoma protuberans (DFSP) is an uncommon locally aggressive mesenchymal tumor with a high local recurrence rate. Atrophic DFSP is a rare variant of DFSP characterized by a non-protuberant lesion. We report on a 23-year-old female, who presented with an atrophic, asymptomatic macule on the right side of her back 2 cm in diameter. Dermatoscopic examination revealed homogenous pigment network on a purplish erythematous background. The histopathological finding of the incisional biopsy material was consistent with DFSP. To our knowledge, this is the second case of atrophic DFSP discussing the dermatoscopic features of this relatively rare condition.

Case presentation

A 23-year-old female presented with an asymptomatic, erythematous, atrophic macule on the upper right side of her back that increased gradually in size within the last four years without any symptoms (Figure 1A). On physical examination the surface of the lesion was smooth and no induration was examined on palpation. There was no history of trauma. Dermatoscopic evaluation of the lesion using 3Gen DermLite-II Pro HR and documented with DermLite-Foto 3Gen (LLC, Dana Point, CA, USA) showed a homogenous pigment network on a purplish erythematous background (Figure 1B). A skin biopsy was taken with the preliminary diagnosis of morphea and cutaneous mastocytosis. Histopathological examination of H&E (hematoxylin and eosin) stained sections revealed epidermal atrophy, increase in melanin in basal keratinocytes and heavy dermal cellular infiltrate composed of spindle-shaped cells arranged in a storiform pattern that extended into the subcutaneous tissue (Figure 2). Immunhistochemical staining for CD34 was positive, while factor XIIIa was negative (Figure 3A, B). The diagnosis of dermatofibrosarcoma protuberans (DFSP) was made according to these findings. The patient was referred to Department of Plastic and Reconstructive Surgery for total excision. Excision was

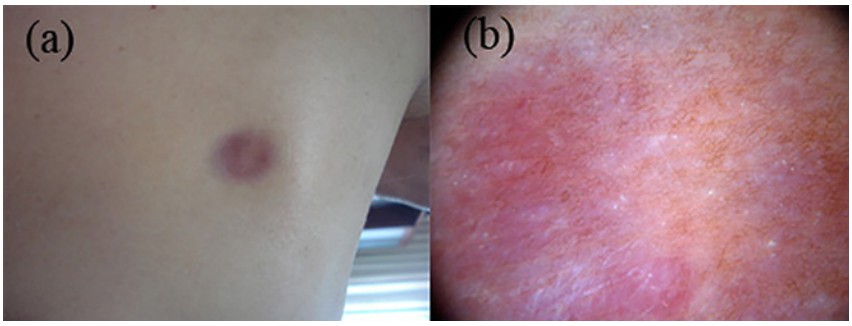

Figure 1. (A) Erythematous, atrophic macule on the right side of upper aspect of the back; (B) dermatoscopic evaluation that shows homogenous pigment network on a purplish erythematous background. (Copyright: ©2015 Akay et al.)

made with 3 cm safe surgical margins. No recurrence has been observed for one year.

Conclusion

DFSP is a rare, slow growing, locally invasive cutaneous neoplasm of fibrohistiocytic origin with intermediate grade malignancy with an incidence between 0.8–5.0 cases per 1 million persons per year [1]. It often shows typically protuberant morphology. The atrophic presentation of DFSP is the rarest variant of this infrequent neoplasm, which was first described in 1985 [2]. The non-protuberant DFSP can be considered the early clinical stage of DFSP before developing the typical protuberant feature unless it remains as a non-protuberant tumor that is called atrophic DFSP. It occurs as an atrophic, asymptomatic plaque that can be difficult to distinguish from morphea, morphea like basal cell carcinoma, scar, lipoatrophy and atrophic dermatofibroma [3]. Although the clinical appearance of atrophic DFSP is different from common protuberant type, histopathological features are similar. Atrophic presentation of the lesions may be due to the marked cellularity of the tumor cells in the dermis and infiltration to the subcutaneous fat [1]. Immunhistochemical staining is important to distinguish DFSP from atrophic dermatofibroma and medallion-like dermal dendrocyte hamartoma. While DFSP cells are positive for CD34 and negative for factor XIIIa, atrophic dermatofibroma cells are usually negative for CD34 and both of CD34 and factor XIIIa are positive in the medallion-like dermal dendrocyte hamartoma [3, 4].

Dermatoscopic studies of DFSP are very rare in the literature. In 2013, Bernard et al reported the first study of dermatoscopic analysis of DFSP in 15 cases. Delicate pigment network, vessels, structureless light brown areas, shiny white streaks, pink background coloration and structureless hypopigmented or depigmented areas were defined as six main dermatoscopic features of DFSP [5]. In 2014, dermatoscopic features such as peripheral dilated vessels forming a mesh-like pattern, milky-red areas, whitish linear structures and fine pigment network were reported in a series of four cases of DFSP [6]. To our knowledge, only one case of atrophic DFSP was presented with dermatoscopic features in the literature [4]. The reported dermatoscopic findings of that case were branching vessels on a yellowish background without pigment network. The authors suggested that the dermatoscopic features might be the result of dermal atrophy and close approximation the subcutis to the epidermis. In contrast, our case showed

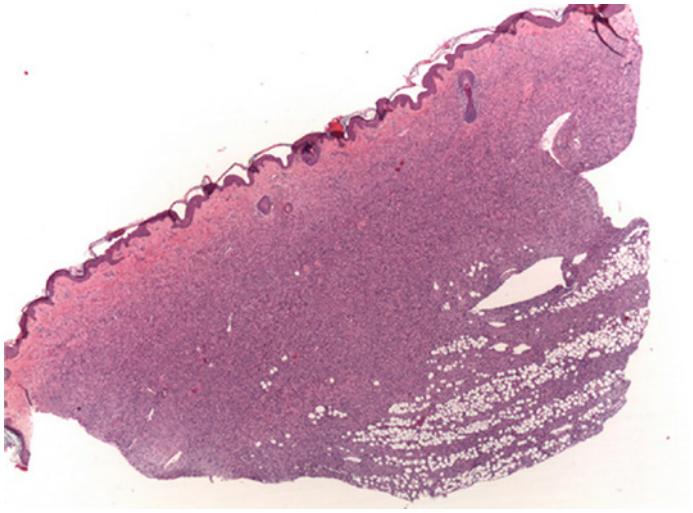

Figure 2. Heavy dermal cellular infiltrate composed of spindle shaped cells arranged in a storiform pattern that extended into the subcutaneous tissue (H&E, X25). (Copyright: ©2015 Akay et al.)

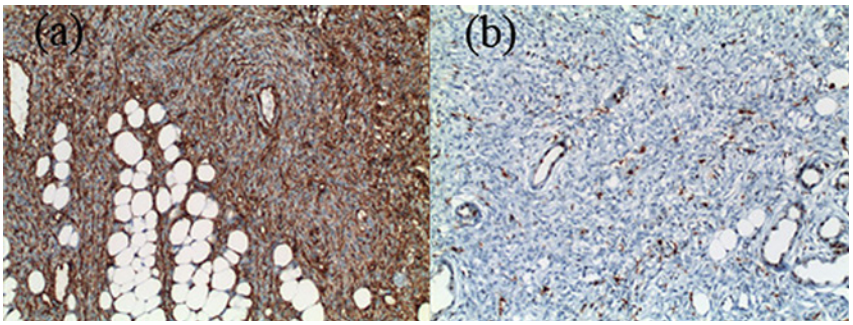

Figure 3. (A) Positive immunhistochemically staining for CD34 (X200); (B) negative immunhistochemically staining for factor XIIIa (X200). (Copyright: ©2015 Akay et al.)

dermatoscopically regular brown lines reticular on a purplish erythematous background. Homogenous brown lines reticular seen in our case correspond to the accumulation of melanin in basal keratinocytes, while the erythematous background is the result of dilated vessels in the dermal plexus. Here, the smooth surface reflects the stratum corneum being normal. The most frequent dermatoscopic pattern associated with dermatofibromas, a well-known and benign cousin of DFSP, is the central white scar-like patch and peripheral delicate pigment network [7]. The histopathologic correlation of white scar-like patch in dermatofibroma is pronounced fibrosis within the papillary dermis. Contrary to dermatofibromas, fibroplasia in the upper half of the dermis is uncommonly observed in DFSP which may explain the absence of white scar-like areas in our case.

In conclusion, we have presented the dermatoscopic findings in an unusual case of atrophic DFSP. Atrophic DFSP should be kept in the differential diagnosis for atrophic and depressed skin lesions, particularly those seen on the trunks of women. Dermatoscopy, a noninvasive method, may not only help to differentiate atrophic DFSP from other skin diseases but may also indicate the need for histopathological examination since the disease has prognostic significance.

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