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Dermatoscopy and Histopathology Correlates of Dermatofibrosarcoma: A Case Report

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Abstract

DFSP is a soft tissue malignant tumor the diagnosis of which is often delayed because of its nonspecific clinical features in early stages of disease. A few clinical investigations have detected several useful diagnostic parameters, including delicate pigmented network, vessels, structureless light brown areas, shiny white streaks, pink background coloration, and structure less poor depigmented areas. In particular, the combination of unfocused vessels with reticulate pigmentation has been suggested as a more distinct pattern of DFSP compared with other skin tumors. The detection of these dermoscopic features could be particularly useful on black skin to recognize DFSP early and to distinguish it from keloids.

Keywords: ***Dermatofibrosarcoma; Dermoscopy; Nodule; Soft tissue tumor***

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Introduction

Dermatofibrosarcoma protuberans (DFSP) is a relatively uncommon fibrohistiocytic tumour of intermediate malignancy. Despite its low metastatic potential, DFSP implies high morbidity due to its great capacity for local infiltration and a high recurrence rate after surgical excision.

Dermoscopy is a non-invasive technique that can be helpful in the diagnosis of skin tumours. We present a case of DFSP with histological confirmation and describe their dermoscopic features.

Case Report

A 53-year-old woman presented with a history over several years of a 40 mm × 25 mm brownish multifocal irregular plaque surmounted by an angiomatous nodule on her trunk (Figure 1). Dermoscopy revealed a central fine pigment network, multifocal milky-red areas, whitish linear structures and peripheral dilated vessels with a mesh-like appearance (Figure 1). A

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histopathological examination showed spindle shaped tumour cells in a storiform pattern invading the dermis and superficial hypodermis. (Figure 2 and Figure 3) Immunostaining was considered positive for CD34 (Figure 4). These findings were consistent with the diagnosis of DFSP. Recurrence has not been observed 3 years after a wide local excision.

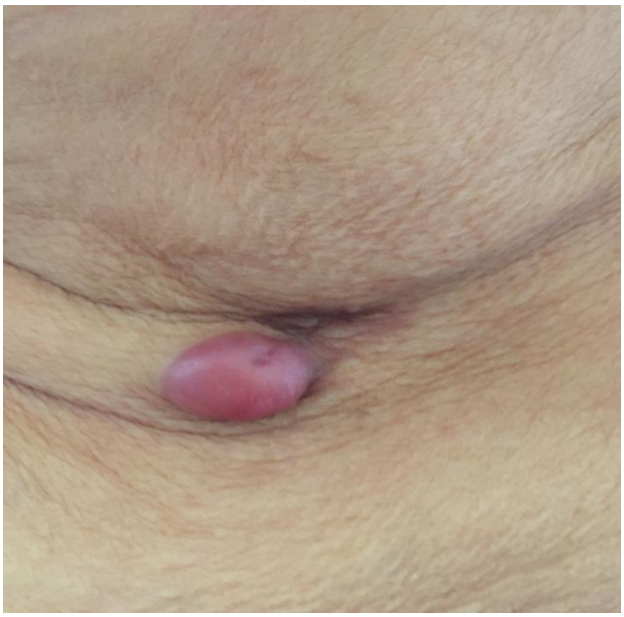

Figure 1: Para-umbilical pigmented plaque surmounted by an angiomatous nodule.

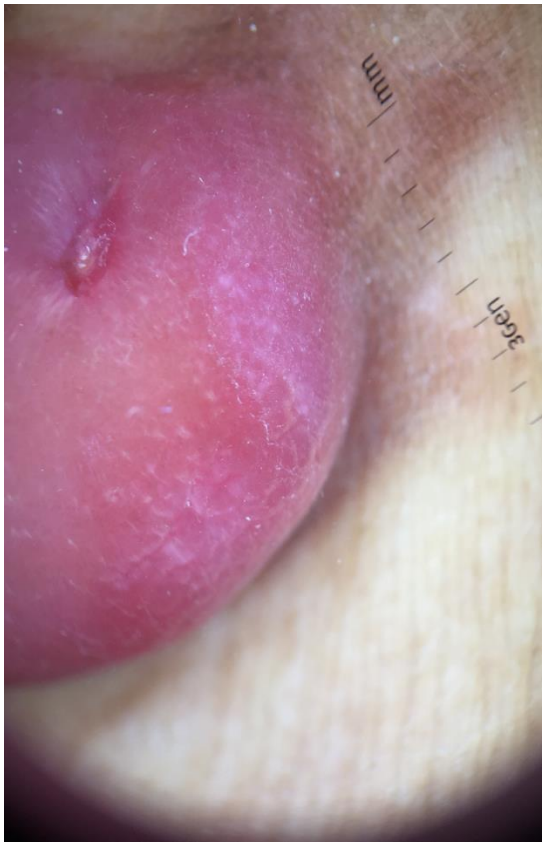

Figure 2: Dermoscopic image showing a fine central pigment network, multifocal milky-red areas, whitish linear structures and dilated mesh-like vessels.

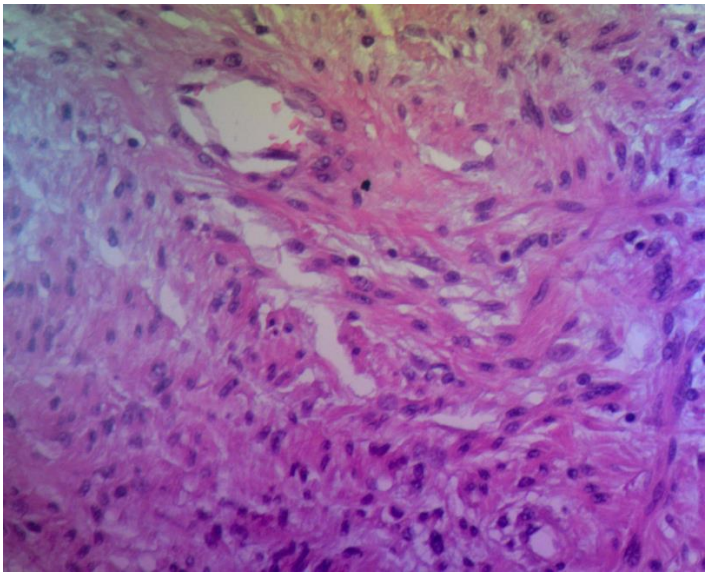

Figure 3: Histological image (H/E, *200) reveals a proliferation of spindle cells with diffuse growth forming a storiform pattern throughout the dermis. The basal cell layer is hyperpigmented and there is an increase in number of superficial dermis vessels.

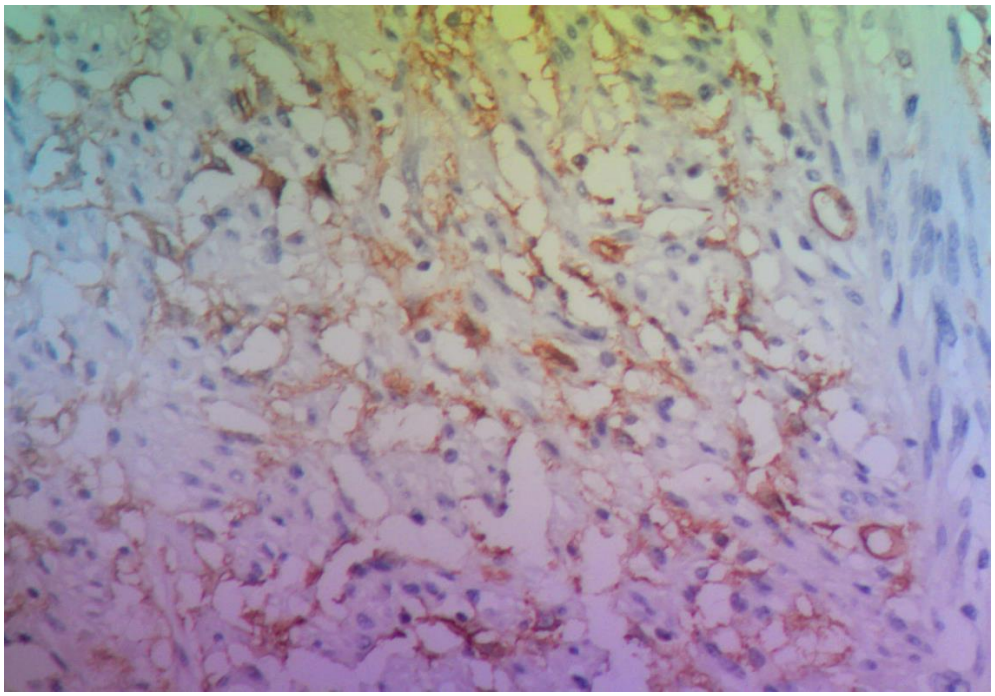

Figure 4: Immunostaining showing a net expression of CD34 by tumor cells.

Discussion

DFSP is a rather uncommon fibrohistiocytic tumour with an intermediate grade malignancy, first described by Darier and Ferrand in 1924 [1]. The incidence of DFSP has been estimated to be 0.8-5 cases/million per year [2]. DFSP usually occurs in adults aged 20-50 years. Most studies show an almost equal sex distribution or a slight male predominance [3]. This tumour mainly occurs on the trunk. DFSP is characterized by locally aggressive growth with a high recurrence rate. Surgical excision either Mohs micrographic surgery or a wide local excision (3 cm margin), remains the mainstay of treatment for DFSP. Due

to recent cytogenetic finding of the essential role of the collagen promoter in driving COL1A1 and PDGFB fusion protein production in the proliferation of DFSP tumour cells, molecular therapy represents an alternative in metastasized or locally advanced cases [4]. Our patient was treated with a conventional wide local excision due to the unavailability in our hospital at that time of either Mohs micrographic surgery or COL1A1-PDGFB translocation detection. Fortunately, no recurrence has been found during follow up after wide local excision in our case. The dermoscopic features described in the abovementioned case should be considered significant. We have found in the literature only two description of the dermoscopic features of DFSP in a report of a case one of pigmented DFSP (Bednar tumour), in which the authors describe a patchy pigment network and dot vessels. However, there is no dermoscopic image of the lesion [5].

Our patient presented with a fine pigment network, although the lesions are not histologically considered Bednar tumours. This pigmented variant of DFSP, described in 1957, accounts for 1per cent of all DFSP and is characterized by a usually scant population of dendritic melanocytes within an otherwise typical DFSP [2]. Like dermatofibromas, the pigment network corresponds to the increase of pigment in the epidermal basal cells rather than a melanocytic proliferation. Consequently, this represents another exception to the rule that the presence of a pigment network constitutes a dermoscopic clue for the diagnosis of melanocytic lesions [6,7]. The most prominent dermoscopic feature in our patient is the mesh-like vessels that are mainly peripherally located. Histologically, they correspond to the great number of dilated vessels that can be found in the superficial dermis of these tumours (Figure 3). An accurate interpretation of vascular structures may be especially useful for the diagnosis of non-pigmented skin tumours.

Several of these tumours present with a particular vascular pattern, such as the string of pearls pattern in clear cell acanthoma [8], the horseshoe-like pattern in Merkel cell carcinoma [9] or the multiple red lacunes with polymorphous vascular pattern in eccrine poroma [10]. To our knowledge, this mesh-like pattern has not been described in association with any specific tumour or inflammatory lesion thus far [11]. Finally, milky-red areas and whitish linear structures histologically correspond to intersecting bands of tumour cells forming a storiform pattern throughout the dermis.

In conclusion, we report here a case of DFSP to share dermoscopic characteristics, such as peripheral dilated vessels forming a mesh-like pattern, milky-red areas, whitish linear structures and fine pigment network. A dermoscopic examination of these lesions may enable an early diagnosis and better prognosis. Nevertheless, a large series describing the dermatoscopic features of DFSP is needed in order to improve our knowledge and early detection of this tumour.

References

1. Darier J, Ferrand M (1924) Dermatofibromes progressifs et recidi-vants ou fibrosarcomes de lapeau. Annales de Dermatologie et de Syphiligraphie 5: 542-562.
2. Sanmartín O, Llombart B, López-Guerrero JA, et al. (2007) Dermatofi-brosarcoma protuberans. Actas Dermo-Sifiliográficas 98: 77-87.
3. Criscione VD, Weinstock MA (2006) Descriptive epidemiology of der- matofibrosarcoma protuberans in the United States, 1973 to 2002. Journal of the American Academy of Dermatology 56: 968-973.
4. Simon MP, Pedeutour F, Sirvent N, et al. (1997) Deregulation of the platelet-derived growth factor β-chain gene via fusion with collagen gene COL1A1 in dermatofibrosarcoma protuberans and giant-cell fibroblastoma. Nature Genetics 15: 95-98.

5. Quigley EA, Marghoob AA, Busan KJ, et al. (2009) A firm red-brown plaque on the arm. Dermatofibrosarcoma protuberans (DFSP), pigmented variant (Bednar tumor). Archives of Dermatology 145(5): 589-594.
6. Zaballos P, Puig S, Llambrich A, et al. (2008) Dermoscopy of der- matofibromas: A prospective morphological study of 412 cases. Archives of Dermatology 144(1): 75-83.
7. Agero ALC, Taliercio S, Dusza SW, et al. (2006) Conventional and polarized dermoscopy features of dermatofibroma. Archives of Dermatology 142: 1431-1437.
8. Kreusch JF (2002) Vascular patterns in skin tumors. Clinics in Dermatology 20(3): 248-254.
9. Ciudad C, Avilés JA, Alfageme F, et al. (2010) Spontaneous regression in merkel cell carcinoma: report of two cases with a descrip- tion of dermoscopic features and review of the literature. Dermatologic Surgery 36(5): 687-693.
10. Avilés-Izquierdo JA, Velázquez-Tarjuelo D, Lecona-Echevarría M, et al. (2009) Dermoscopic features of eccrine poroma. Actas Der- mosifiliogr. 100(2): 133-136.
11. Vazquez-Lopez F, Kreusch JF, Marghoob AA (2004) Dermoscopic semiology: further insights into vascular features by screening a large spectrum of nontumoral skin lesions. The British Journal of Dermatology 150: 226-231.