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Case Presentation

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A Case of Atrophic Pigmented Dermatofibrosarcoma Protuberans and Short Review

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is an uncommon cutaneous sarcoma characterized histologically by spindle cells arranged in a storiform pattern, which has a high local recurrence rate, low metastatic rate and low mortality. Although, several uncommon subtypes of DFSP including pigmented or atrophic variants have been described, atrophic pigmented dermatofibrosarcoma protuberans is extremely rare, making it much more difficult to identify clinically. Here, we describe a case of atrophic pigmented dermatofibrosarcoma protuberans in a 28-year-old female and review other cases of atrophic pigmented DFSP that have been reported in the literature from 1997 in PubMed.

Keywords: Atrophic; Dermatofibrosarcoma protuberans; Pigment

Introduction

Dermatofibrosarcoma Protuberans (DFSP) is an uncommon cutaneous sarcoma characterized histologically by spindle cells arranged in a storiform pattern, which has a high local recurrence rate, low metastatic rate, and low mortality [1]. Although several uncommon subtypes of DFSP including pigmented or atrophic variants have been described [2], atrophic pigmented dermatofibrosarcoma protuberans is extremely rare, making it much

more difficult to identify clinically. Here, we describe a case of atrophic pigmented dermatofibrosarcoma protuberans.

Case Presentation

A 28-year-old female presented with an asymptomatic, slightly depressed, bluish lesion on her back near the waist that had been slowly progressing for 20 years and did not receive any treatment. There was no history of trauma to the area. She was otherwise healthy and denied systemic symptoms or relevant family history. On dermatological examination, there was a soft, 3 cm×1.4 cm, erythematous-to-bluish ill-defined, slightly depressed plaque on the left back near the waist, and subcutaneous capillaries visible **(Figure 1a).** Histopathological analysis on low-power view revealed fascicles of densely packed spindle cells extending around fat tissue with a reduced dermal thickness **(Figure 1b).** The monomorphic spindle cells had elongated darkly staining nuclei and bland cytoplasm **(Figure 1c).** Scattered pigmented cells were also noticed **(Figure 1d).** Immunohistochemical staining of spindle cells was positive for CD34 **(Figure 1e),** while negative for CK-P, SOX-10, CD31, CD3, CD20, Desimin and S-100 **(Figure 1f-i).** Based on the clinicopathological features, a diagnosis of atrophic pigmented DFSP was made, and wide local excision was performed.

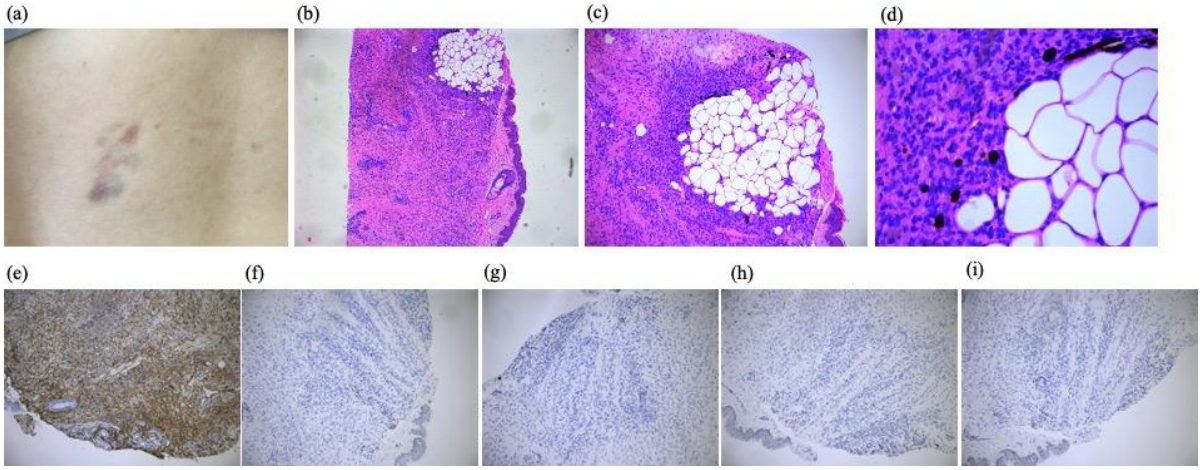

Figure 1: A soft, 3 cm×1.4 cm, erythematous-to-bluish ill-defined, slightly depressed plaque on the left back near waist (a); Fascicles of densely packed spindle cells extending around fat tissue with a reduced epidermal thickness (b); The monomorphic spindle cells had elongated darkly staining nuclei and bland cytoplasm (c) and scattered pigmented cells were also noticed (d); Immunohistochemical staining of spindle cells was positive for CD34 (e), negative for CD3 (f), CD20 (g), Desimin (h) and S-100 (i).

Discussion

The pigmented and atrophic variants of DFSP are rare, accounting for <5% **[3]** and 1.7% **[1]** of all cases, respectively. And atrophic pigmented dermatofibrosarcoma protuberans are extremely rare. The atrophic variant usually appears as a depressed plaque rather than protuberant nodules which are easily misdiagnosed. Pigmented dermatofibrosarcoma protuberans, also known as Bednar tumors, usually present as a bluish-black discoloration similar to a bruise, which develops into a slow-growing, painless plaque or tumor **[4].** In our case, the patient had the features of both pigmented and atrophic DFSP in the same lesion by immunopathogenesis, so the

diagnosis of atrophic pigmented dermatofibrosarcoma protuberans was established. So far, 8 cases of atrophic pigmented dermatofibrosarcoma protuberans in addition to ours have been reported in PubMed from 1997 till now **(Table 1) [2,3,5-8].**Various misdiagnoses were considered because of their atypical clinical presentations, such as lipoatrophy, hemangioma, hyperpigmentation, and neurofibroma **[6,9,10].** In our case, a misdiagnosis of lipoatrophy was made at the initial visit. DFSP typically occurs in young to middle-aged adults. The epidemiology of DFSP has no sex difference, and the vast majority occurred in the trunk or extremities **[1].** In these 8 cases, female: male ratio is 5:3. Trunk (5/8) and extremities (2/8) are the most frequently involved sites. DFSP has characteristic morphology, of storiform islands of bland spindle cells, and immunohistochemically, it shows diffuse positivity of CD34. The mainstay of treatment is surgical excision, either Wide Local Excision (WLE) or Mohs Micrographic Surgery (MMS). Other treatments include targeted immunotherapy and Radiotherapy. DSFP have a high local recurrence rate and need close clinical follow-up. In our case, local enlarged resection with a surgical margin of about 3 cm was used and is still in follow-up.

Table 1: Cases of atrophic DFSP reported in PubMed from 1997 to 2023.

Age, years	Sex	History, years	Location	Lesion morphology	Size (mm)	Misdiagnosed	Reference
24	F	2	Left infraorbital	Bluish atrophic lesion	NA	NA	Chuan, M.T. et al.
34	F	15	Left buttock.	Pigmented plaque	11× 12	NA	Taura M, et al.
7	M	NA	Left wrist	Red-brown, irregular congenital atrophic patch	20×40	Lipoatrophy	Zhang et al.
8	F	5	Left forearm	Bluish-black scleroatrophic plaque	10×10	Hemangioma	Zhang et al.
33	F	10	Left upper back	Asymptomatic, bluish and slightly depressed lesion	16×13	Post inflammatory hyperpigmentation	Lin et al.
44	M	3	Right back	NA	25×25	Neurofibroma	Xu et al.

26	M	10	Left back	Asymptomatic, smooth, bluish black plaque with central atrophy	30×25	NA	Bai et al.
28	F	20	Back near waist	Asymptomatic, slightly depressed, bluish lesion	30×14	Lipoatrophy	In this report

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Conflicts of Interest: we (all authors) declare no conflict of interest.

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