Atrophic pigmented dermatofibrosarcoma protuberans misdiagnosed as hyperpigmentation

Sir,

Dermatofibrosarcoma protuberans is a rare cutaneous fibroblastic sarcoma of intermediate malignancy characterized histologically by spindle cells arranged in a storiform pattern.[1] Although, several uncommon subtypes of dermatofibrosarcoma protuberans 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 misdiagnosed as hyperpigmentation.

A 33-year-old female had an asymptomatic, bluish and slightly depressed lesion on her left upper back for ten years which was previously diagnosed as postinflammatory hyperpigmentation and did not receive any treatment. Three years ago after her last child birth, she noticed that the lesion began to gradually enlarge and developed peripheral erythema. No history of trauma could be identified. She was otherwise healthy and denied systemic symptoms or relevant family history. On dermatological examination, there was a firm, 1.6 × 1.3 cm, erythematous-to-bluish ill-defined, slightly depressed plaque on the left upper back [Figure 1a]. Non-polarized dermoscopic evaluation with DERMOSCOPY-II (Dermat, Beijing) at 20x magnification revealed linear vessels within yellowish background with bluish structureless areas and shiny white streaks. No pigment network was observed [Figure 1b]. Histopathological analysis on low-power view revealed fascicles of densely packed spindle cells extending around fat tissue with a reduced dermal thickness [Figure 2a]. Cytologically, the monomorphic spindle cells had elongated darkly staining nuclei and bland cytoplasm with minimal mitotic figures [Figure 2b]. Scattered dendritic cells abundant in melanin were noticed [Figure 2b]. Immunohistochemical staining of spindle cells was positive for CD34 [Figure 2c] and vimentin, while negative for factor XIIIa and S-100. Melanin-laden dendritic cells were positive for S-100 [Figure 2d]. Modified Mohs micrographic surgery was performed to excise the lesion followed by secondary healing. No recurrence was observed at one-year follow-up.

Based on the clinicopathological features, more than ten variants of dermatofibrosarcoma protuberans including atrophic and pigmented variants have been described.[2] Atrophic dermatofibrosarcoma protuberans was first described by Lambert in 1985 characterized by a slow-growing depressed plaque which revealed decreased dermal thickness by >50% compared with the surrounding dermis histologically.[1,3] According to a single-center experience, atrophic variant accounts for only 1.7% (16/937) of all dermatofibrosarcoma protuberans cases.[4] Pigmented dermatofibrosarcoma protuberans, also known as Bednar tumor, usually presents as a bluish plaque and is characterized by melanin-laden cells histologically.[1] It predominantly occurs in Black population and accounts for approximately 1–5% of all dermatofibrosarcoma protuberans cases.[1] With both characteristic histological features seen in our case, the diagnosis of atrophic pigmented dermatofibrosarcoma protuberans was established. A typical dermatofibrosarcoma protuberans lesion appears as an indurated protuberant plaque or nodule which may mimic dermatofibroma, neurofibroma, leiomyoma, keloid and desmoid tumors and other soft-tissue sarcomas such as Kaposi sarcoma and fibrosarcoma.[5] The differential diagnosis of atrophic dermatofibrosarcoma protuberans, however, consists of another group of disorders including morphea, anetoderma, morphea-like or sclerosing basal cell carcinoma, atrophic dermatofibroma, atrophic scar, lipoatrophy, lymphocytoma etc.[6] When it presents with bluish pigmentation, differential diagnoses of bruise, blue nevi, melanocytic nevi and malignant melanoma should also be taken into consideration.

So far, seven cases of atrophic pigmented dermatofibrosarcoma protuberans in addition to ours have been reported in PubMed, all presenting with a depressed pigmented plaque, three of which developed a nodule within the plaque [Table 1].[4,7-10] Due to their atypical presentations, various incorrect diagnoses were considered, including lipoatrophy,

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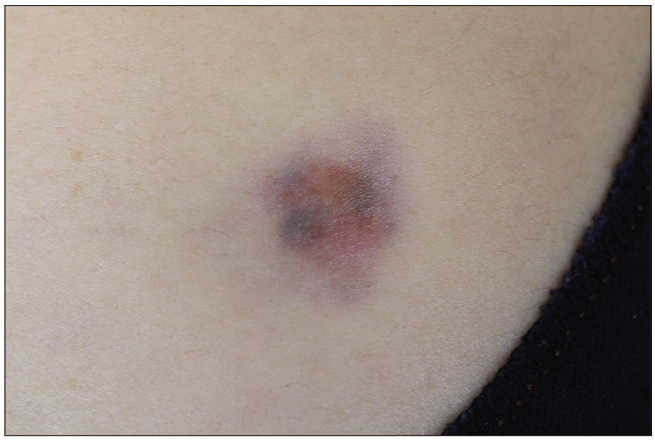

Figure 1a: A firm, 1.6 × 1.3 cm, erythematous-to-bluish ill-defined, slightly depressed plaque seen on the left upper back

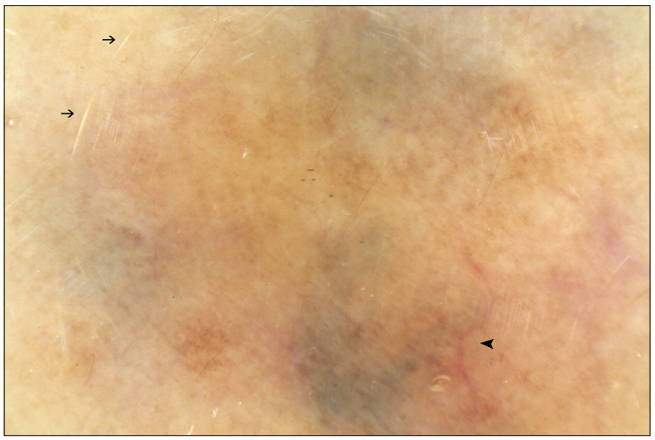

Figure 1b: Non-polarized dermoscopy with DERMOSCOPY-II (Dermat) at 20x magnification revealed linear vessels (arrowhead) within yellowish background with blue structureless areas and shiny white streaks (arrows)

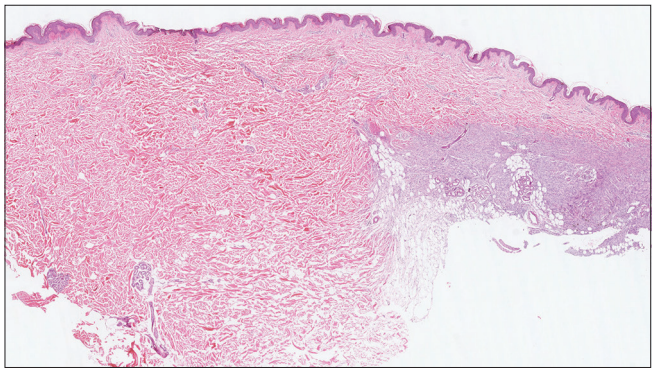

Figure 2a: Histopathologically, fascicles of densely packed spindle cells extending around fat tissue with reduced dermal thickness by about half on low-power view (Hematoxylin-eosin; ×15)

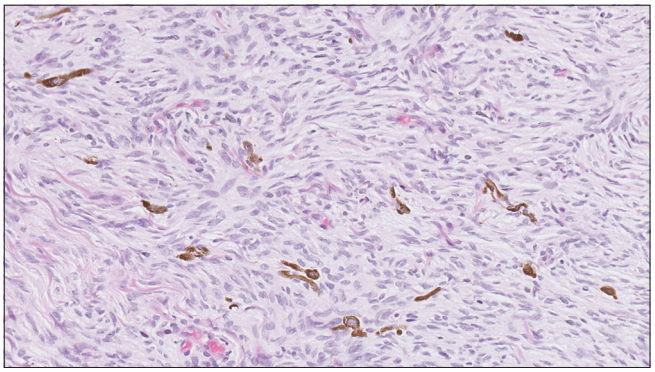

Figure 2b: Monomorphic spindle cells display elongated darkly staining nuclei and bland cytoplasm with minimal mitotic figures. Scattered dendritic cells abundant in melanin also seen (Hematoxylin-eosin; ×200)

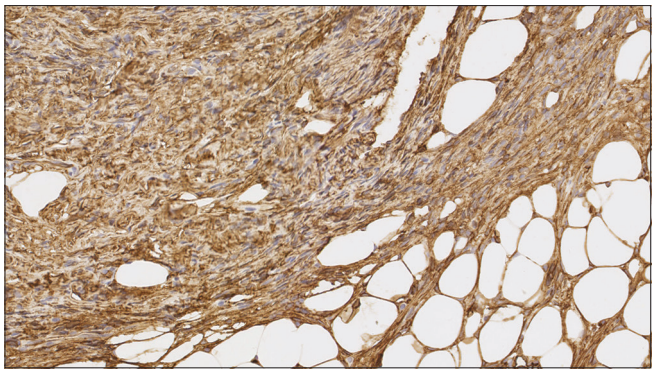

Figure 2c: Immunohistochemistry showed CD34 + cells infiltrating into subcutis and forming honeycomb-like pattern (anti-CD34; ×200)

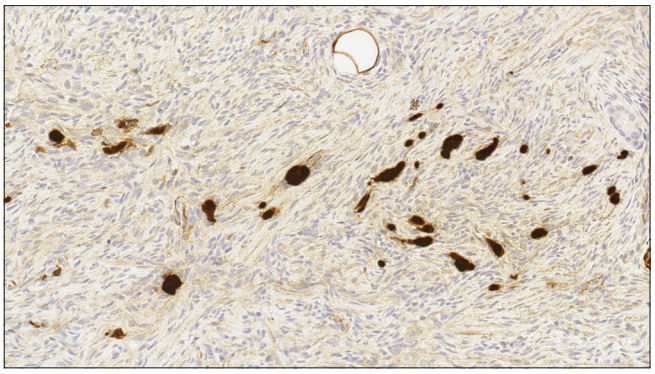

Figure 2d: Immunohistochemistry showed spindle cells were negative for S-100, in contrast, melanin-bearing dendritic cells were positive for S-100 (anti-S-100; ×200)

hemangioma and neurofibroma.[4,9] In our case, misdiagnosis of hyperpigmentation was made at initial visiting and only observation was recommended. It was gradual enlargement of the lesion that made us reconsider the diagnosis. Therefore, any solitary hyperpigmented lesion that does not resolve spontaneously or evolves on follow up should be biopsied to exclude malignancy even if it is not suspected clinically.

Dermoscopy is a valuable non-invasive tool to differentiate a variety of cutaneous pigmented or non-pigmented tumors. However, it is not widely employed in the diagnosis of dermatofibrosarcoma protuberans. Bernard *et al*.[11] summarized six dermoscopic features based on 15 classical cases of dermatofibrosarcoma protuberans: delicate pigment network, vessels, structureless light

Table 1: Cases of atrophic pigmented dermatofibrosarcoma protuberans reported in PubMed

Age	Sex	Location	Size (mm)	Clinical presentations	Clinical differential diagnosis	Reference
24	F	Infraorbital area	NA	Well-demarcated bluish depressed lesion	NA	Chuan *et al*., 1997[7]
16	F	Thigh	12×15	Multiple papules, plaques and nodules superimposed over several atrophic or indurated plaques	NA	Sathyanarayana *et al*., 2004[10]
34	F	Buttock	11×12	Pigmented plaque	NA	Taura *et al*., 2016[8]
7	M	Wrist	20×40	Hard hemispherical nodule in a red-brown irregular atrophic patch	Lipoatrophy	Zhang *et al*., 2018[9]
8	F	Forearm	10×10	Nodule over a bluish-black atrophic plaque	Hemangioma	Zhang *et al*., 2018[9]
7	M	Forearm	5×5	NA	NA	Xu *et al*., 2019[4]
44	M	Back	25×25	NA	Neurofibroma	Xu *et al*., 2019[4]

F: Female, M: Male, NA: Not available

brown areas, shiny white streaks, pink background color and structureless hypo- to depigmented areas which are presented in a multicomponent pattern, suggesting the diagnosis of malignancy. Another recent study summarized dermoscopic features of 32 dermatofibrosarcoma protuberans cases as follows: features of vessels, pigment network and pinkish background presented in 26 (81.3%), 25 (78.1%) and 21 (65.6%) patients, respectively.[12] On a literature search, we could not come across any specific dermoscopic features of atrophic pigmented dermatofibrosarcoma protuberans. Vessels and white streaks seen in our case were similar to the description of classic types. Structureless yellowish background may be a distinctive finding in atrophic type which might be the result of dermal atrophy and approximation of the subcutis to the epidermis. Bluish pigmentation likely corresponded to the distribution of melanin-laden cells in the dermis in pigmented dermatofibrosarcoma protuberans.

Modified Mohs micrographic surgery was performed to achieve complete resection with minimal sacrifice of normal tissue. No differences in terms of treatment options and prognosis between classical and atrophic or pigmented dermatofibrosarcoma protuberans have been delineated. However, atypical presentations often lead to misdiagnosis, delayed surgery as well as inadequate resection margins. Knowledge of the clinical characteristics dermoscopic features of this variant may aid in early diagnosis of atypical dermatofibrosarcoma protuberans.

Declaration of patient consent

The patient's consent is not required as the patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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