Dermatofibrosarcoma protuberans: Two rare variants

Sir,

Dermatofibrosarcoma protuberans is a dermal neoplasm with low malignant potential. Histopathologically, it is a poorly circumscribed neoplasm characterized by storiform arrangement of spindle-shaped cells in short interlacing fascicles.[1] We hereby describe two rare variants of dermatofibrosarcoma protuberans.

Case 1

A 42-year-old woman presented with an asymptomatic, slowly progressive lesion on her right shoulder since last 25 years. Dermatological examination revealed a single ill-defined, atrophic, indurated, nontender, erythematous papulonodular plaque sized 6 cm × 4 cm with few intervening areas of bluish discoloration [Figure 1]. Punch biopsy from an erythematous nodule demonstrated an unencapsulated, poorly demarcated neoplasm extending from the papillary dermis to the base of specimen, thus obscuring the entire dermis and subcutis. [Figure 2a]. The overlying epidermis showed flattening and loss of rete ridges. The neoplasm was densely packed with spindle-shaped cells arranged in multiple, irregular, interwoven fascicles, resulting in a storiform pattern. These spindle-shaped cells were monomorphic their hyperchromatic slender nuclei showing negligible pleomorphism [Figure 2b]. Near the lower end, some interspersed melanin-containing dendritic cells were noted after appropriate staining (Fontana-Masson stain). [Figure 2c]. Immunohistochemical analysis confirmed the presence of two distinct cell populations- the spindle-shaped cells showing diffuse positive staining with CD34 and negative staining with S100 [Figure 2d] while the melanin-bearing cells stained positively with S100 and negative for CD34 [Figure 2e].

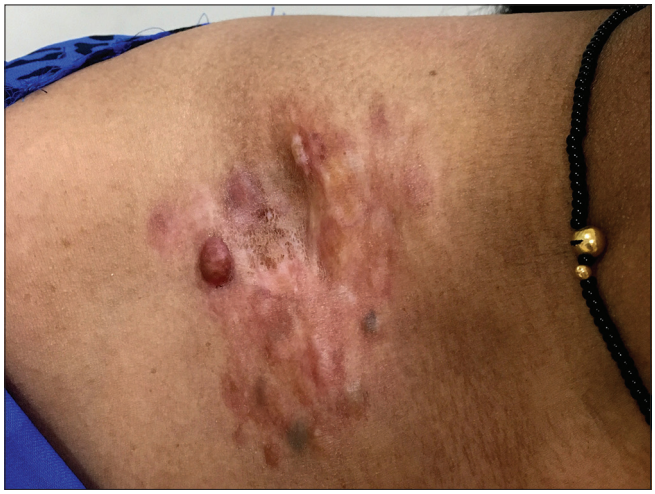

Figure 1: An ill-defined, atrophic, indurated, erythematous papulonodular plaque with some areas of bluish discoloration present on the right shoulder

Based on the above findings, a diagnosis of Bednar's tumor (pigmented dermatofibrosarcoma protuberans) was made.

Case 2

A 19-year-old woman presented with an asymptomatic lump on her right thigh for the last 5 years gradually increasing in size. On local inspection, the overlying skin was normal without any visible lump. However, palpation revealed a deep, firm, ill-defined, nontender, movable subcutaneous mass with proportions 3 cm × 5 cm [Figure 3]. Morphea and subcutaneous zygomycosis were considered as differentials. Histopathological examination revealed a well-circumscribed neoplasm, though microscopically infiltrative, limited almost exclusively to the subcutis, with a well-defined horizontal, upper margin [Figure 4a]. The neoplasm was composed of monomorphic, spindle-shaped cells with elongated non-pleomorphic nuclei, arranged in short interlacing fascicles [Figure 4b]. The intervening collagen was coarse and abundant. The neoplasm infiltrated the subcutis in a honey-comb pattern reaching the base of the specimen. The neoplastic cells also surrounded a hair bulb with focal dermal involvement [Figure 4a]. On immunohistochemical analysis,

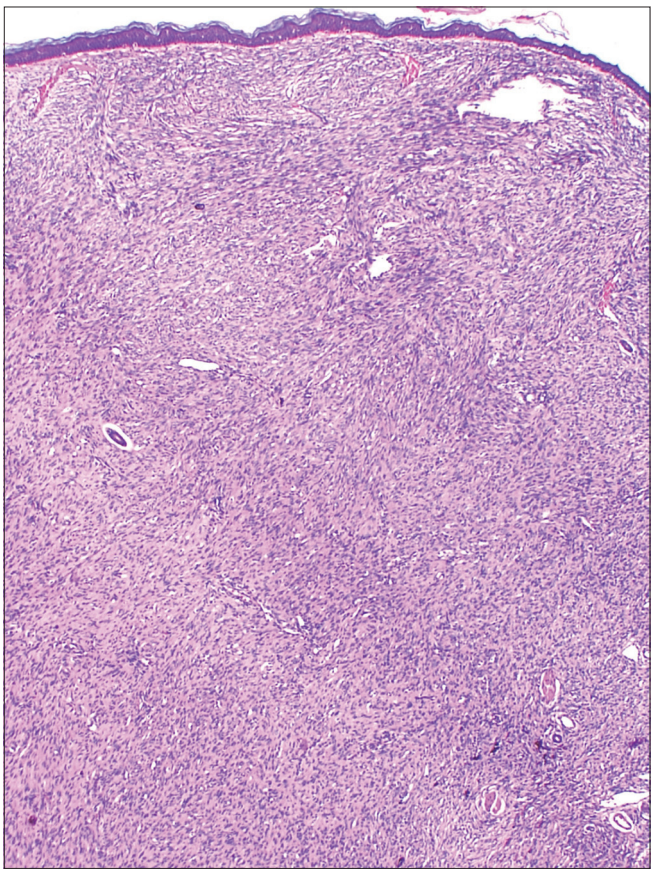

Figure 2a: An unencapsulated, poorly demarcated neoplasm, composed of spindle shaped cells arranged in storiform pattern, with a few melanin containing dendritic cells towards the base of the neoplasm (hematoxylin and eosin, ×25)

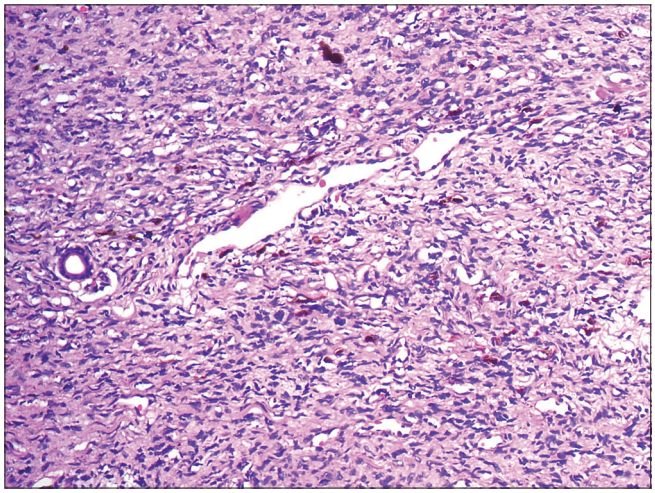

Figure 2b: Monomorphic spindle shaped cells with hyperchromatic nuclei, along with a few scattered melanin containing dendritic cells (hematoxylin and eosin, ×100)

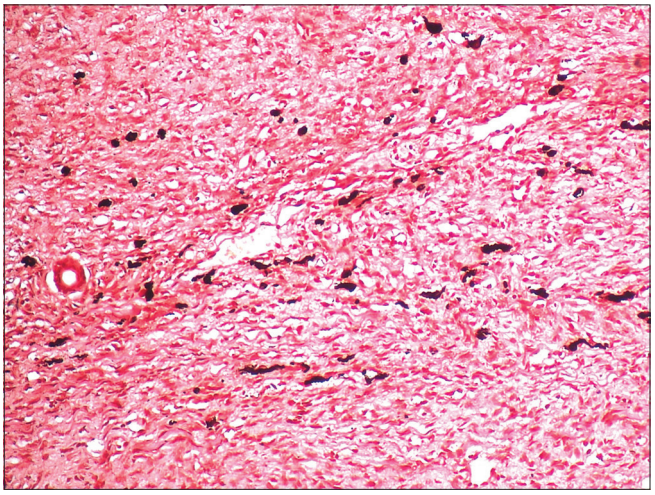

Figure 2c: Fontana-Masson stain highlighted the melanin-bearing dendritic cells (Fontana-Masson stain, ×100)

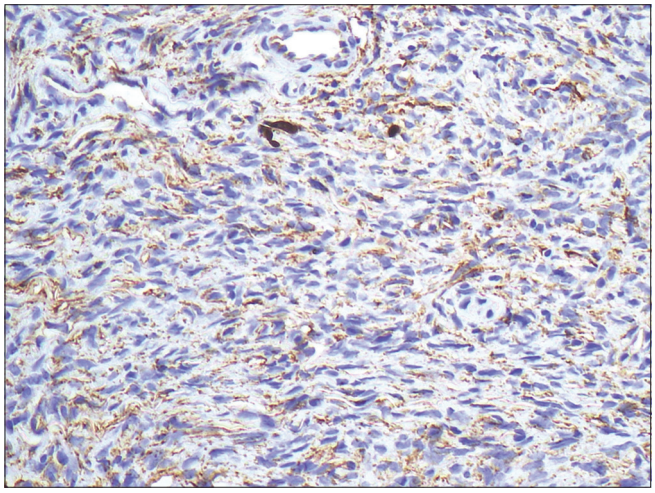

Figure 2d: Spindle-shaped cells showed diffuse positive staining with CD34 antibody (CD34 marker, ×200)

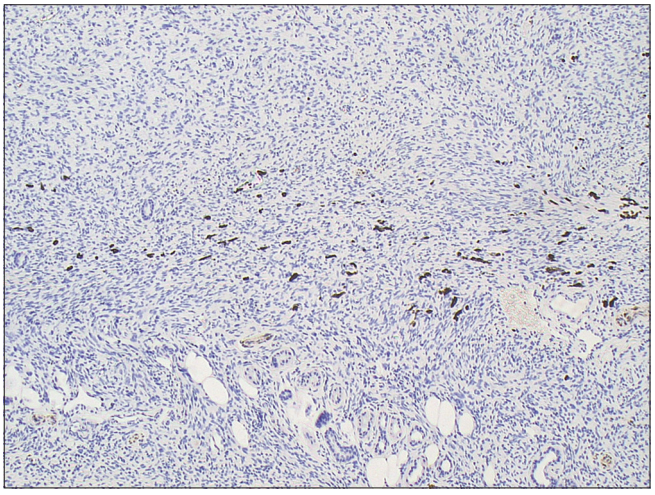

Figure 2e: Melanin-bearing dendritic cells showed positive staining with S100 antibody (S100 marker, ×50)

the neoplastic cells stained diffusely positive with CD34 and negative with S100 [Figure 4c].

On the basis of above analysis, a diagnosis of subcutaneous dermatofibrosarcoma protuberans was made. Bednar's tumor (pigmented dermatofibrosarcoma protuberans) represents 1–5% of all dermatofibrosarcoma protuberans cases.[2] It differs from classic dermatofibrosarcoma protuberans only by the presence of scattered melanin-bearing dendritic cells. Clinically, t can be nonpigmented or pigmented (blue/black), depending on the amount of melanin pigment.[1] The histogenesis of melanin-containing dendritic cells remain controversial. Neuroectodermal differentiation and melanocytic colonization from epidermis are the two proposed theories.[3], Neuromesenchymal origin has also been suggested, as corroborated by the association of Bednar's tumor with dermal melanocytosis (nevus of Ito).[4]

Histopathologically, it is characterized by spindle-shaped cells, arranged in a storiform pattern, admixed with a variable population of melanin-containing dendritic cells. On immunohistochemistry, spindle-shaped cells and the melanin-containing dendritic cells stain positively with CD34 and S100 respectively. Histopathological differential diagnoses include other pigmented spindle cell neoplasms [Table 1].

Conventionally, dermatofibrosarcoma protuberans is a dermal neoplasm with frequent extension into subcutaneous fat; however, recently, a new variant, subcutaneous dermatofibrosarcoma protuberans has been recognized, where the neoplasm is confined exclusively or almost exclusively to subcutaneous fat with minimal and clinically inapparent dermal involvement.

Clinically, subcutaneous dermatofibrosarcoma protuberans manifests as an asymptomatic, nonprotuberant, nonatrophic,

Table 1: Histopathological differential diagnoses of Bednar's tumor (pigmented dermatofibrosarcoma protuberans)

Cutaneous pigmented spindle cell neoplasm	Immunohistochemical staining and/or special staining for differentiation
1. Pigmented neurofibroma It is characterized by the presence of heterogeneous population of plump fusiform, elongated spindle, and oval Schwann cells, lack of typical storiform pattern, presence of epithelioid cells and Wagner-Meissner-like bodies, though seen occasionally, and frequent myxoid change	Diffusely immunoreactive for S100 with scattered CD34 positive cells
2. Pigmented atypical fibroxanthoma It is characterized by the presence of xanthomatous cells with vesicular nuclei, variably extensive areas of intratumoral hemorrhage and hemosiderin containing neoplastic cells	Factor XIIIa positivity, CD 10 positivity, S100 negative Iron stains (colloidal iron, Prussian blue stain) positivity

Table 2: Histopathological differential diagnoses of subcutaneous dermatofibrosarcoma protuberans

Subcutaneous spindle cell neoplasms	Immunohistochemical staining with CD34 and S100 antibodies
Atypical lipomatous neoplasm (well-differentiated liposarcoma) It is made up of a distinctive admixture of mature fat and irregular, fibrous septa containing pleomorphic, hyperchromatic cells. Similar hyperchromatic cells are frequently present within the vessel walls	CD34 positive
Spindle cell lipoma It consists of spindle cells that are admixed with mature adipocytes and characteristic bundles of eosinophilic collagen. Spindle cells are somewhat smaller with more ovoid nuclei when compared with those of DFSP	CD34 diffusely positive
Solitary fibrous tumor It is characterized by a deeply located, well-circumscribed mass which has a prominent branching; "staghorn" vascular pattern; a "patternless" growth pattern without well-formed storiform pattern	CD34 diffusely positive
Deep dermatofibroma It may show prominent storiform pattern and limited involvement of adjacent adipose tissue, but lacks diffuse fat infiltration It consists of spindle cells that are plumper and more eosinophilic, along with a variable population of multinucleated giant cells, siderophages and foamy macrophages	Lacks diffuse CD34 positivity, though focal CD34 positivity frequently present
Diffuse neurofibroma It is ill-defined and diffusely infiltrates in the dermis and subcutaneous fat, extending along connective tissue septa It envelopes the normal adnexal structures, without destroying them Presence of abundant myxoid matrix (may simulate myxoid DFSP) and numerous mast cells Wagner-Meissner bodies are characteristic, often found on careful inspection	Lacks diffuse CD34 positivity, though CD34 may be expressed by a population of cells S100 diffusely positive

DFSP: Dermatofibrosarcoma protuberans

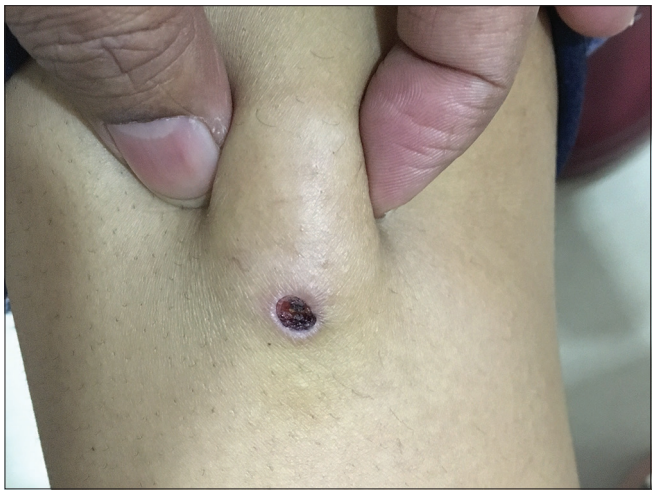

Figure 3: Palpation revealed a deep, firm, ill-defined subcutaneous mass on the right thigh

well- or ill-defined, indurated, subcutaneous mass without any visible skin change.[5] Histopathologically, it is characterized by a dense storiform arrangement of monomorphic spindle-shaped cells with slender nuclei, confined almost exclusively to the subcutis. Another unusual feature of this variant is its frequently well-circumscribed nature (although microscopically infiltrative). Bague and Folpe documented 15 cases of subcutaneous dermatofibrosarcoma protuberans, and found all the cases to be well-circumscribed.[6] Recently, Llombart *et al*. studied 18 cases of subcutaneous dermatofibrosarcoma protuberans and observed 10 cases to be well circumscribed and eight cases as poorly delimited and infiltrative.[5]

Immunohistochemistry with CD34 antibody reveals the neoplastic cells to be diffusely positive.[5,6] Hence, except for the deep location and well-circumscribed nature, the histopathological and immunohistochemical features of

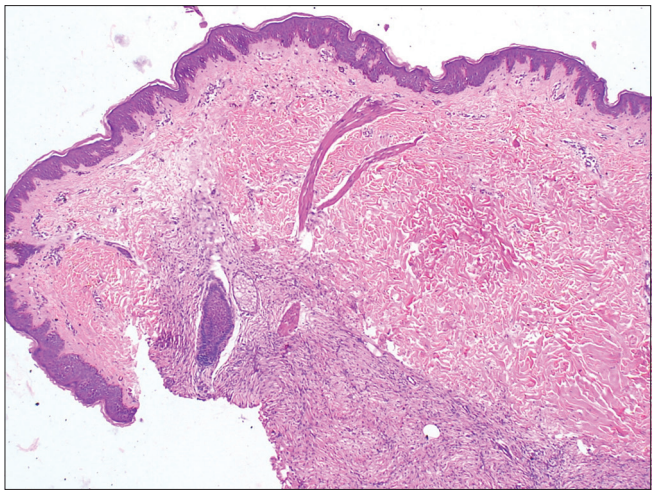

Figure 4a: A well circumscribed neoplasm, confined nearly exclusively to the subcutis, with a well-defined, horizontal, upper margin, with neoplastic cells surrounding a hair bulb, with focal dermal involvement (hematoxylin and eosin, ×25)

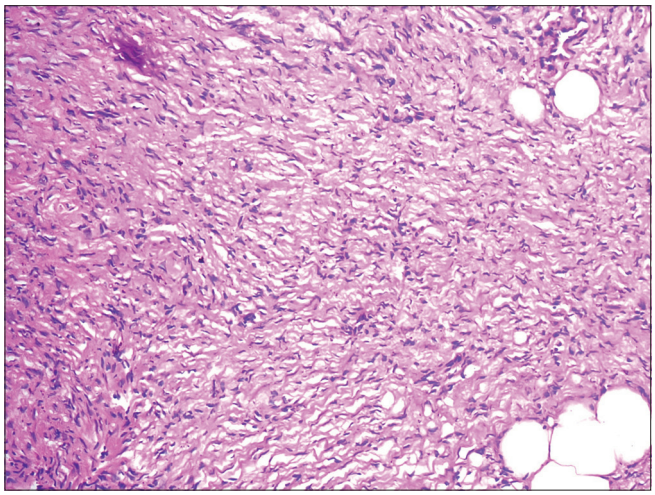

Figure 4b: Monomorphic, spindle-shaped cells with elongated nuclei arranged in short interlacing fascicles with coarse and abundant intervening collagen (hematoxylin and eosin, ×100)

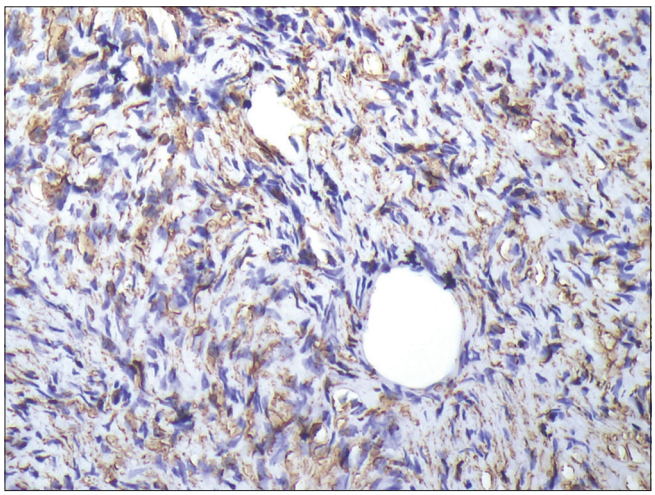

Figure 4c: Spindle-shaped cells showed diffuse positive staining with CD34 antibody (CD34 marker, ×200)

subcutaneous dermatofibrosarcoma protuberans are identical to cutaneous (conventional) dermatofibrosarcoma protuberans.

Histopathological differential diagnoses of subcutaneous dermatofibrosarcoma protuberans include other subcutaneous spindle cell neoplasms [Table 2].[6] These cases highlight the importance of careful recognition of stereotypical histopathological features of dermatofibrosarcoma protuberans and prudent use of ancillary immunohistochemistry (CD34 antibody) to accurately diagnose these rare, unusual variants of a low malignant neoplasm. In addition, awareness of atypical clinical presentation and deep biopsy are needed to diagnose it's rarer variants.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Sarina Jain, Shuken Dashore[1], Bharat Singhania[2], Uday S. Khopkar, Bhushan Darkase

Department of Dermatology, Seth GS Medical College and KEM Hospital, Mumbai, Maharashtra, [1]Dr Dashore's DHL Clinic, Indore, Madhya Pradesh, [2]Singhania Skin Clinic, Raipur, Chhattisgarh, India

Correspondence: Dr. Sarina Jain,
Department of Dermatology, Main Hospital Building, Ward 17-18,
Ground Floor, KEM Hospital, Parel, Mumbai - 400 012,
Maharashtra, India.
E-mail: sarinajaingoyal@gmail.com

References

1. Llombart B, Serra-Guillén C, Monteagudo C, López Guerrero JA, Sanmartín O. Dermatofibrosarcoma protuberans: A comprehensive review and update on diagnosis and management. Semin Diagn Pathol 2013;30:13-28.
2. Dupree WB, Langloss JM, Weiss SW. Pigmented dermatofibrosarcoma protuberans (Bednar tumor): A pathologic, ultrastructural, and immunohistochemical study. Am J Surg Pathol 1985;9:630-9.
3. Ding JA, Hashimoto H, Sugimoto T, Tsuneyoshi M, Enjoji M. Bednar tumor (pigmented dermatofibrosarcoma protuberans). An analysis of six cases. Acta Pathol Jpn 1990;40:744-54.
4. Amonkar GP, Rupani A, Shah A, Deshpande R. Bednar tumor: An uncommon entity. Dermatopathology (Basel) 2016;3:36-8.
5. Llombart B, Serra-Guillén C, Rubio L, Nagore E, Requena C, Traves V, *et al.* Subcutaneous dermatofibrosarcoma protuberans, a rare subtype with predilection for the head: A retrospective series of 18 cases. J Am Acad Dermatol 2017;77:503-110.
6. Bague S, Folpe AL. Dermatofibrosarcoma protuberans presenting

as a subcutaneous mass: A clinicopathological study of 15 cases with exclusive or near-exclusive subcutaneous involvement. Am J Dermatopathol 2008;30:327-32.

Access this article online

Quick Response Code:

Website: www.ijdvl.com

DOI: 10.4103/ijdvl.IJDVL_923_17

How to cite this article: Jain S, Dashore S, Singhania B, Khopkar US, Darkase B. Dermatofibrosarcoma protuberans: Two rare variants. Indian J Dermatol Venereol Leprol 2019;85:204-8.

Received: February, 2018. **Accepted:** October, 2018.