Case Report **Doi:** 10.18231/j.occ.2024.003

A rare case of Bednar tumor

Abhijit Bhanji,[1] Amrita Neelakantan[1] ✉
[1]Laboratory Director, Dept. of Histopathology, Lifenity Health Ltd, Mumbai, Maharashtra India

Email-id: amritaneel@yahoo.co.in

How to cite the article: Bhanji A, Neelakantan A. A rare case of Bednar tumor. *Onco Critical Care.2024*;2(1).07-09

Keywords: Fibrohistiocytic, Thecharacteristic, Protruberans

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue sarcoma that arises in the dermis.[1] It is more commonly found on the trunk and proximal extremities. [1] It is a slow-growing fibro histiocytic tumor of a low to intermediate grade malignancy, with a high propensity for a local recurrence and a low rate of distant metastasis.[2]

A rare variant of dermatofibrosarcoma protuberans is Bednar tumor which is a pigmented variety described first in 1957, by Bednar under the name of storiform neurofibroma.[3,4] It accounts for less than 5% of dermatofibrosarcoma protuberans variants.[4] It has been reported in approximately 1 to 5% of DFSP and has a predilection for the back and shoulders of young to middle-aged adults, with sporadic cases reported in the pediatric age group.[2]

The histology is unique because of the presence of pigment-laden dendritic cells scattered among spindle shaped cells with a storiform pattern.[5] The characteristic honeycomb appearance results from irregular tentacle-like projections infiltrating the underlying subcutaneous tissue, traversing the septa and fat, leading to fat entrapment.[1]

Extensive local infiltration and extending well beyond the visible lesion, characterizes this lesion. The distant metastasis is rare unless fibro-sarcomatous transformation occurs.[1] We present a case of a 38-year-old male patient who presented with a pigmented forearm swelling.

Case Report

A 38-year-old man presented with a history of a slow growing swelling on his right forearm for the last 1 year. The lesion appeared well defined, firm with regular margins. He had no significant personal and family history, and no recent medical history of the trauma of the affected area. On clinical examination, a dark-brown to blackish lesion of firm consistency, measuring 6 x 5 cm, was observed on the right arm. There was no pain, itching or signs of local inflammation. All routine blood investigations including complete hemogram, bleeding time, clotting time, random blood sugar, serology were within normal limits. Clinical diagnosis of a melanoma or soft tissue sarcoma was made. The lesion was excised with wide margins and sent for histopathology.

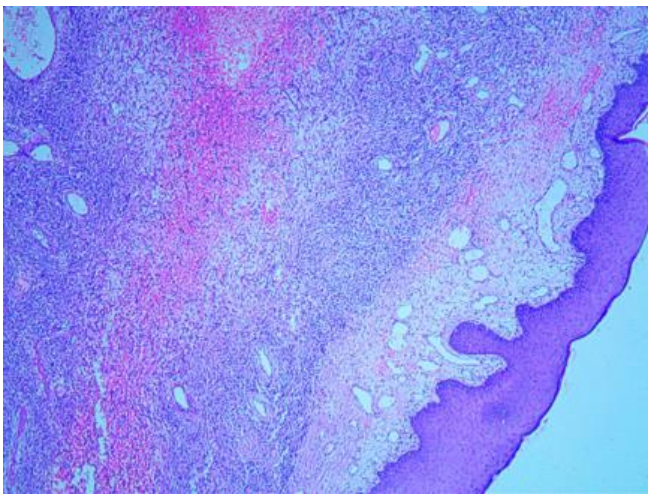

Fig 1: H&E 40x; Dermal tumour with Grenz zone

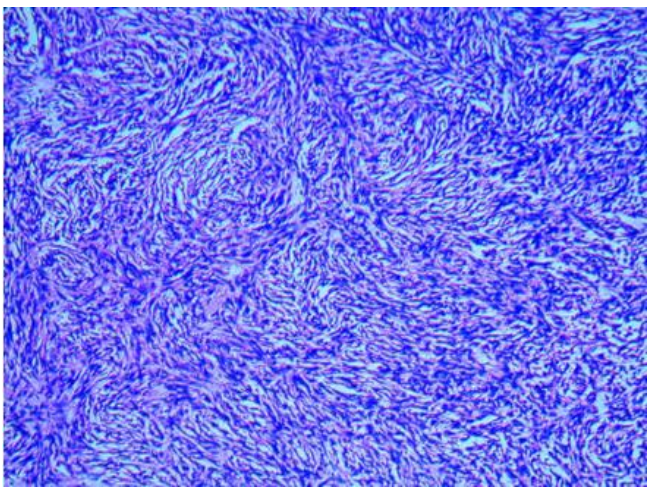

Fig 2: H&E 100x; Storiform architecture

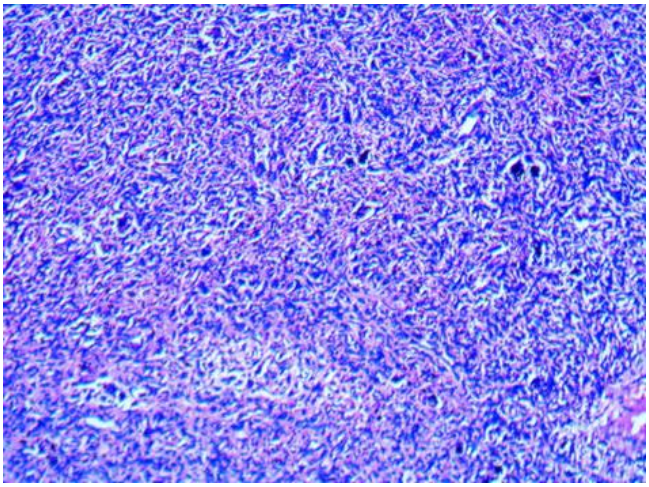

Fig 3: H&E 100x; Pigmented spindle shaped melanocytic cells

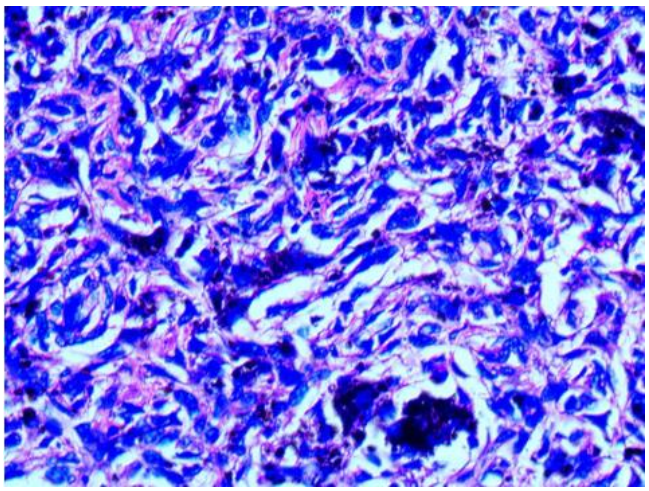

Fig 4: H&E 400x; Pigmented spindle shaped melanocytic cells

Gross examination revealed a skin covered tissue piece measuring 8 x 7 x 6 cm. On cut section, a diffuse, grey white tumour was noted, with clear margins on gross. The histopathological examination revealed a dermal tumour with sparing of superficial dermis focally (Grenz zone) (Fig 1). There was storiform arrangement of the spindle-shaped cells (Fig 2) admixed with the melanin- containing cells (Fig 3,4). Necrosis was not present and mitotic activity was low. The margins were free of the tumour. Provisional diagnosis of low grade spindle cell tumour was made, and further characterization by immunohistochemistry (IHC) was done. A panel of IHC was performed and the tumor cells were positive for clusters of differentiation 34 (CD34) and negative for alpha smooth muscle actin (SMA), S-100, Human Melanoma Black (HMB)-45, Caldesmon and Melan A. Based on the histomorphology and immunohistochemistry, a diagnosis of pigmented dermatofibrosarcoma was rendered.

Discussion

Dermatofibrosarcoma Protruberans is a rare cutaneous neoplasm with the reported prevalence of 0.1-1% among all cutaneous malignant tumors.[2] Besides conventional Dermatofibrosarcoma Protruberans, more than 10 clinicopathological subtypes have been reported which include myxoid, juvenile, granular cell, palisading, and pigmented subtype. The pigmented subtype is known as Bednar tumor. It shares it's histopathological, immunohistochemical and cytogenetical features with the conventional Dermatofibrosarcoma Protruberans, the difference being the presence of the melanin-containing dendritic cells.[2]

Though the histogenesis of pigmented Dermatofibrosarcoma Protruberans is widely debated, neuro-ectodermal differentiation or melanocytic colonization are the two proposed theories for histogenesis for the Bednar tumour.[4]

The melanin is responsible for the pigmented nature of this tumour.[3] The melanocytes may also be derived from the hair follicle melanocytes.[2] Few studies believe that these tumors may arise due to local traumas, such as burns, vaccination scars etc.[6]

It has been proposed that dermatofibrosarcoma protuberans may be of perineural fibroblastic origin based on the ultrastructural demonstration of a basal lamina around tumour cells, intercellular junctions and rather limited rough endoplasmic reticulum.[9] Furthermore, the presence of dendritic melanocytes in pigmented variants has often been used as additional presumptive evidence of neuroectodermal origin.[9] Immunohistochemical studies reveal a positive reaction to CD 34 and vimentin in most of the tumour cells, and are negative for HMB-45 and protein S-100. However, the melanin-containing cells may react positively to protein S-100.[8]

The absence of S-100 protein from the pigmented melanin-containing spindle cells has been attributed by some authors to the perineural rather than true Schwann cell differentiation.[9]

Differential diagnoses for this entity include pigmented neurofibroma, dermatofibroma, psammomatous melanotic schwannoma, and malignant melanoma.[10]

Pigmented neurofibroma does not appear dark colored or black due to only the microscopic presence of melanin. It also does not show a storiform pattern and is negative for CD34. Dermatofibroma is distinguished from pigmented variant of dermatofibrosarcoma protruberans by the presence of foamy macrophages, absence of pigment, less prominent storiform pattern, less uniform immunostaining with CD34, and CD68-positive histiocytes. Psammamatous melanotic schwannoma shows psammoma bodies, and tumor cells are S-100 positive and CD34 negative. Histopathology of malignant melanoma shows large degree of atypia and mitoses, and can be ruled out even though they are clinical mimics of each other.[10]

Desmoplastic (neurotrophic) variant of malignant melanoma shows neurotropism, focal melanocytic junctional activity, and diffuse and strong S-100 protein, HMB 45 immunoreactivity, CD34 negativity, whereas the pigmented dermatofibrosarcoma protruberans does not show any junctional activity, and shows strong CD 34 immunoreactivity.[8]

The differentiation of Bednar tumour from other pigmented cutaneous and soft tissue neoplasms is important because it has a less aggressive behavior, although with some propensity for local recurrence and an occasional rare incidence of metastasis.[8]

Studies reported a recurrence rate of 17% among cases of Bednar tumors. In addition, they reported an average interval of 9 years for recurrence (range, 9 months to 23 years).[8] The recommended treatment for Bednar tumors is wide excision with more than 2-3 cm margins of visibly uninvolved tissue and inclusion of the superficial fascia in the specimen.[8] Our case was a pigmented growth, which was excised with wide margins, and showed the histologic and IHC prolife matching a Bednar tumour.

The present case report explores an uncommon differential diagnosis to melanoma or other pigmented malignant tumors, which more aggressive than Bednar tumour. The accurate diagnosis of this entity is important to avoid radical treatment protocols and provide a better prognosis.

Source of Funding
None.

Conflict of Interest
None.

References

1. Menon G, Brooks J, Ramsey ML. Dermatofibrosarcoma Protuberans.
2. Cekić S, Jović A, Popović D, Zlatanović Z, Vidović N, Kocić H, Tiodorović D. Bednar Tumor–a Case Report. Serbian Journal of Dermatology and Venereology. 2020;12:59-62.
3. Jaishankar J, Krishnan RA, Kumar KS, Raghavan V. A Case of Bednar Tumour in the Sacral Region-A Rare Pigmented Variant of Dermatofibrosarcoma Protuberans. J Clin Diagn Res. 2022;16:PD05-06.
4. Kalonia T, Dhanta A, Singh A, Kumar A, Malik A, Kumari N. A report of two cases of Bednar tumour, an uncommon tumour of the skin: a report of two cases. Pan Afr Med J. 2022;42.261
5. Patnayak R, Prayaga A, Anuradha SV, Ahmed F, Jena A, Gupta V. Pigmented dermatofi brosarcoma protuberance (Bednar tumor). Eur J Dermatol. 2008;18:98-100.
6. Kaur M, Dalal V, Bansal A. Bednar tumour: an infrequent diagnosis. Brit J Med Pract. 2015 ;8:13-6.
7. Tan AW, Tan SH. Dermatofibrosarcoma protuberans: a clinicopathological analysis of 10 cases in Asians. Aust J Dermatol. 2004;45:29-33.
8. Mondal I, Chaudhuri A, Samaddar A, Das N, Nag D. Pigmented Dermatofibrosarcoma Protuberans (Bednar Tumor) Masquerading Clinically as Malignant Melanoma: A Case Report. Ann Appl Bio-Sci. 2017;4:C1-3.
9. Fletcher CD, Theaker JM, Flanagan A, Krausz T. Pigmented dermatofibrosarcoma protuberans (Bednar tumour): melanocytic colonization or neuroectodermal differentiation? A clinicopathological and immunohistochemical study. Histopathology. 1988;13:631-43.
10. Tiwary AK. Bednar tumor: a rare neuromesenchymal neoplasm in a rare location. Iran J Dermatol. 2021;24:346-9.