Bednar tumor: a rare neuromesenchymal neoplasm in a rare location

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Bednar tumor or pigmented dermatofibrosarcoma protuberans is a rare slow-growing dermal tumor of neuromesenchymal origin having low malignant potential. It usually presents as a black, firm plaque or exophytic nodule on the back or shoulder, mostly in black people in their third to fourth decades of life. The head, neck, and extremities are less common sites. Herein, we reported a 55-year-old female presenting with a well-defined, dark-colored, ulcerated, painful plaque on the fifth toe of the right foot that had developed since two years beforehand. Malignant melanoma and dermatofibroma were considered as clinical differential diagnoses. An incisional biopsy was done. Histopathologic evaluation showed dermal proliferation of plump elongated cells and spindled cells (with oval to elongated hyperchromatic nuclei and scanty eosinophilic cytoplasm) arranged in storiform pattern and sheets. Melanin-laden dendritic cells were also present, interspersed with neoplastic elongated and spindle-shaped cells. Based on these clinical and histopathologic features, a diagnosis of Bednar tumor was made.

Keywords: neoplasm, Bednar tumor, dermatofibroma

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INTRODUCTION

Bednar tumor is a rare pigmented variant of dermatofibrosarcoma protuberans (DFSP), with low-to-intermediate grade malignant potential. It mostly presents as a slow-growing, solitary, well-defined, dark-colored, firm plaque or nodule, commonly involving the upper trunk in middle-aged black individuals [1]. Other less common sites are the head, neck, and limbs. Herein, we report a case of Bednar tumor involving the fifth toe of the right foot in an Indian female.

CASE PRESENTATION

A 55-year-old female presented with a painful, black-colored, ulcerated lesion on the fifth toe of the right foot. It started with an asymptomatic, hyperpigmented papule two years back, which slowly increased in size and became ulcerated and secondarily infected. Although the patient was treated with many systemic antibiotics (cephalosporins and metronidazole) by local medical practitioners on multiple occasions, the lesion never healed. On local cutaneous examination, the presenting lesion was seen as a well-circumscribed, bluish-black, ulcerated, tender plaque of approximately 3 cm × 2.5 cm in size involving the medial side of the little toe of the right foot (Figure 1). It also appeared macerated due to improper bandaging and contact with water. The plaque surface was firm at the margin and soft at the center, oozing pus. There was no history of trauma, surgery, or preexisting dermatoses at the same site. Inguinal lymph nodes were not enlarged. All routine investigations and laboratory parameters were within normal limits and non-contributory. There were no radiological abnormalities in the

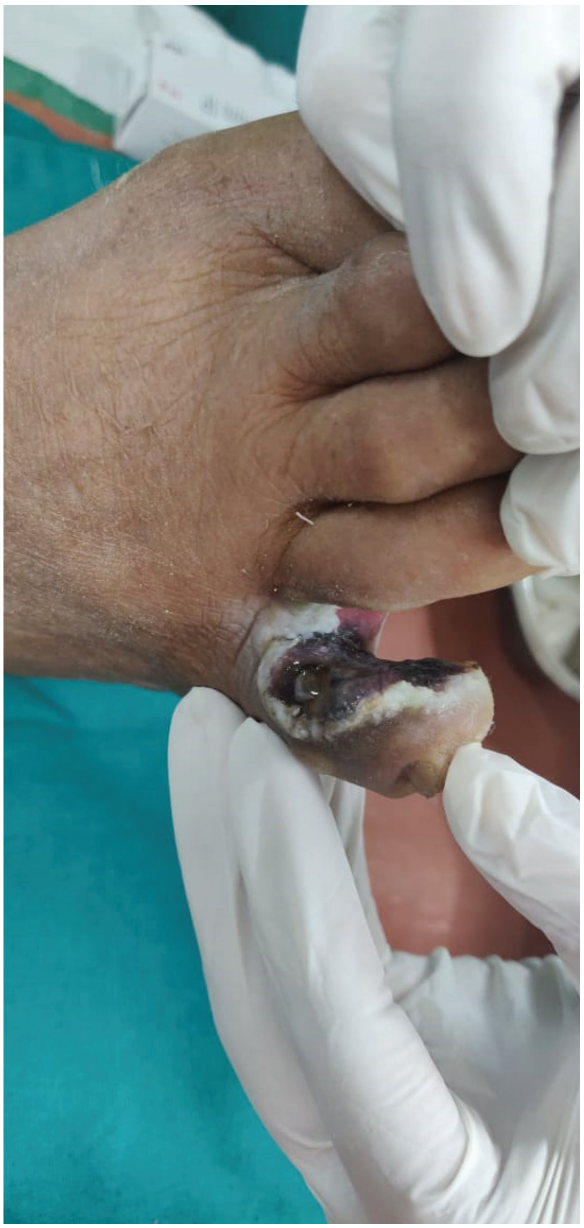

Figure 1. A well-circumscribed, black-colored, ulcerated plaque on the medial side of the 5th toe of the right foot.

affected toe. Based on the clinical presentation, malignant melanoma and dermatofibroma were kept as differentials. An incisional biopsy was taken from the margin of tumor for histopathologic examination to confirm the diagnosis.

Histopathologic evaluation showed dermal proliferation of plump elongated cells and spindled cells (with oval to elongated hyperchromatic nuclei and scanty eosinophilic cytoplasm) arranged in

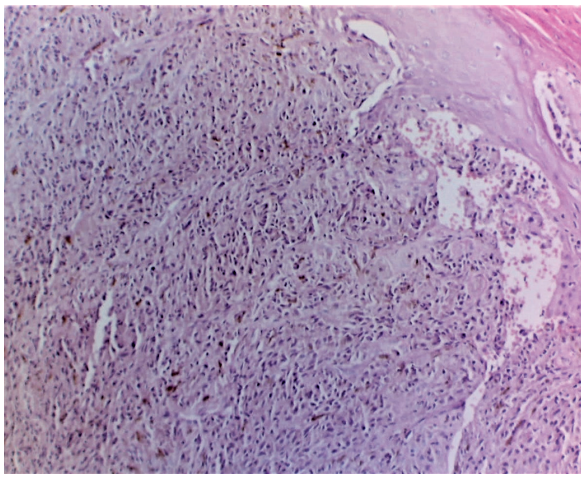

Figure 2. Plump elongated and spindled cells arranged in sheets/storiform pattern in the whole dermis along with many interspersed melanin-laden dendritic cells embedded in the collagenous stroma (H & E, × 200).

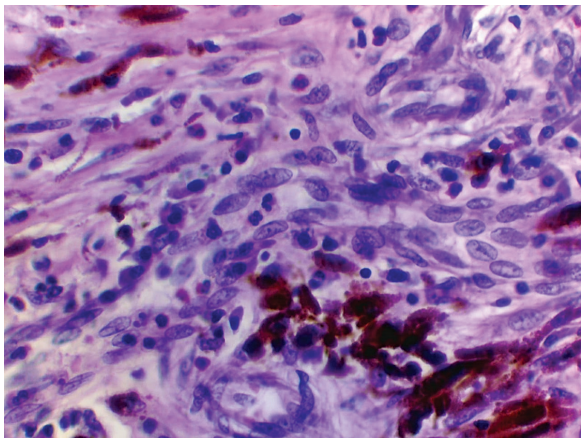

Figure 3. Neoplastic cells with oval and elongated hyperchromatic nuclei and scanty eosinophilic cytoplasm (H & E, × 400).

storiform pattern and sheets (Figure 2). Many interspersed melanin-laden dendritic cells were also noted. Intervening stroma revealed delicate and thick collagen fibers. Mitotic activity and cytologic atypia were not seen (Figure 3).

Although immunohistochemical staining could not be performed on the specimen due to the financial constraint of the patient, clinical and classical histopathologic findings were sufficient to diagnose it as pigmented dermatofibrosarcoma protuberans or Bednar tumor. Then, the patient was referred to the oncology department for wide excision, but she did not turn up in our department after that.

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive neoplasm with low to intermediate malignant potential accounting for about 1% of soft tissue sarcomas [2]. It was first identified as a progressive and recurrent dermatofibroma by Darier and Ferrand in 1924. Hoffman coined the term DFSP in 1925 due to its tendency to develop protruding nodules [1]. There are several histologic variants of this tumor such as pigmented, myxoid, granular cell, palisaded, atrophic, DFSP with fibrosarcomatous areas (DFSP-FS), DFSP with areas of giant cell fibroblastoma (GCF) or foci of myoid/myofibroblastic differentiation, and the sclerosing variant [3].

Pigmented dermatofibrosarcoma protuberans or Bednar tumor, first described by Bednar in 1957 as storiform neurofibroma, is also a rare variant constituting approximately less than 5% of all cases of DFSP. It mostly occurs in the third and fourth decades of life with a slight male preponderance and is common in black people. The back and shoulder are most commonly involved, followed by the extremities, head, and neck [2].

The cellular origin of Bednar tumors is still uncertain. Previous histologic study and immunoreactivity for S-100, presence of melanin-containing dendritic cells, and cells suggestive of Schwannian differentiation suggested it to be of neuroectodermal origin with secondary reactive melanocytic colonization from the dermo-epidermal junction [4]. As there are two different subsets of cell populations within the tumor, namely spindle shaped fibroblasts (strongly immunoreactive for CD34 and negative for S100) and dendritic pigmented cells (negative for CD34 and strongly immunoreactive to S100), the current concept is to consider it of neuromesenchymal origin with bi-directional differentiation [5].

Clinically, it presents as a slow-growing, solitary, hyperpigmented plaque with an irregular surface. Depending upon the density of the melanin-containing dendritic cells, its color may vary from blue to black [2]. Occasionally, it can be non-pigmented due to very scanty pigment. Over a period of months or years, the plaque may become exophytic and multinodular, slowly invading the subcutaneous tissue or fascia [3]. Malignant transformation and distant metastasis are very rare.

Histopathologically, Bednar tumor is characterized by irregular, short, intersecting fascicles of spindle-shaped tumor cells embedded in the collagenous stroma, arranged in a storiform/spiral pattern, and admixed with a variable population of melanin-containing dendritic cells. The tumor cells show little or no pleomorphism with a low mitotic rate [6]. On immunohistochemistry, spindle-shaped cells and melanin-containing dendritic cells stain positive with CD34 and S100, respectively [7].

Differential diagnoses include pigmented neurofibroma, dermatofibroma, psammomatous melanotic schwannoma, and malignant melanoma. Pigmented neurofibroma does not appear dark-colored or black due to only the microscopic presence of melanin. Additionally, it does not show a storiform pattern and is negative for CD34. Dermatofibroma is distinguished from Bednar tumor 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 is quite different from Bednar tumor, so it can easily be ruled out [1].

The treatment approach and prognosis vary with the location of the lesions. Wide surgical excision is the most successful procedure for truncal tumors. Lesions on the head, neck, and extremities are difficult to excise with wide margins without compromising cosmesis. Additionally, postsurgical recurrence is high. Radiation or chemotherapeutic agents such as imatinib mesylate can reduce tumor size [8]. Moh's microsurgery is a better alternative to wide excision as it minimizes the surgical anatomic defects.

CONCLUSION

As melanoma and dermatofibroma are much more common entities than Bednar tumors, histopathology is required to confirm the eluding diagnosis. To add, the rarity of this case and the involvement of an atypical site encouraged us to report this case.

Conflict of Interest: None declared.

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