Case Report

Pigmented Dermatofibrosarcoma Protuberans (Bednar Tumor) Masquerading Clinically as Malignant Melanoma: A Case Report

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ABSTRACT

Bednar tumor is a rare neoplasm of intermediate malignant potential, which account for less than 5% of dermatofibrosarcoma protuberans (DFSP). This tumor is a pigmented variant of DFSP. The diagnosis is commonly made in early to mid adult life except in cases with melanin containing cells. The case report presents a 19 years old female who presented with a painless slow-growing 3.5×3×1.2 cm blackish mass on the dorsal aspect of her right foot. Histopathological examination of the biopsy specimen revealed typical features of a Bednar tumor, the diagnosis was confirmed through immunohistochemical study.

Keywords: Dermatofibrosarcoma Protuberance (DFSP), Bednar Tumour, Immunohistochemistry

Introduction

Bednar tumor, a variant of dermatofibrosarcoma protuberance (DFSP), is a rare neoplasm of intermediate malignant potential described by Bednar in 1957. It accounts for less than 5% of all cases of dermatofibrosarcoma protuberans (DFSP).1,2 The lesion comprised of heavily melanin pigmented dendritic spindle cells arranged in a storiform pattern with elongated nuclei and a scant to moderate amount of cytoplasm.3 Mitotic activity was sparse. The lesion has an infiltrating margin and involved the sub-cutaneous tissue. Immunohistochemical stains in most cases show positivity for vimentin and CD34.4,5,6 We report a rare case of a Bednar tumor in a 19-year-old patient.

Case Report

A 19-year-old female presented with a slow growing painless soft tissue mass on the dorsum of the right foot which was present since childhood as single black spot. Gross examination revealed single irregular globular partially skin covered tissue piece, measuring 3.5 cm in the greatest dimension.

Light microscopic examination of hematoxylin-eosin stained sections, of biopsy specimen, showed spindle cell tumor with moderate cellular pleomorphism occupying the dermis and subcutaneous tissue. There is grenz zone between overlying epidermis and tumor mass. The tumor cells show intracytoplasmic brown pigments of melanin. The basal layer of epidermis does not show any junctional activity. However there is increase melanin pigment in the dermis and presence of melanophages in the papillary dermis. Fatty tissue and dermal appendages are entrapped within the tumor. (Fig.1A, B, C)

Immunohistochemically, the spindle tumor cells were diffusely positive for vimentin and CD34 antigen. (Fig.2A, B). The melanin containing pigment cells were positive for S-100 protein (Fig. 2C) where as tumor cells show negativity for HMB-45 (Fig. 2D).Thus, the diagnosis of Bednar tumor (pigmented DFSP) was confirmed.

Fig. 1: A-Photomicrograph showing spindle cell tumor occupying dermis and subcutaneous tissue and entrapped adipose tissue (X40).B, C- Photomicrograph showing intracytoplasmic brown pigment within spindle tumor cells (X200).
Initially designated “storiform neurofibroma” by Bednar in 1957, this variant of dermatofibrosarcoma protuberans contains abundant melanotic pigment. It usually occurs after 20 years of age, but it is also found in the children, and called as congenital Bednar tumor.\textsuperscript{1,2} Some reports suggest a slight male predominance, others demonstrate an equal distribution between the sexes. It has been described in all ethnic groups; however it is generally more prevalent in blacks.\textsuperscript{7} The lesions present as slow growth, over a period of months to years. The most frequent location is in the trunk, also involved upper and lower extremities, or the head and neck region.\textsuperscript{7}

Bednar tumors differ from the typical cases of DFSP by the presence of heavily melanin pigmented dendritic cells, the histiogenesis of which continues to be debated. Several investigators have suggested that this tumor is derived from neuroectodermal cells according to the ultrastructural and immunohistochemical findings as well as the presence of melanosome containing cells.\textsuperscript{7}

In immunohistochemical studies most of the tumor cells exhibit a positive reaction to CD 34 and vimentin, and are negative for HMB-45 and protein S-100. However, those cells containing melanin may react positively to protein S-100.\textsuperscript{6}

However, to date it is uncertain whether Bednar tumors are simply colonized by the melanin bearing cells or the tumor is derived from putative neuromesenchyme.

The differential diagnoses include other benign or malignant cutaneous pigmented neoplasms such as pigmented (melanotic) neurofibroma, psammomatous melanotic schwannoma, and desmoplastic (neurotrophic) melanoma.\textsuperscript{8}

A pigmented neurofibroma can be confused with a Bednar tumor because the melanin laden cells of both processes are similar. However, the Bednar tumor exhibits a more extensive storiform growth pattern, has greater immunoreactivity for CD34 and lacks diffuse proliferation of S-100 protein positive Schwann cells.

Psammomatous melanotic schwannoma is rather circumscribed, heavily pigmented with psammomatous bodies and diffusely positive for S-100 protein, whereas the Bednar tumor is poorly circumscribed and composed of CD34 positive spindle shaped cells with scattered pigmented cells.

Desmoplastic (neurotrophic) melanoma shows a neurotropism, focal melanocytic junctional activity, and diffuse and strong S-100 protein, HMB 45 immunoreactivity, CD34 negativity, whereas the Bednar tumor does not show any juctional activity, diffuse and strong CD 34 immunoreactivity.

In addition to a careful histology examination, immunohistochemical study for CD34 is the most useful marker for differentiating a Bednar tumor from other cutaneous pigmented tumors.
The typical DFSP has a rate of recurrence ranging from 20-50% among reports with a long-term follow up. The biologic behaviour of Bednar tumor is less aggressive than typical DFSP. However the rare case of metastatic Bednar tumor has been reported in literature. Mochzuchi cited a recurrence rate of 17% among reported cases of Bednar tumors. In addition, they reported an average interval of 9 years for recurrence (range, 9 months to 23 years). Although it is difficult to assess the biologic behavior of this tumor in pediatric patients because of the rarity of cases.

The recommended treatment for DFSP or Bednar tumors in the adult or pediatric patient is Mohs’ micrographic surgery, wide excision with more than 2-3 cm margins of visibly uninvolved tissue and inclusion of the superficial fascia. Mohs’ surgery is one of the many methods of obtaining complete margin control during removal of a skin cancer (CCPDMA- Complete circumferential peripheral and deep margin assessment using frozen section histology).

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