Case Report

Bednar tumor

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ABSTRACT

Pigmented dermatofibrosarcoma protuberance (DSFP), also known as Bednar tumor or formerly known as pigmented storiform neurofibroma is a rare variant of DSFP. Bednar first described it in 1957. The histology is striking because of the presence of multipolar pigment laden dendritic cells scattered among spindle shaped cells with a storiform pattern, the latter being characteristic of DSFP. The spindle cells are positive for CD_{34}, and Vimentin and negative for S_{100}. The pigment represents melanin. We describe such a tumor because of its rarity and very striking microscopic appearance.

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In 1957, Bednar[1] described a type of cutaneous tumor characterized by indolent growth, a prominent storiform pattern and the presence of melanin pigment. Some of these lesions are non pigmented, and these resemble the conventional dermatofibrosarcoma protuberance (DSFP) with which they share many gross and clinical features. The tumor usually occurs on the trunk, especially the shoulder region. The other sites include head and neck, upper and lower extremities. Most of the tumors are slow growing. Grossly they are broad based and have a fleshy slate gray or black lobulated appearance. Microscopically, they are composed of spindle shaped cells arranged in tight whorls that give a distinct storiform appearance. Scattered in between these cells, bipolar or multipolar dendritic cells are seen, which show mature melanosomes and premelanosomes on electron microscopy. This paper records our findings, and the varied immunohistochemical and electron microscopic findings are discussed.

Case Report. A 35-year-old male patient presented with a slow growing nodular swelling on the right forearm of 2 years duration. On examination, it was skin covered, measuring 3x2 cm with softening and ulceration in the center. The surrounding skin showed evidence of Wassam treatment. The tumor was excised and sent for histopathological examination. The specimen was a circular skin covered piece with a raised nodule on the surface measuring 1.5 cm in circumference. The cut surface showed a slate gray fleshy lesion, with a broad base measuring 2.5x2x2 cm. Microscopy showed a cellular tumor composed of spindle shaped cells arranged in a definite storiform pattern. The cells had hyperchromatic oval to spindle shaped nuclei with occasional mitotic figures. Scattered in between, there was characteristic deeply pigmented bipolar or multipolar cells with tentacle like processes extending into the surrounding stroma (**Figure 1**). Immunohistochemical stains showed positivity of the spindle shaped cells for vimentin and CD_{34}. Cytokeratin, smooth muscle actin and S_{100} were negative. We concluded that this was a pigmented DSFP (Bednar tumor).

Discussion. Bednar tumor accounts for less than 5% of all cases of DFSP.[2] It is more common in black patients and clinically indistinguishable from the latter.[3] It was originally described by Bednar in 1957, who thought that this tumor

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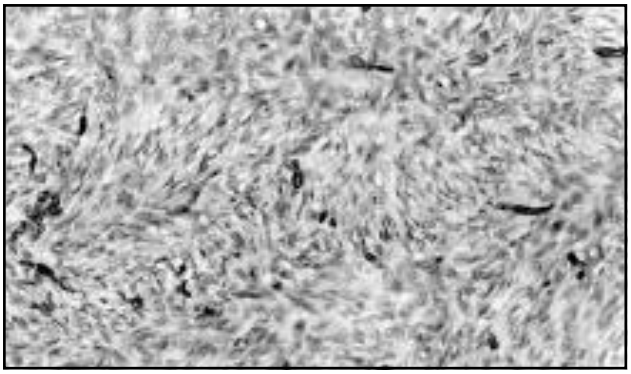

Figure 1 - Hematoxylin and eosin stain showing spindle shaped cells arranged in storiform pattern mixed with multipolar pigmented cells.

represented neural lesions. However, their non-pigmented portions are identical to the conventional DFSP. Microscopy shows a cellular lesion composed of spindle shaped cells arranged in short fascicles that form a distinct storiform pattern, similar to the conventional DFSP. But, the striking feature is the presence of pigmented bipolar or multipolar dendritic cells with tentacle like processes emanating from a nucleus containing zone. The pigment is tinctorially similar to melanin. The differential diagnosis includes fibrous histiocytoma, neurofibroma, malignant melanoma and cellular blue nevus. This tumor is positive for Vimentin and CD_{34}. S_{100} will be positive in neurofibroma and melanoma, but negative in this tumor. Fibrous histiocytoma is CD_{34} negative.

Some authors noted that in the dendritic melanin containing cells, antibodies against Vimentin were positive whereas HMB_{45} was negative.[4] Some investigators have reported positive reactions for antibodies against S_{100} and neuron specific enolase in the pigmented cells, whereas one case was reported negative.[5] Our case showed negative staining against S_{100}. Kagoura et al[6] demonstrated that Factor XIIIa was expressed on tumor cells around the melanin containing cells, which were positive for S_{100} and Vimentin. This suggested that the phenotype of the tumor cells around the melanin containing cells differ from the other tumor cells, probably resulting from the relationship of the tumor cells and the melanin containing cells.

Ultrastructurally, the tumor is composed mainly of nondescript spindle shaped mesenchymal cells resembling fibroblasts. A few pigment-laden cells are localized with the general appearance similar to the non pigmented cells.[7] Controversy persists with regard to the nature of the pigment laden cells. Non uniform ultrastructural and inconsistent immunohistochemical findings have generated diverse theories regarding the histogenesis. Dupree[1] proposed a neuroectodermal origin based on electron microscopy findings in which the pigmented cells with premelanosomes, mature melanosomes and definite basal lamina were consistent with Schwann cells.[1]

Recently, based on immunohistochemical studies, a new theory of dual cell origin was proposed. The author speculated that Bednar tumor originated from 2 different cell lines; CD_{34} positive spindle cells of mesenchymal origin and pigmented cells of neuroectodermal origin.[8] Seo et al[7] in their study, were unable to demonstrate ultrastructurally, the intermediate or transitional forms between the fibroblastic and the pigmented cells, thus, favoring the hypothesis of dual cell origin. More studies have to be carried out at an ultrastructural level to gain information on the histogenesis of Bednar tumors, especially the pigmented cells. Until then, the true nature of this tumor will remain elusive.

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