

Case Studies

Bednar Tumor: An Uncommon Entity

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Key Words

Bednar tumor · Pigmented dermatofibrosarcoma protuberans · Differentiation

Abstract

Bednar tumor is an uncommon variant of dermatofibrosarcoma protuberans. Also known as pigmented dermatofibrosarcoma protuberans, this tumor is of intermediate grade. It is seen in adults and has a predisposition to affect the shoulder region. We report a rare case of Bednar tumor in a 40-year-old female patient. The diagnosis of Bednar tumor must be considered while reporting pigmented subcutaneous spindle cell lesions.

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Published by S. Karger AG, Basel**Introduction**

Bednar tumor or pigmented dermatofibrosarcoma protuberans (DFSP) is a rare, aggressive, cutaneous tumor that constitutes 1–5% of all DFSPs [1, 2].

Case History

A 40-year-old female presented with right shoulder swelling present since few months and gradually increasing in size. A complete excision was performed without any prior fine needle aspiration cytology or needle biopsy. The tumor was just beneath the skin, fairly circumscribed, non-capsulated extending into the subcutaneous fat and measuring 5 × 4 × 4 cm. Grossly, focal areas of pigmentation were noticed towards the periphery of the tumor (fig. 1). On histopathology, the tumor was a spindle cell lesion abutting the skin and invading the underlying subcutaneous fat. The cells were arranged in storiform pattern and short interlacing fascicles. There were scattered pigment-laden cells throughout the lesion (fig. 2). On higher magnification, the spindle cells were uniform appearing with a moderate amount

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Fig. 1. Gross photograph showing a fairly circumscribed lesion with areas of pigmentation towards the periphery of the tumor.

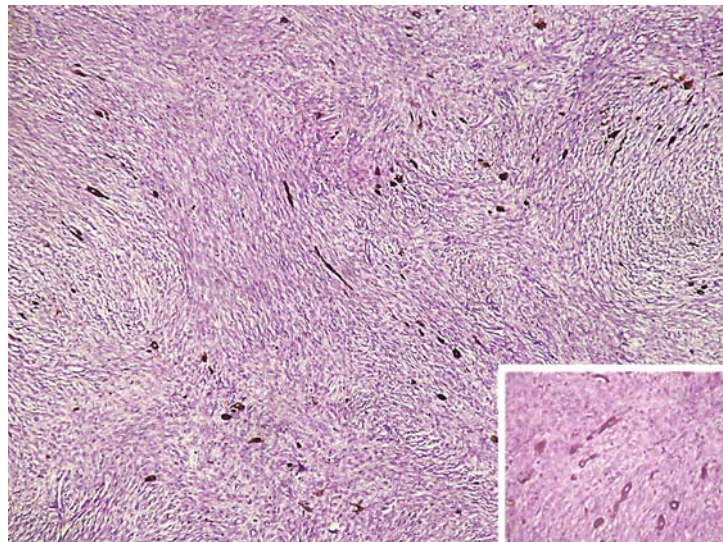


Fig. 2. Microphotograph showing a spindle cell lesion; cells being arranged in storiform pattern and short interlacing fascicles (HE, ×100) with numerous scattered pigment-laden cells throughout the lesion seen in the inset (HE, ×400).

of eosinophilic cytoplasm and plump nuclei with tapering edges. There were 1–2 mitotic figures per 10 high power fields. The pigment-laden cells showed coarse brown black pigment obscuring the nuclei and positivity for Masson-Fontana stain. No areas of necrosis or hemorrhage were noted. The above-mentioned features lead to the diagnosis of Bednar tumor. Currently the patient is on regular follow-up without any recurrences.

Discussion

Bednar tumor is a rare pigmented variant of DFSP first described in 1957. It is considered to be a tumor of intermediate grade and very infrequently reported in the literature [1–5]. It is seen in young to middle-aged adults in the fourth decade with very occasional cases in the pediatric age group [4, 5]. The preferred sites are the shoulder region as in our case as well as the trunk, extremities and the head and neck [1–5]. Grossly, the tumors have been described to be multilobular with pigmentation, irregular surface and firm nodules growing deep within

the subcutaneous tissue. In this case, the cut surface of the fairly circumscribed tumor showed trabeculations and peculiar pigmentation in the subepidermal portion, which has not been described in the literature.

The pigment-laden dendritic cells distinguish this lesion from conventional DFSP. It has to be distinguished from other pigmented cutaneous spindle cell lesions like pigmented neurofibroma, psammomatous melanotic schwannoma, neurocristic cutaneous hamartoma and desmoplastic malignant melanoma. However, our case had classic features of DFSP, and the above lesions were ruled out on histopathology itself.

Neuro-ectodermal differentiation or melanocytic colonization are the two proposed theories for histogenesis for the Bednar tumor. It has also been reported in association of dermal melanocytosis (nevus of Ito), and based on the immunohistochemistry, the cell of origin is thought to be a neuromesenchymal cell. Bednar tumor can rarely undergo malignant transformation in form of fibrosarcoma with repeated recurrences and distant metastasis. Hence, a close follow-up of these cases is always necessary, which is being done in our case. To conclude, though rare, it is important for the histopathologists to be aware and recognize this unusual entity and distinguish it from other pigmented spindle cell lesions.

Statement of Ethics

Ethics committee approval and patient consent has been obtained. Published research complies with the guidelines for human studies and animal welfare regulations. The patient has given informed consent, and the study protocol has been approved by the institute's committee on human research.

Disclosure Statement

The authors have no conflicts of interest to disclose.

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