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DERMOSCOPY OF THE MONTH
Bednar Tumor – a Case Report

Slađana Cekić[1*], Andrija JOVIĆ[1], Danijela POPOVIĆ[1], Zorana ZLATANOVIĆ[1], Nataša VIDOVIĆ[2], Hristina KOCIĆ[1], Danica TIODOROVIĆ[1,3]

[1]Clinic of Skin and Venereal Diseases, Clinical Center of Niš, Serbia
[2]Center for Pathology and Pathological Anatomy, Clinical Center Niš, Serbia
[3]Faculty of Medicine, University of Niš, Serbia

*Correspondence: Slađana Cekić, E-mail: srcekic@mts.rs

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Abstract

Bednar tumor is a rare pigmented type of the dermatofibrosarcoma protuberans characterized histologically by the coexistence of two distinct cell populations, including spindle-shaped cells and melanin-containing dendritic cells. We report dermoscopic features of Bednar tumor observed in a 54-year-old female patient. The dermoscopy of Bednar tumor revealed a multicomponent pattern composed of a homogeneous blue-gray pigmentation with shiny white lines, structureless light-brown pigmented areas and a peripheral pigment network. The dermoscopic features observed in the present case are consistent with reported dermoscopic descriptions of Bednar tumor. Although dermoscopy may be suggestive of the diagnosis of Bednar tumor, pathohistological examination remains a gold standard for diagnosis.

Key words: Dermatofibrosaroma; Diagnosis; Dermoscopy; Skin Neoplasms; Case Reports; Treatment Outcome

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a slow-growing fibrohistiocytic tumor of a low-grade to intermediate malignancy, with a high propensity for a local recurrence and a low rate of distant metastasis (1-4). The pigmented DFSP, also known as Bednar tumor, is a unique subtype of DFSP, characterized histologically by the coexistence of two distinct cell populations, including spindle-shaped cells and particular melanin-containing dendritic cells (1). 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 (1, 5). Herein, we report dermoscopic features of a rare case of Bednar tumor.

Case Report

A 54-year-old woman presented with a 1-year history of solitary asymptomatic nodule on her right forearm that had demonstrated a slow but progressive enlargement over the previous 3 months. Her personal and family history regarding skin cancers was unremarkable. The patient did not report any recent medical history of the local trauma of the affected area. The clinical examination revealed a slightly elevated dark-blue to partially brown nodule of firm consistency, measuring 8x10 mm **(Figure 1 A).** Dermoscopically, the lesion displayed a multicomponent pattern composed of a homogeneous blue-gray pigmentation with shiny white lines, structureless light-brown pigmented areas and a peripheral patchy pigment network (**Figure 1 B).** Based on the clinical and dermoscopic appearance of the lesion, our provisional diagnoses were: atypical dermatofibroma, combined nevus, blue nevus and melanoma. The surgical excision with margins of 2-3 mm was performed. The histopathological examination revealed a storiform arrangement of the spindle-shaped cells admixed with the melanin-

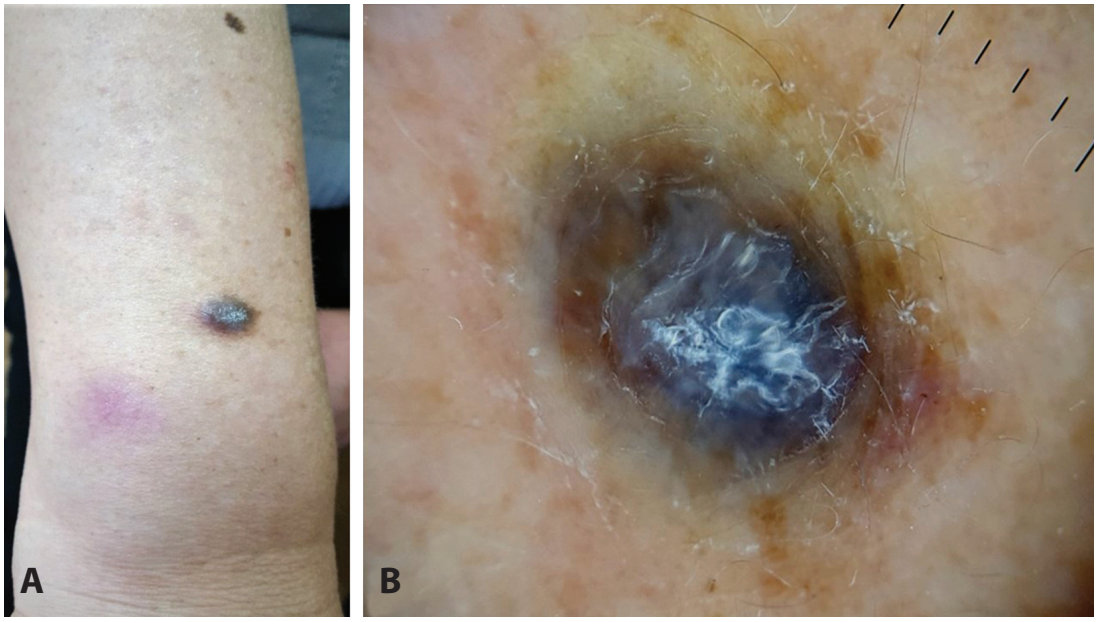

Figure 1. A. Dark-blue to partially brown nodule presented on the right forearm; **B.** Dermoscopic examination revealed a multicomponent pattern composed of homogeneous blue-gray pigmentation, shiny white lines, structureless light-brown pigmented areas and peripheral patchy pigment network

containing dendritic cells **(Figure 2 A, B)**. The additional immunohistochemical examination showed expression of CD 34 by the spindle-shaped cells **(Figure 2 C),** while pigmented cells were positive for S100 **(Figure 2 D),** corresponding to a diagnosis of the pigmented DFSP or Bednar tumor. Consequently, the patient underwent a surgical re-excision with 3 cm safety margins. During the period of 2 years of regular follow-up, there has been no evidence of recurrence.

Discussion

DFSP is a rare cutaneous neoplasm with the reported prevalence of 0.1-1% among all cutaneous malignant tumors (3). Besides conventional DFSF, more than 10 clinicopathological subtypes have been reported including myxoid, giant cell fibroblastoma or juvenile, granular cell, atrophic, palisading, as well as pigmented subtype. The pigmented subtype of DFSF or Bednar tumor shares similar pathohistological, immunohistochemical and cytogenetical features with the conventional DFSF, except for the presence of the melanin-containing dendritic cells (1). The origin of the melanin-containing dendritic cells in this tumor remains unclear: whether it originates from the neoplastic neuroectodermal differentiation, or represents an incorporated and/or colonized epidermal or hair follicle melanocytes (2-4).

A typical clinical appearance of Bednar tumor includes a solitary or multiple papules or nodules, mostly arising in the indurated plaque of tumor (typical protuberant morphology), exhibiting brown-red, blue or bluish-black coloration (1, 5). Usually, patients report a longstanding history of a slow-growing and asymptomatic lesion until it infiltrates the underlying muscle, fascia, or bones, causing discomfort. Initially, lesions may lack a protuberant clinical appearance only revealing a visible pigmentation (5, 6). At this early stage, Bednar tumor should be differentiated from dermatofibroma, blue nevus, melanoma and cutaneous metastases of various tumors. Moreover, Bednar tumor has been described in association with the history of a preceding local trauma, insect bite and vaccine scars (6, 7).

Given the rarity of DFSP, dermoscopic features of these lesions, especially in the context of Bednar tumor, have been insufficiently defined. In the largest study so far, Bernard et al. (8) performed a dermoscopic evaluation of 15 biopsy proven cases of DFSP whereas six der-

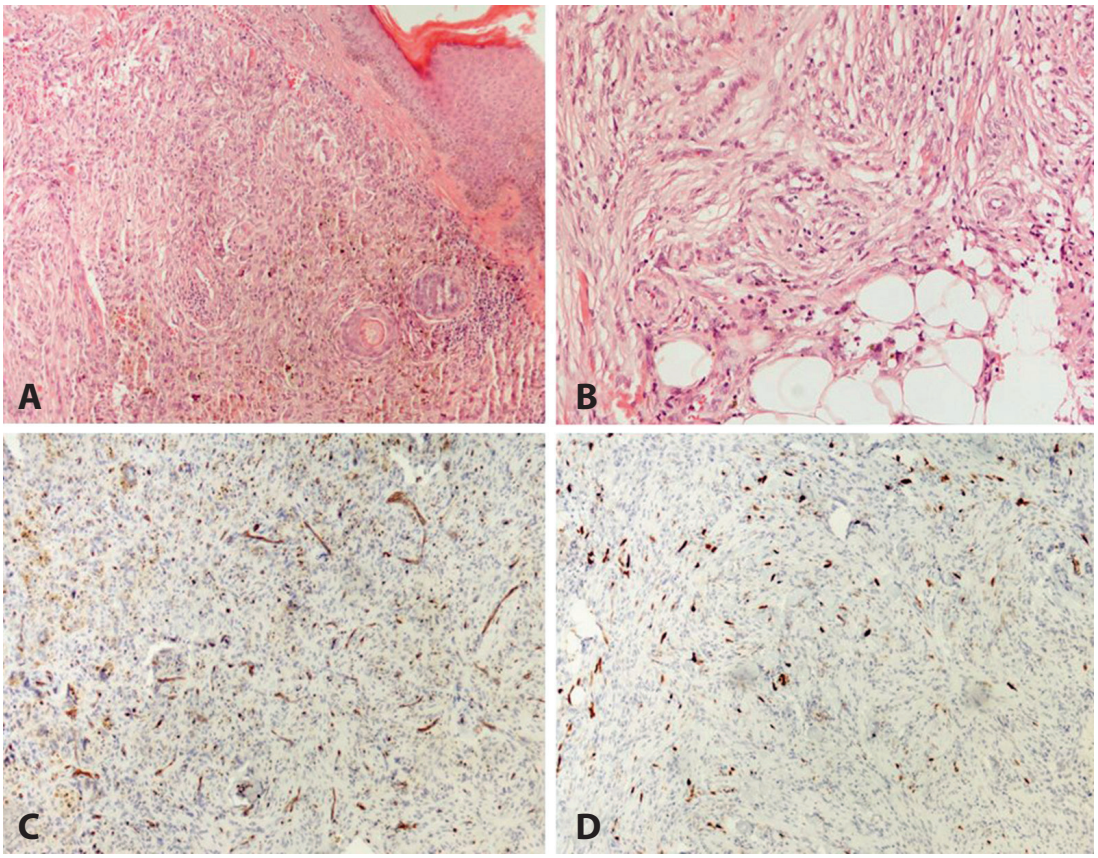

Figure 2. A. Spindle cells arranged in tight storiform pattern admixed with a small population of dendritic cells with melanin (hematoxylin and eosin; H&E x 20); **B.** Infiltration of the subcutaneous tissue (hematoxylin and eosin; H&E x 40); **C.** CD34 positive expression of spindle cells; **D.** Melanin-containing dendritic cells positive for S100

moscopic features of DFSP were observed, including: pigmented network, vessels, structureless light-brown areas, shiny white streaks, pink background coloration and structureless hypo- or non-pigmented areas. The previous study included a dermoscopic description of one Bednar tumor which revealed all 6 dermosocopic features mentioned above. Notably, the majority of the lesions analyzed in this study revealed a multicomponent pattern, which is highly suspicious of malignancy (8). In addition, Maeda et al. (9) reported another case of Bednar tumor which displayed similar dermoscopic features that had been observed by Bernard et al., except for a delicate pigmented network and vessels. In that article, authors stated that combination of "color variegation" and "blue-whitish veil lesions without a peripheral pigment network", in addition to the conventional dermoscopic features of DFSP, should suggest a diagnosis of Bednar tumor (9). Furthermore, Bednar tumor can mimic blue nevus both clinically and dermoscopically, revealing a homogeneous black-bluish pigmentation with white-veil structures (10).

The dermoscopic features observed in the present case are consistent with the reported dermoscopic descriptions of Bednar tumor (8-10). In this report, the blue-gray pigmentation was a prominent dermoscopic feature and it may reflect the histopathological finding of the melanin deposition in the dermis, while shiny white streaks may correlate to collagen alterations in the underlying stroma. Those dermoscopic features are commonly reported in the melanoma and blue nevus. The structureless light-brown pigmented areas and peripheral patchy pigment network may correspond to a distribution of melanin in the upper dermis and in the basal layer of the epidermis, respectively.

In conclusion, we presented a rare case of Bednar tumor with a dermoscopic description which is in line with previous reports. Although dermoscopy may be suggestive of the diagnosis of Bednar tumor, the pathohistological examination remains a gold standard for diagnosis.

Abbreviations

DFSP-dermatofibrosarcomaprotuberans

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Bednar tumor – prikaz slučaja

Sažetak

Bednarov tumor je retka pigmentna varijanta dermatofibrosarkoma protuberans koji histopatološki karakterišu istovremeno dve različite ćelijske populacije, uključujući vretenaste ćelije i dendritične ćelije koje sadrže melanin. Prikazujemo dermoskopske karakteristike Bednarovog tumora sagledane kod pacijentkinje starosti 54 godine. Dermoskopskim pregledom uočen je multikomponentni obazac sačinjen od homogene plavosive pigmentacije sa sjajnim beličastim linijama, bez strukturnih svetlobraon pigmentnih područija i periferne pigmentne mreže. Dermoskopske karakteristike uočene u ovom slučaju u skladu su sa objavljenim dermoskopskim opisima Bednarovog tumora. Iako dermoskopija može sugerisati dijagnozu Bednarovog tumora, histopatološki pregled predstavlja zlatni standard za dijagnozu.

Ključne reči: Dermatofibrosarkom; Dijagnoza; Dermoskopija; Kožne neoplazme; Prikazi slučajeva; Ishod Terapije

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