CASE REPORT

Pigmented dermatofibrosarcoma protuberans, Bednar tumor: Case report and Dermatoscopic findings

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ABSTRACT

Pigmented dermatofibrosarcoma protuberans, or Bednar tumor (BT) accounts for 1% of DFSP cases. It is a low-intermediate grade connective tissue neoplasm, characterized by slow growth, and a high tendency to local recurrence. Its etiology is related to a genetic alteration that affects the platelet-derived growth factor beta and the collagen type 1 alpha 1 gene, generating the COL1A1-PDGFB fusion gene. There are few Dermatoscopic reports describing its pigmented variant. A clinical case of Bednar tumor is presented, along with its Dermatoscopic findings.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a very rare locally aggressive soft tissue tumor, associated with high recurrence after surgical excision.[1] In addition to its classical form, more than 10 clinicopathological variants have been described, among which is the pigmented form.[2]

Pigmented dermatofibrosarcoma protuberans, or Bednar tumor (BT) as it is also known, accounts for 1% of DFSP cases. Its incidence is equal in men and women and is more frequent in black patients. Its etiology is related to a genetic alteration affecting the platelet-derived growth factor beta and the collagen type 1 alpha 1 gene, generating the COL1A1-PDGFB fusion gene.[3] Although there are some Dermatoscopic reports of DFSP, very few describe the pigmented variant. A clinical case of Bednar tumor is presented, along with its Dermatoscopic findings.

CLINICAL CASE

A 35-year-old female patient with a 4-year-old clinical picture presents with asymptomatic hyperpigmented grayish-blue round macule with diffuse margins, measuring approximately 1 cm, located on the left arm. In the last 6 months, such lesion evolves to an infiltrated, pigmented, bluish plaque, presenting with hard, central elevated region. (Fig. 1). The patient reports these past months the lesion has increased in size rapidly. Dermoscopy reveals (fig. 2) some pink, blue and light brown areas, as well as underlying hypopigmented areas. An incisional biopsy is performed, and the histopathological study reports pigmented dermatofibrosarcoma protuberans (Bednar tumor). This tumor compromises lateral and deep surgical margins. Consequently, the patient is referred to surgery for wide local excision. Resection is performed with safety

margins of 5cm. Both the 6 months and the year follow-ups, show no recurrences.

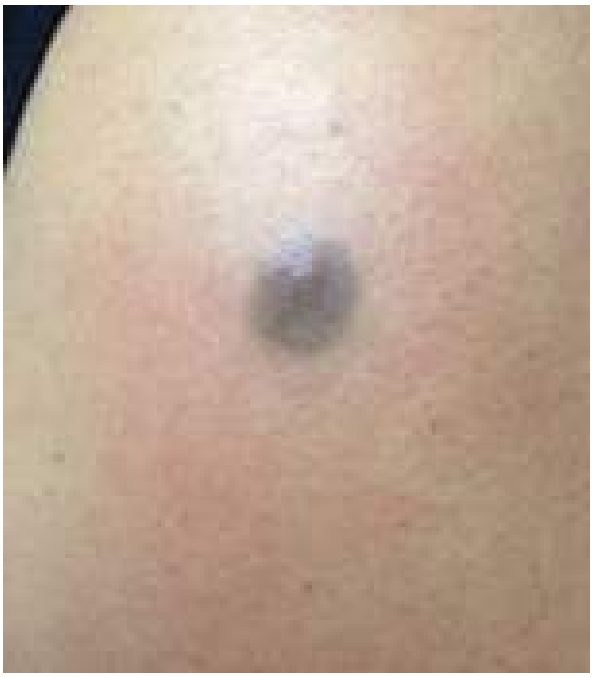

Figure 1. Clinical picture of the lesion, showing infiltrated, pigmented, blue plaque with central elevated area.

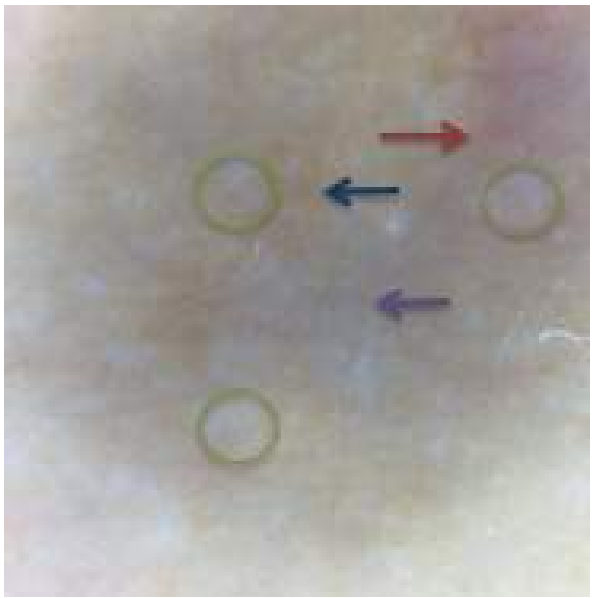

Figure 2. Dermoscopic picture showing delicate pigment network (blue arrows), pink background (red arrows), bluish areas (purple arrow) and hypopigmented areas (green circles).

DISCUSSION

Bednar tumor was described in 1957 by Blahoslav Bednar, who observed the presence of melanin in four cases of skin tumors of indolent growth and a whorl-like pattern in their histopathology. Consequently, they were considered as a variant of dermatofibrosarcoma protuberans.[4]

It is a low-intermediate grade connective tissue neoplasm, characterized by slow growth, and a high tendency to local recurrence. There is a high tendency to local recurrence, but metastases are very rare, occurring mainly hematogenously, with the lung being the site of greatest involvement.[5,6]

Its histogenesis is not clear, since it originates from an undifferentiated mesenchymal cell, with the ability to differentiate into fibroblast or histiocyte. It is also considered to have a neuroectodermal origin, due to the presence of dendritic cells.[3,7]

The incidence is equal in both men and women, and manifest more frequently between the second and fifth decade of life, although it can also appear in childhood. The predilection sites are the trunk and proximal part of the limbs. The condition manifests more frequently in black patients.[8,9] Local trauma, such as burns or bites, often precedes the appearance of this tumor.[3]

Clinically, a multinodular, infiltrated, pigmented plaque or tumor is observed. It is hard, of variable size, usually greater than 2 cm. It may sometimes appear as a sclerotic or atrophic plaque. Macroscopically, Bednar tumor has been described with different background colors, ranging from gray-whitish to black-bluish.[8]

Histopathology reveals dendritic cells with different proportion of melanin. These are S100 positive and interspersed between CD34-positive spindle cells, forming a whorl-like pattern. The presence of dendritic cells is what distinguishes BT from the classic form of DFSP.[10]

Dermatoscopic reports of DFSP are scarce, and within its findings have been described a pigment network, along with vessels, pink areas, underlying hypopigmented areas, bright white stretch marks, and light brown areas. Regarding the pigmented variant, a bluish background coloration has also been described. This is due to dendritic cells with melanic content.[11–14]

The pigment network corresponds to the increase of pigmented basal epidermal cells, instead of melanocytic proliferation, being an exception to the rule that the pigment network is an indicator of melanocytic lesions.[11]

The present case revealed predominant presence of bluish coloration in the background, with hypopigmented and pink areas, in addition to a delicate pigment network. Although blood vessels are not observed in the present case, it may be due to the bluish coloration, which hinders their visibility, as has been described in other cases.[12]

The treatment of Bednar Tumors is complete surgical excision, either by wide excision with minimum margins of 3cm, or Mohs surgery. The latter is considered the ideal treatment due to a lower rate of recurrence. Some authors report that radiotherapy reduces the risk of local recurrence in patients with DFSP, which, due to the characteristics of the tumor, already has a high probability of recurrence.[6,15]

Dermatoscopy is a useful tool for the detection of tumor lesions. Although there are no specific dermatoscopic findings for Bednar Tumors, the present case reports serve as a reference for diagnostic help and differentiation from other pigmented lesions.

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