Bednar Tumor: Treatment With Mohs Micrographic Surgery

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Bednar tumor is a rare variant of dermatofibrosarcoma protuberans (DFSP). However, the presence of pigment-bearing cells distinguishes it from DFSP. We report a case of Bednar tumor occurring in a 52-year-old white woman and review the typical clinical and histopathologic findings. To our knowledge, this is the second reported case of Bednar tumor successfully treated with Mohs micrographic surgery. The patient remains free of recurrence 5 years after treatment. Because of the clinical similarity between Bednar tumor and DFSP, we recommend Mohs micrographic surgery for treatment of this rare tumor.

CASE REPORT

A 52-year-old white woman presented with a lesion on her upper back, which had enlarged slowly during the previous 6 years. The lesion was asymptomatic, but the patient desired removal for cosmetic reasons. The patient had no previous history of skin cancer.

On physical examination, a 1.5-cm, firm, flesh-colored, asymptomatic nodule was noted on the right upper back (Figure 1). The lesion was nontender and had a cystic appearance. No other significant findings were noted upon complete skin examination, and the lesion was clinically diagnosed as an epidermal inclusion cyst.

During excision, an unencapsulated grey mass of tissue was found. Histopathologic results revealed a bland, infiltrating, spindle-cell neoplasm arranged in a storiform pattern (Figure 2). Scattered throughout the tumor were dendritic pigmented cells (Figure 3). The tumor infiltrated through subcutaneous tissue in a honeycomb pattern. The majority of the tumor stained with CD34 (Figure 4). The scattered pigmented cells stained with S-100 and Melan-A; HMB-45 and factor X111a results were negative.

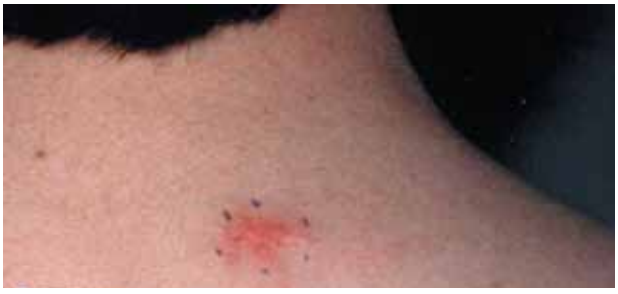

Figure 1. Patient with a 1.5-cm, multinodular, flesh-colored tumor on the right upper back.

After the diagnosis of Bednar tumor was made, the patient was referred for Mohs micrographic surgery (MMS). Two layers of MMS (approximately 12 mm) were required to remove the tumor. An additional layer was performed and processed in a horizontal manner with permanent sections and stained with CD34 to confirm the clear margins (Figure 4). The cells stained positive with S-100 and Melan-A (Figures 5 and 6) and negative with HMB-45 immunostains. A layered closure was performed. The patient has shown no sign of recurrence 5 years posttreatment.

COMMENT

Pigmented dermatofibrosarcoma protuberans (DFSP), or Bednar tumor, was first described in 1956.[1] It is considered

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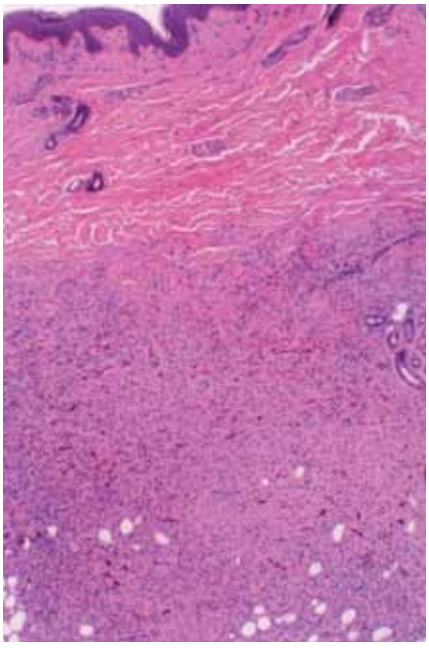

Figure 2. Biopsy specimen demonstrating a bland, infiltrating, spindle-cell neoplasm arranged in a storiform pattern (H&E, original magnification x10).

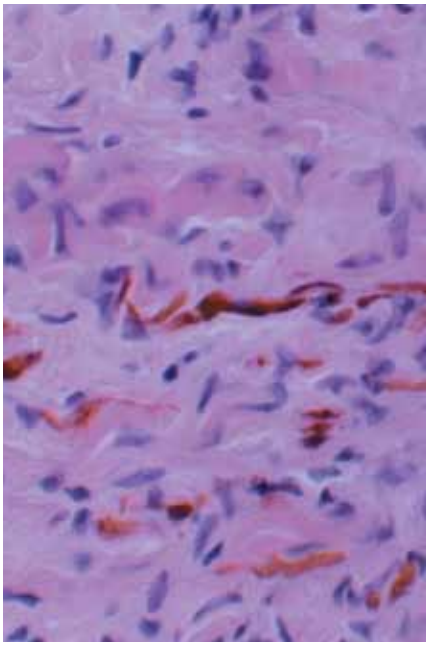

Figure 3. Higher power view demonstrating scattered dendritic melanocytes (H&E, original magnification x40).

a rare variant of DFSP, comprising less than 5% of the total number of cases.[2] As in DFSP, the pigmented variant has been reported to occur in the site of prior immunization, suggesting that Bednar tumor also may arise in sites of previous trauma.[3] The tumor presents mainly on the trunk, with an equal incidence in men and women. Bednar tumor can occur at any time from birth to age 66 years but presents most commonly in young to middle-aged adults.[4] The typical clinical appearance of Bednar tumor is a firm multinodular plaque. Histopathology shows a uniform array of spindle cells in a storiform pattern with variable amounts of pigment. In some cases, the amount of pigment causes the lesion to appear black. In other cases, the pigment may only be detected microscopically. The tumor may show honeycomb infiltration of subcutaneous adipose tissue, and electron microscopy reveals mature and immature melanosomes within pigmented cells. Bednar tumor stains positively with CD34 and is HMB-45 negative. The pigmented cells have been shown to stain with S-100, neuron specific enolase, and glial fibrillary acid protein.[2] Inconsistent staining results have been reported with Melan-A and MART-1.

The histogenesis of the pigment-bearing cells found in Bednar tumor is the subject of controversy. It has been proposed that pigment is produced by epidermal melanocytes, which have colonized the tumor.[4] Another hypothesis is that the presence of pigment-bearing cells indicates that DFSP is derived from neuroectoderm. To support the latter theory, Bednar[5] presented 3 cases of pigmented DFSP occurring within nevi. Enzinger and Weiss[2] demonstrated mature and immature melanosomes in the pigment-containing cells of Bednar

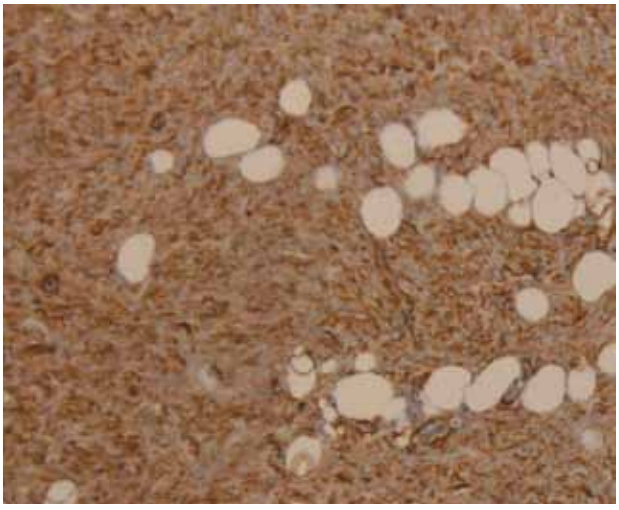

Figure 4. Strong diffuse CD34 staining of spindle cells (CD34 immunostaining, original magnification x10).

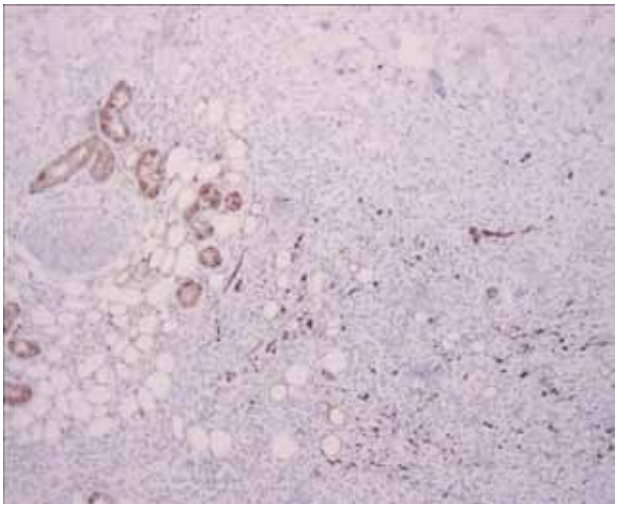

Figure 5. Scattered S-100 positive–staining pigmented cells (S-100 immunostaining, original magnification x10).

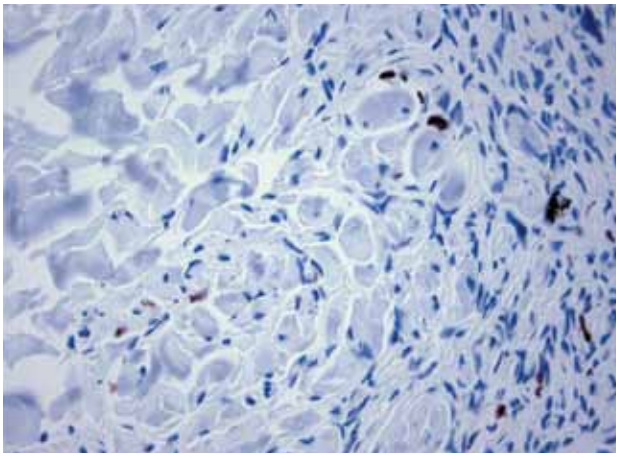

Figure 6. Scattered Melan-A positive–staining pigmented cells (Melan-A immunostaining, original magnification x10).

tumor, further supporting the theory of neuroectodermal derivation. There have been multiple reports in the literature of Bednar tumors in which the pigment-containing cells reacted positively to S-100.[4]

Bednar tumor is considered a tumor of intermediate malignancy. Although the rarity of this tumor has made it difficult to study, it is assumed, based on histopathologic similarity, to have the clinical characteristics of a typical DFSP. Bednar tumor is locally invasive, with slow growth and infrequent metastasis. There is a high rate of local recurrence after excision. Ding et al[4] found that Bednar tumor showed a lower rate of local recurrence than DFSP. A review of the literature revealed only 3 reports of Bednar tumor metastasis.[6-8]

The treatment of choice for DFSP is MMS. Several studies have shown a lower recurrence rate with MMS than with traditional 3-cm surgical margins.[9-11] With traditional wide excision, recurrence rates range from 10% to 20%. Recurrence rates for DFSP treated with MMS range from 2% to 6.6%.[9,11] DFSP typically tracks between collagen bundles and may extend deep into fascia and muscle. These tumor extensions are difficult to appreciate clinically and may be missed with traditional histopathologic tissue sectioning. MMS has the advantage of close examination of all tissue margins, better tissue conservation, and potentially superior cosmetic results.

Because of the documented sucess of MMS in the treatment of DFSP and the clinical similarities between DFSP and Bednar tumor, we chose to treat our patient with MMS. To our knowledge, there is one other report in the literature of a Bednar tumor treated with the MMS. This patient was tumor free 9 months posttreatment.[3] Our patient remains free of recurrence 5 years after treatment. In light of the slow growth of Bednar tumors, long-term follow-up is necessary.

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