

Case report



BEDNAR'S TUMOR – A RARE VARIANT OF DERMATOFIBROSARCOMA PROTUBERANS

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ABSTRACT

Bednar's tumor is a rare pigmented variant of dermatofibrosarcoma protuberans (DFSP)—a cutaneous sarcoma of low to intermediate malignancy, characterized by a tendency for local recurrence. Histologically, it is defined by the presence of spindle-shaped cells arranged in a storiform pattern and melanin-containing dendritic cells. Clinically, the lesions often resemble melanoma or other pigmented dermal tumors.

We present the case of a 66-year-old male with a long-standing, asymptomatic pigmented lesion in the right subclavicular region. Histopathological examination confirmed the diagnosis of Bednar's tumor based on its characteristic morphology and immunohistochemical profile: diffuse CD34 expression in spindle cells and S-100 positivity in melanin-laden components. Radical surgical excision with a 2 cm margin was performed. Follow-up over a two-year period revealed no evidence of recurrence or metastasis, as confirmed by PET/CT.

This case highlights the importance of early diagnosis, immunohistochemical distinction from melanoma, and the need for a multidisciplinary approach in managing rare cutaneous sarcomas with atypical presentation.

Keywords: dermatofibrosarcoma protuberans, Bednar's tumor, pigmented tumor, Immunohistochemistry, PET scan,

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive cutaneous sarcoma, accounting for approximately 1% of all soft tissue sarcomas and about 18% of cutaneous sarcomas [1]. The annual incidence of DFSP ranges between 0.8 and 4.5 cases per million individuals [2].

Bednar's tumor is a pigmented variant of DFSP, observed in approximately 1–5% of all cases [3]. It is characterized by the presence of melanin-containing dendritic cells among the spindle-shaped tumor cells [4].

We present the case of a 66-year-old male with pigmented DFSP (Bednar's tumor) located in the right subclavicular region. The patient underwent radical surgical treatment and was followed for two years without evidence of local recurrence or metastasis. Differential diagnostic considerations are discussed, including distinction from melanoma and other pigmented lesions. A brief review of the current diagnostic and therapeutic approach to DFSP and its variants is provided.

CASE PRESENTATION

We present the case of a 66-year-old male in good general condition, with no complaints at the time of examination. The skin in the right subclavicular region exhibited an irregular, grayish-brown surface covered with multiple telangiectasias. Scattered whitish nodules with smooth surface and firm consistency, measuring between 3 and 5 mm in diameter, were noted (Fig. 1). Centrally within the plaque, a nodule approximately 15 mm in size was visualized, showing increased firmness and multiple telangiectasias, non-tender upon palpation (Fig. 2). The patient reported that the skin changes had been present for over 10 years.

Fig. 1. Right subclavicular region.



Fig. 2. Nodules with a smooth surface and firm consistency



Physical examination findings: vesicular breath sounds without rales, regular heart rate without murmurs, blood pressure 130/80 mmHg, soft and non-tender abdomen. The only comorbidity identified was hypertensive heart disease, without signs of congestive heart failure.

A skin biopsy was performed from the lesion in the right subclavicular region. Histopathological examination of the excised specimen revealed: a skin fragment with adnexal structures and areas of epidermal atrophy, a mildly increased number of melanocytes in the basal layer, and a poorly demarcated dermal lesion composed of monomorphic oval, elongated, and wavy cells arranged around collagen bundles with no giant cells (Fig. 3). Areas of increased cellularity involving the subcutaneous fat were present (fenestrated pattern), along with scattered melanocytes (Fig. 4). Resection margins traversed cellular areas.

Fig. 3. Histologically, there is an area showing a predominance of oval-shaped cells. HE x 40

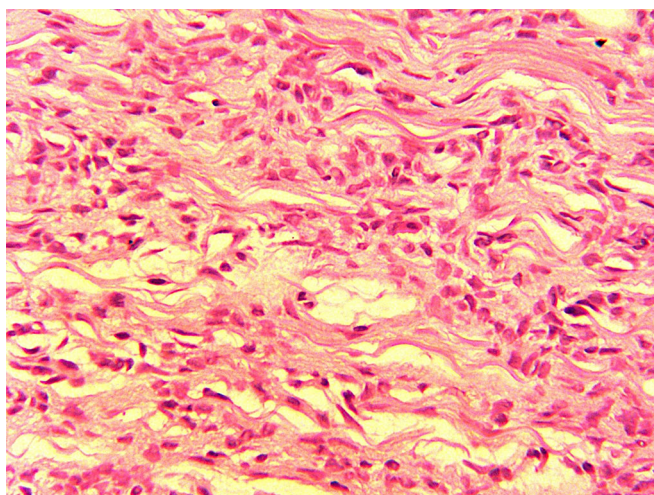
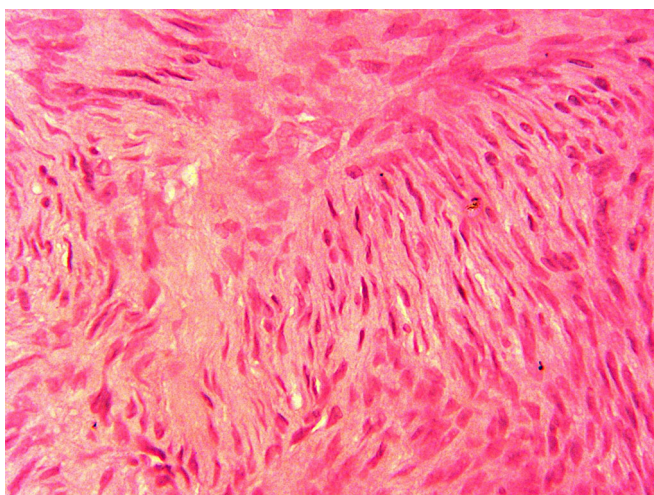


Fig. 4. Elongated cells arranged in interlacing bundles HE x 40



Histopathology of the excised specimen:

- Macroscopic findings: skin fragment measuring 6 × 7 cm, with irregular grayish surface and scattered whitish nodules (3–4 mm).

- Microscopic findings: skin and subcutis with zonal epidermal atrophy, pigment deposits in the basal layer, dilated vessels, and diffusely distributed soft tissue formations composed of monomorphic oval cells, without giant cells. Closest resection margin: 3 mm. Confirmed diagnosis: dermatofibrosarcoma protuberans – Bednar’s tumor.

Immunohistochemistry showed diffuse CD34 expression (Fig. 5), p53 expression in cellular regions, and S-100 positivity in melanocytes (Fig. 6). The morphological diagnosis was consistent with Bednar’s tumor – the pigmented variant of dermatofibrosarcoma protuberans.

Fig. 5. Immunohistochemistry: diffuse CD34 expression x 40

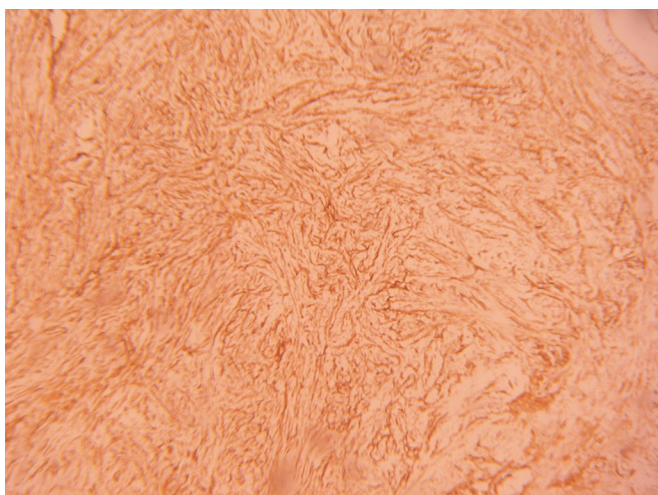
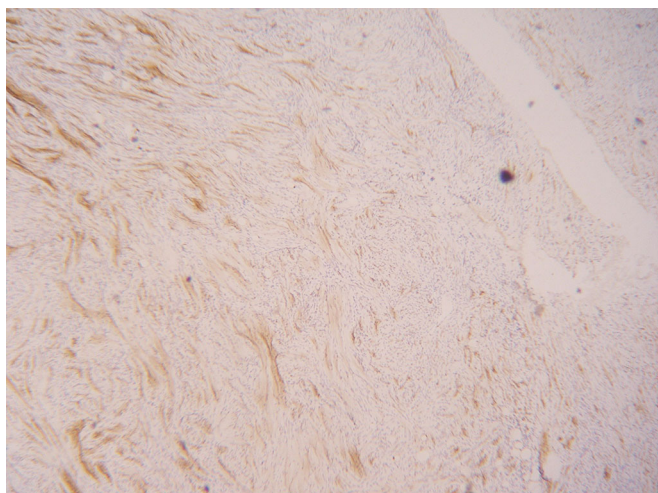


Fig. 6. Immunohistochemistry: focal S-100 expression x 40



At follow-up examination three months postoperatively, a linear scar measuring 8–9 cm was noted in the right clavicular region, with mild skin atrophy and telangiectasias. Regional lymph nodes (supraclavicular, infraclavicular, cervical, and axillary) were non-palpable.

Four months after the intervention, a follow-up PET/CT scan was performed with 2.7 mCi ¹⁸F-FDG (uptake time: 115 minutes). No metabolically active zones suspicious for local recurrence, regional lymphadenopathy, or distant metastases were identified. Clinical stage: cN0, cM0.

Additional findings: a small apical lesion in the left lung, appearing as a fibrotic focus; follow-up with contrast-enhanced CT was recommended in 3–6 months. Evidence of benign prostatic hyperplasia.

RESULTS

In April 2023, radical surgical excision of the skin lesion was performed with a 2 cm margin of healthy tissue under local anesthesia with Sol. Lidocaini 1%. Intraoperatively, multiple poorly defined tumor formations were observed in the right clavicular region. The postoperative period was uneventful, with primary wound healing (Fig. 7). Based on the clinical, histopathological, and immunohistochemical findings, the diagnosis of dermatofibrosarcoma protuberans, Bednar's tumor variant, was established.

Fig. 7. Two years after the surgery



DISCUSSION

Bednar's tumor represents a rare pigmented variant of dermatofibrosarcoma protuberans—a mesenchymal soft tissue tumor primarily involving the dermis and subcutaneous adipose tissue, and in rare cases extending into the underlying muscles and fasciae. The disease was described as a distinct clinicopathological entity as early as the late 19th century, and the term “dermatofibrosarcoma protuberans” was officially introduced in 1925 [5].

DFSP typically presents as a slowly growing, firm plaque or nodule, most commonly located on the trunk of young adults [6]. Although it can occur at any age, the peak incidence is around 40 years [7]. Geographic variations in incidence have been reported—for instance, in Eastern France, the frequency exceeds 3 cases per 1 million people annually [8]; in Sweden, it is approximately 4.0–4.4 cases per million per year [9]; and in Denmark, it reached up to 5.3 cases per million during the period 2000–2012 [10].

The etiology of DFSP is not fully understood, but a specific chromosomal translocation leading to fusion of the COL1A1 and PDGFB genes is believed to play a central role. This results in overexpression of platelet-derived growth factor and stimulation of tumor proliferation via activation of

signaling pathways such as MAPK and PI3K [11].

The pigmented subtype of DFSP—Bednar’s tumor—is distinguished by the presence of melanin-containing dendritic cells in addition to the classic spindle cells arranged in a storiform pattern [3,12]. The origin of the pigmented cells remains under discussion, with hypotheses including differentiation from melanocytes or mesenchymal progenitor cells with melanocytic potential [12].

Diagnosis is established through skin biopsy and histological examination, with the immunohistochemical profile playing a critical role. The tumor’s spindle cells are diffusely positive for CD34, while the pigmented cells express S-100 protein [3,13]. Melan-A and HMB-45 are typically negative, aiding in the distinction from melanomas and blue nevi [13]. The differential diagnosis of DFSP includes a broad spectrum of solid firm skin tumors (Table 1).

Table 1. Differential Diagnosis of Bednar’s Tumor [18,19].

Diagnosis	Clinical Presentation	Histology	Immunohistochemistry
Bednar’s tumor (pigmented DFSP)	Slowly growing plaque/nodule, often grayish-brown in color, telangiectasias, history >10 years	Spindle cells in storiform pattern, melanin-containing dendritic cells, subcutaneous infiltration (“honeycomb”)	CD34 (+) diffuse, S-100 (+) in melanocytes, Factor XIIIa (-), p53 (+) in cellular areas
Melanoma (spindle cell type)	Pigmented lesion, rapid growth, ulceration, pruritus, bleeding	High cellularity, nuclear pleomorphism, mitoses, melanin pigment, often with epidermal component	S-100 (+), HMB-45 (+), Melan-A (+), CD34 (-)
Pigmented dermatofibroma	Small, firm, often hyperpigmented nodule, stable over time	Collagen bundles, epidermal hyperplasia, “collagen trapping,” foamy histiocytes	Factor XIIIa (+), CD34 (-), S-100 (-/weak)
Fibrosarcoma (superficial)	Rapidly growing tumor, often painful, occasionally ulcerated	“Herringbone” pattern of spindle cells, mitoses, necrosis	Vimentin (+), CD34 (-/weak), S-100 (-), p53 (+)
Neurofibroma / Schwannoma	Soft, often non-inflamed nodule along nerve paths	Hypocellular tumor with spindle cells, neurofibrillary bundles	S-100 (+), CD34 (+/-), EMA (-), neurofilament (+)
Cutaneous leiomyosarcoma	Often painful nodule, dermal or subcutaneous location	Spindle cells with eosinophilic cytoplasm, “cigar-shaped” nuclei	SMA (+), Desmin (+), CD34 (-), S-100 (-)

DFSP is classified as a tumor of low to intermediate malignancy—with low metastatic potential but a high propensity for local recurrence [5,14]. Due to its infiltrative nature and subclinical extension, the preferred treatment modality is Mohs micrographic surgery, which offers maximal margin control with minimal loss of healthy tissue. In the absence of Mohs surgery availability, wide local excision with at least a 2–3 cm surgical margin is recommended [15].

In cases of inoperable, recurrent, or metastatic tumor, systemic therapy with a tyrosine kinase inhibitor (imatinib mesylate) is an effective alternative, approved for the treatment of DFSP harboring the COL1A1–PDGFB fusion gene [16,17].

The prognosis following complete surgical excision is favorable; however, if the surgical margin is inadequate, the risk of local recurrence remains high—up to

50% of cases. Metastases are rare, occurring in fewer than 5% of patients [6].

CONCLUSION

Bednar’s tumor represents a rare pigmented variant of dermatofibrosarcoma protuberans, combining the histological and molecular characteristics of classic DFSP with the presence of melanin-containing cells. Its clinical presentation often mimics that of other pigmented cutaneous lesions, including malignant melanoma, necessitating careful differential diagnosis and thorough histopathological and immunohistochemical evaluation.

The presented clinical case illustrates the typical course of the disease—characterized by a long-standing history, minimal symptoms, and a localized cutaneous lesion with distinctive morphology. Timely histological diagnosis and appropriate surgical treatment with radical

resection margins led to a favorable outcome, with no evidence of recurrence or metastatic spread on imaging follow-up.

Despite the favorable prognosis following complete resection, Bednar's tumor carries a potential for local aggressiveness and recurrence, particularly in cases of insufficient surgical margins. Therefore, long-term patient fol-

low-up is essential. The optimal therapeutic approach requires multidisciplinary collaboration among dermatologists, pathologists, oncologists, and surgeons.

The present case adds to the limited literature on the subject and underscores the importance of early recognition and appropriate management of this rare DFSP subtype, aiming to prevent diagnostic and therapeutic delays.

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