Dermatofibrosarcoma Protuberas: Case Report of a Bednar Tumor
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Case Report
A twenty-one year old Caucasian male was referred to our dermatology clinic for evaluation of a 2.5 x 1.4 cm black nodule located on his right anterior arm (Figure 1). The patient and his family were very concerned that he may have malignant melanoma as his maternal grandmother recently died from it. The lesion had been present for three years, and over the past several months had been growing in size. There were no associated symptoms with the lesion, and it previously has not been treated.

An excisional biopsy with 2 cm margins was obtained. Histopathology demonstrated a large asymptomatic deep spindle-cell neoplasm extending into the septa and lobules of the subcutis. Within the dermis, there were interlaced bundles of fibroblastic spindle-cells with pumil nuclei in a cartwheel or storiform appearance. There was also intermittent deposits of melanin containing dendritic cells (Figure 2). Immunohistochemical stains were diagnostically positive for CD34 (Figure 3), while S-100, MART-1, Factor X:11a (Figure 4), HMB-45 and AE1/AE3 were all negative. A diagnosis of a pigmented Dermatofibrosarcoma Protuberas, or Bednar tumor, was made. The patient was evaluated by an oncologist and declined radiation treatment. He was tumor free at his 6 month follow-up.

Discussion
Dermatofibrosarcoma protuberans (DFSP) is a soft tissue sarcoma that can be locally aggressive, but has a low risk of metastasis. DFSP is considered rare as it causes 0.1% of all malignancies and about 1% of all soft tissue sarcomas.1,2 In the United States, DFSP has an incidence between 0.8 and 4.5 cases per million individuals annually.3 While DFSP can develop at any age, it is most often seen around the third and fourth decades.4 African Americans have a higher incidence of DFSP compared to Caucasians.5 In a study of 2885 cases over 30 years, DFSP was found in twice as many African Americans than Caucasians, with relatively equal incidence in females and males.6 DFSP tumors can occur in various locations on the body, but it has a higher propensity to develop on the chest and trunk.7 Multiple subtypes of DFSP have been identified including the following: DFSP with areas of giant cell fibroblastomas, DFSP with fibromatosarcoma areas, myxoid, granular, atrophic and others.8 The rare pigmented variant of DFSP is known as the Bednar tumor. Bednar tumors predominantly occur in African Americans and account for 15-50% of all DFSP.9 The tumor displays spindle cells arranged in a storiform pattern with scattered melanin-bearing dendritic cells causing the tumor to appear blue or black.8

The appearance of a DFSP tumor can vary given the slow growth for an extended period before entering a rapid growth phase. In the early stages, DFSP typically presents as an asymptomatic, non-painful red or brown plaque with a firm texture that is generally fixed to the skin, but not underlying tissue.9 Clinical variabilities do exist in the pretumor/pretumor stage. DFSP develops during childhood, it can be morphoea-like with a white or brown indurated plaque resembling morphoea, nevusseous basal cell carcinomas, scar or dermatofibroma plaque.10 DFSP is often associated with a translocation on the 17q22 ring chromosome.11 The distal long arm of chromosome 17 houses the 17q25 translocation breakpoint, potentially causing an extra copy number and/or poor regulation of an oncogene, a possible contributory to the neoplastic initiation or progression of DFSP.12

Figure 1. A 2.5 x 1.4 cm black well-circumscribed black nodule.

Figure 2. 2x magnification of the asymmetrical spindle-cell neoplasm extending through the dermis into the septa and lobules of the subcutis with intermittent deposits of melanin. (H&E)

Conclusion
In conclusion, although DFSP is considered a rare malignancy, clinicians should be aware of its variants and be knowledgeable of its treatments and workup. Even more rare is a Bednar variant of DFSP, as seen in our patient. When properly treated and with plenty of patient counseling, the prognosis of DFSP is quite good.

References