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# Perianal Pigmented Variant of Dermatofibrosarcoma Protuberans: A Case Report

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## INTRODUCTION

- Dermatofibrosarcoma protuberans (DFSP) is a soft tissue sarcoma that accounts for 0.1% of cancers
  - Incidence of 4.1 cases per million person-years in the US
  - Incidence greater than 5.0 cases per million person-years in Detroit<sup>1</sup>
- Most commonly occurs on the trunk and extremities
  - Less than 1% occur in the genital region<sup>1</sup>
  - Only 3 perianal DFSP cases reported in the literature<sup>2-4</sup>
- Histopathology characteristics include storiform spindle cells with minimal atypia, CD34 positive, factor XIIIa and S100 negative, and can be associated with the presence of COL1A1-PDGFR $\beta$  fusion gene<sup>5</sup>
- Bednar or pigmented type occurs in less than 5% of DFSP and is more common in African Americans<sup>1,5</sup>
- Locally aggressive with historically high local recurrence rates up to 50%<sup>6,9</sup>
- Current NCCN guidelines recommend 2 to 4 cm margins to the investing fascia<sup>6</sup>

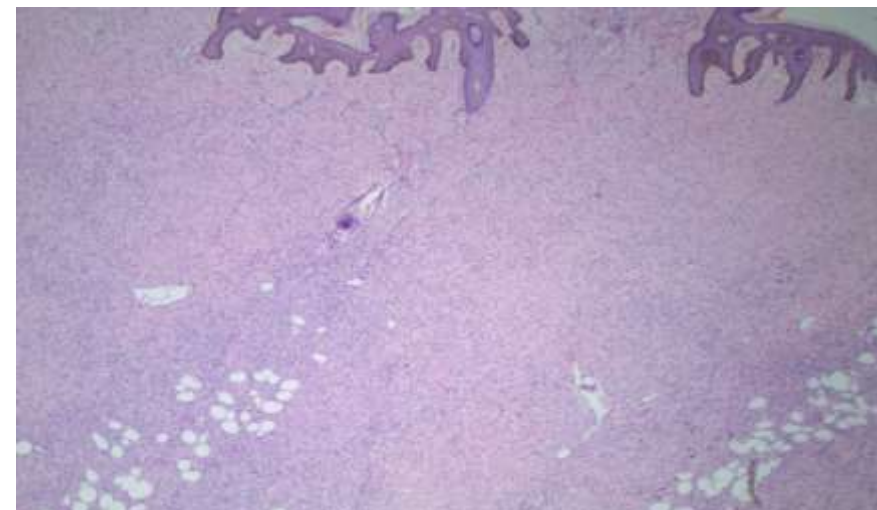
## CASE PRESENTATION

- 44-year-old African American female presented with mildly pruritic, non-tender, non-draining left buttock lesion first noted 2 years before
  - Firm 1 cm hyperpigmented scar-like papule thought to be from prior furunculosis or epidermoid cyst
  - Elected for continued observation
- 1 year later, lesion reevaluated and found to have with increasing hyperpigmentation and size; patient referred for surgical evaluation
  - Single 2x2 cm indurated, non-tender, hyperpigmented perianal nodule at the 10 o'clock position was identified
  - Initial excisional biopsy unexpectedly diagnosed Bednar DFSP with histology of storiform spindle cells, CD34 positive, S100 negative, and dendritic melanin-containing cells
- Subsequently, patient underwent definitive wide local excision with primary closure
- However, circumferential margins were limited to 1 cm due to proximity to anal verge
- Follow-up MRI at one-year post-excision without evidence of recurrence
- Currently undergoing annual skin exams with primary care physician for surveillance

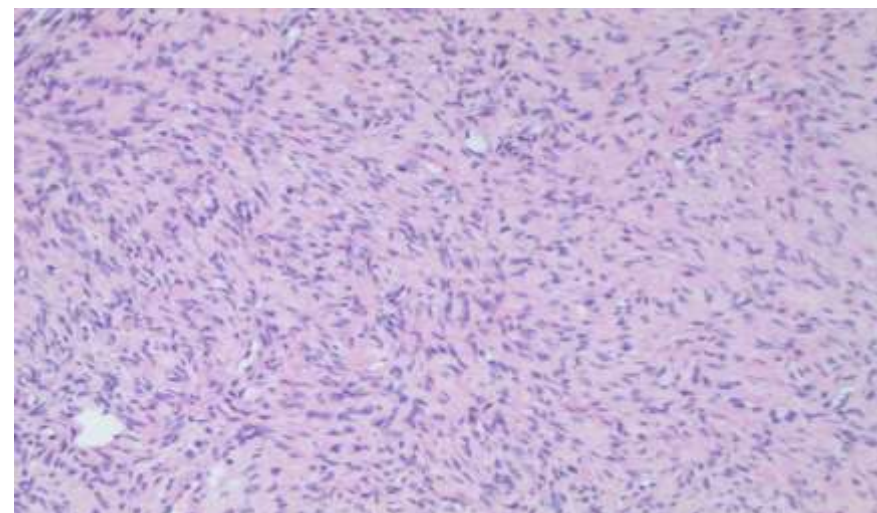
## FIGURES



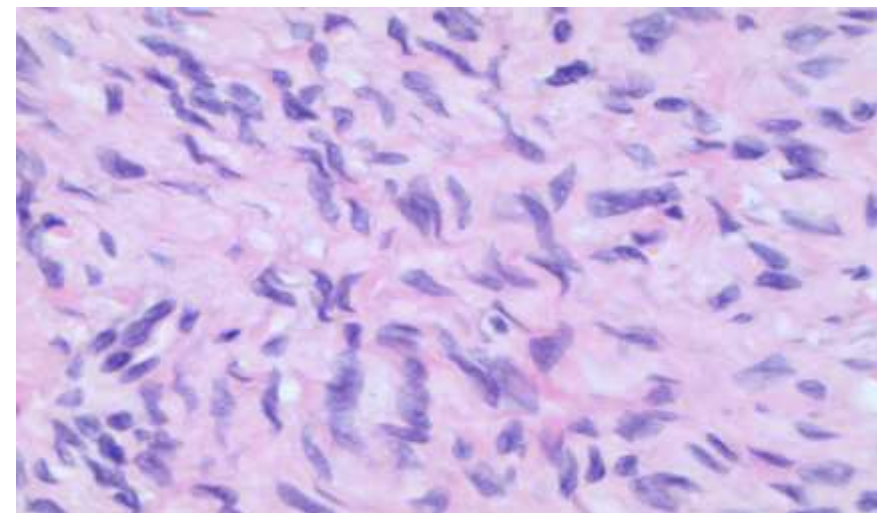
**Figure 1.** Indurated hyperpigmented nodule on initial surgical evaluation after 3 years of symptoms



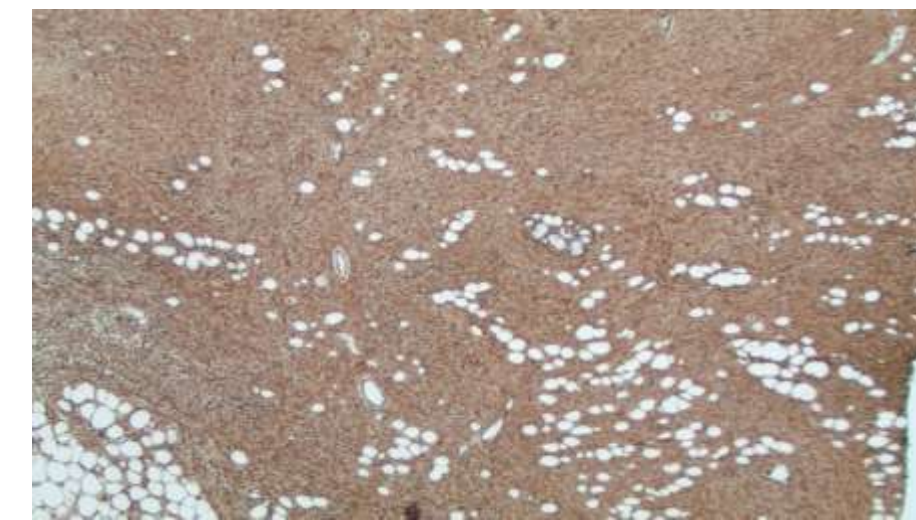
**Figure 2.** Hematoxylin and eosin, characteristic "honeycomb" pattern



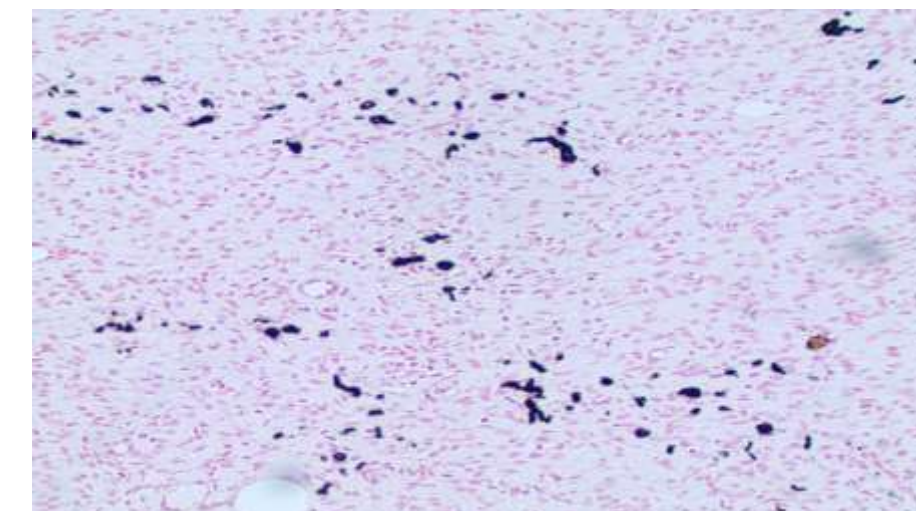
**Figure 3.** Hematoxylin and eosin, storiform pattern of neoplastic cells



**Figure 4.** Hematoxylin and eosin, uniform spindle cells without atypia



**Figure 5.** While nonspecific and seen in spindle cell tumors, CD34 immunohistochemical stain strongly and diffusely positive consistent with DFSP



**Figure 6.** Fontana-positive melanin pigment in dendritic cells consistent with Bednar variant that occurs in <5% of DFSP



**Figure 7.** 1 year post-operative, well healed incision with proximity to anal verge evident

## DISCUSSION

- DFSP is malignant fibroblastic tumor traditionally associated with high rates of local recurrence and rarely metastases
- Its indolent growth pattern can be associated with initial misdiagnosis or delayed diagnosis and treatment, as was the case here
- Treatment is surgical excision with meticulous histologic examination of the specimen to ensure adequate margins
  - DFSP infiltrates subcutaneous tissues through finger-like extensions complicating analysis of margins
  - Proximity to the anal verge limited attainable margins to 1 cm while avoiding incontinence
- Mohs micrographic surgery has been utilized to obtain negative margins with similar recurrence rates as compared to wide local excision with careful microscopic examination
  - Recent data suggests recurrence rates as low as 1% in both approaches<sup>9,10</sup>
- Reconstruction or tissue undermining should be avoided until negative histologic margins are confirmed
- Imatinib, a protein tyrosine kinase inhibitor, can be used for recurrent or unresectable DFSP with t(17;22) translocation<sup>6</sup>
- Radiation therapy has been utilized with 86-93% 10 year disease free survival and could be employed if complete resection is not feasible<sup>7,8</sup>

## CONCLUSIONS

- This is the first perianal Bednar DFSP case reported and one of few perianal DFSP cases found in the literature
- This case highlights the indolent course of DFSP delaying diagnosis as well as difficulties achieving recommended surgical margins while preserving continence

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