BRIEF ARTICLE

Dermatofibrosarcoma Protuberans Presenting as a Subcutaneous Cystic Nodule

Chris Logas, DO[1], Austin Dunn, DO[2], Danielle Lazarra, DO[1], Justin Rubin, DO [1], Eli Saleeby MD, Brad P. Glick, DO[1,3]

[1] Larkin Community Hospital – Palm Springs Campus, Dermatology Residency Program, Hialeah, FL
[2] Center for Clinical and Cosmetic Research - Aventura, FL
[3] Glick Skin Institute, Margate, FL

ABSTRACT

A 41-year-old African American male complains of a painful cyst located on his right shoulder and lasting 2 for years. The specimen was excised and stained positive for CD34 and negative for SOX-10, confirming the diagnosis of Dermatofibrosarcoma Protuberans (DFSP). The initial specimen had positive extension at both the deep and lateral margins. Complete removal of the tumor required 3 stages of MMS resulting in a 10 cm x 7.6 cm final defect, extending to muscle. As demonstrated in this case, DFSP has an erratic growth pattern and requires meticulous histopathological examination to ensure clear margins. MMS is the preferred method of treatment. Cases of DFSP have high rates of recurrence and require regular follow-up for 3-5 years after treatment.

INTRODUCTION

Dermatofibrosarcoma Protuberans (DFSP) is a rare soft tissue tumor that was first described by Derier and Ferrand in 1924[1]. It most often occurs on the trunk of young to middle aged patients and has an incidence of 4.1 per million adults accounting for 0.1% of all malignancies [2–4]. It often presents as a slow growing, indurated plaque and progresses into a violaceous to red-brown firm nodule fixed to the skin with occasional reddish blue to reddish-yellow surrounding discoloration. Tumors can measure from one centimeter upwards to several centimeters in diameter [2,5,6]. Although occurrence of DFSP is preceded by trauma in approximately 10% of cases, it is unclear if there is a direct correlation with development [4]. DFSP is considered a low-grade sarcoma and regarded as relatively benign with a low rate of metastasis (0.5%) and high 2-to-5-year survival rate (92-97%)[2,6]. While DFSP has an overall good prognosis, the rate of recurrence is high (20-50%), requiring lifelong surveillance of patients following diagnosis. Surgical excision is the most common treatment but often proves incomplete due to the infiltrative growth pattern and fingerlike projections characteristic of the tumor. Thus, Mohs micrographic surgery (MMS) is the preferred treatment to ensure a clear margin [2]. We report a clinical case of a patient diagnosed with DFSP and subsequently treated with MMS.

CASE REPORT

A 41-year-old African American male with a past medical history significant for pre-diabetes and hypertension presented to the

clinic with a complaint of a cyst located on his right shoulder for 2 years (Figure 1A).

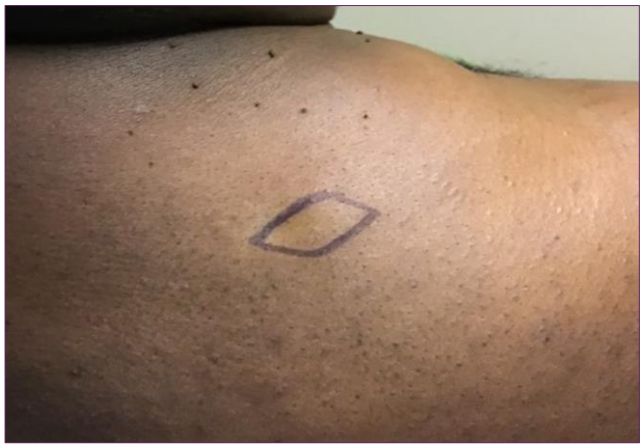

Figure 1. A) Initial presentation of a cystic nodule

The nodule had been asymptomatic, but recently increased in size and became painful. This patient denied any associated symptoms including fever, chills, drainage, or increased warmth to the touch of lesion. Physical exam revealed a large subcutaneous nodule about 2 cm in size, tender to palpation. The lesion was surgically excised a week later. Histologically, the specimen was a markedly mesenchymal neoplasm occupying the reticular dermis, as well as the subcutis with invasion into the fat in a honeycombing fashion (Figure 2).

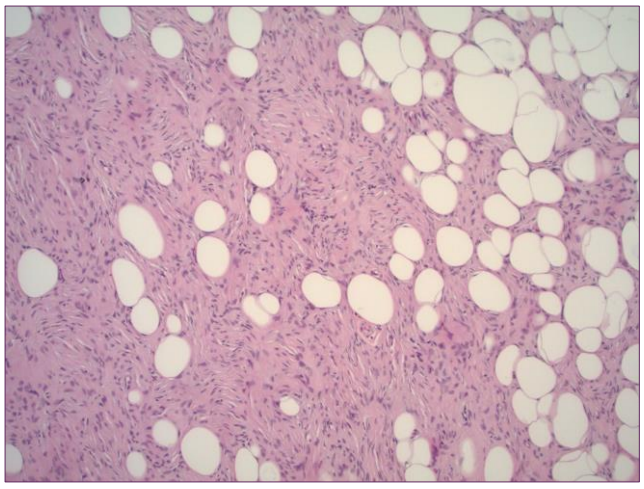

Figure 2. H&E X10 after initial resection of tumor

The neoplasm was extremely cellular with little intervening collagen. The cells were thin and filiform in shape, with slender, wavy nuclei, and in foci showed prominent whirling. The specimen stained positive for CD34 and negative for SOX-10, confirming the diagnosis of DFSP (Figure 3).

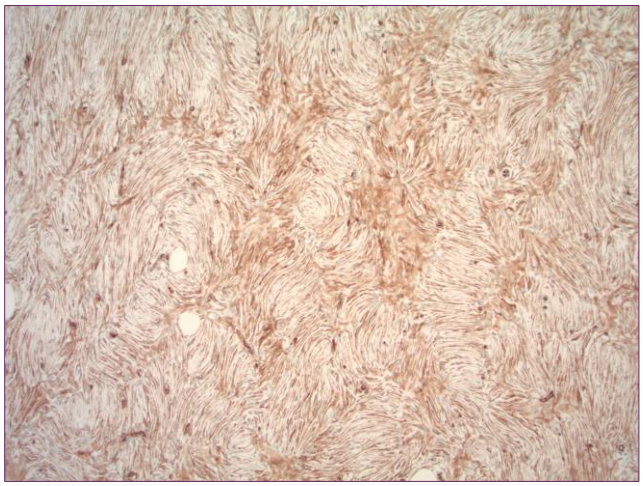

Figure 3. CD34 staining after initial resection of tumor

The initial specimen had positive extension at both the deep and lateral margins. Complete removal of the tumor required 3 stages of MMS resulting in a 10 cm x 7.6 cm final defect, extending to muscle, (figure 1B).

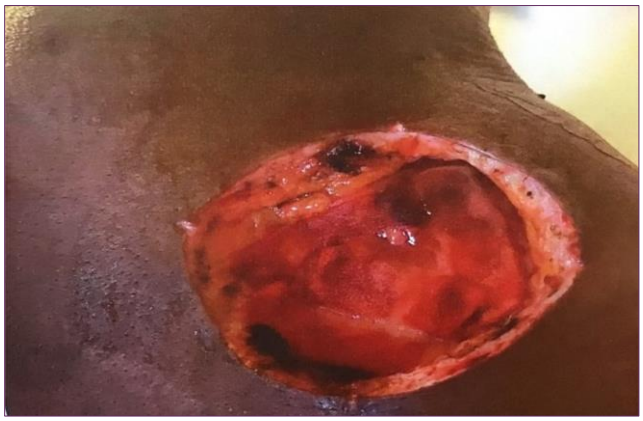

Figure 1. B) Surgical defect after 3 stages of MM

Histology of stages 1 and 2 exhibited a dense, cellular tumor proliferation composed of spindled fibroblasts intersecting collagen

bundles within the dermis and invading the subcutaneous fat. Perineural invasion was not noted. The patient was referred to plastic surgeon for repair. A skin graft was determined to provide the best cosmetic and functional result. Upon one month follow-up, the surgical defect and skin graft appeared well-healed. The patient underwent genetic, PET/CT imaging, and tumor marker testing which were all negative for any pathological abnormalities or metastatic disease.

DISCUSSION

As in this case, diagnosis of DFSP may be challenging. A punch biopsy containing epidermis, dermis, and subcutaneous fat can lead to an accurate diagnosis, but excisional biopsy is the preferred diagnostic method [1]. Due to the erratic growth pattern of DFSP and the need for meticulous histopathological examination to ensure clear margins, MMS is the preferred method of treatment[4]. Recurrence rate decreased by 6.2% when using MMS over WLE (7.3% compared to 1.1%)[4]. A disadvantage of MMS is that tumor cells can be confused with normal spindle cells of the dermis. CD34 staining can offer some assistance but is of variable use[4].

A unique genetic transformation in DFSP allows for systemic therapy with Tyrosine Kinase inhibitors. Approximately 90% of tumors are associated with the (17;22) translocation, which leads to formation of the COL1A1-PDGFB fusion protein[2]. Increased PDGFB expression leads to autocrine activation, tumor growth and development [4]. Imatinib targets the PDGF receptor, providing an alternative treatment option. In imatinib-resistant DFSP, other tyrosine kinase inhibitors such as sunitinib and sorafenib may be beneficial [2]. Adjuvant radiotherapy can be used if an excised tumor has positive margins and re-excision is not possible [1,2].

CONCLUSION

Due to the high rate of recurrence of DFSP, evaluation for metastasis and close, regular follow-up is critical when developing appropriate treatment plans. Patients are recommended to have 3-to-6-month interval follow-ups for 3-5 years after initial diagnosis [4].

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Corresponding Author:
Austin Dunn, DO
Center for Clinical and Cosmetic Research
Aventura, FL
2925 Aventura Blvd., Suite 205
Aventura, Florida 33180
Phone: (765) 808-3631
Email: a.dunn@admcorp.com

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