

Dermatofibrosarcoma Protuberans of the Neck: A Case Report

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Abstract

Background: Dermatofibrosarcoma protuberans (DFSP) is rare soft tissue sarcoma but has a locally aggressive nature. Although most cases are of low grade and have a rare metastasis rate, all DFSP variants have a tendency to show local recurrence. Wide excision with negative margins is the treatment of choice. **Aim:** To report a case of DFSP who presented with an asymptomatic slow growing tumor similar to etiologies such as hypertrophic scars or other benign soft tissue tumors. **Case Presentation:** A 68-year-old male presented with a large soft tumor located at the left posterior neck. Local excision was done under the preoperative impression of a benign tumor such as lipoma or sebaceous cyst. However the diagnosis of DFSP was made upon histological examination and the patient underwent another surgery to achieve free resection margins under general anesthesia as well as adjuvant radiotherapy. **Conclusion:** DFSP is a malignant tumor that is diagnosed histopathologically. Due to the low incidence rate, slow-growing nature, and non-alarming initial presentation features, diagnostic delay or even misdiagnosis is not uncommon.

Keywords

Dermatofibrosarcoma Protuberans, Soft Tissue Sarcoma, Surgery

1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is an uncommon locally aggressive cutaneous soft tissue sarcoma of the dermis layer of the skin [1] with an incidence of 0.8 - 4.5 cases per million per year [2] [3]. Approximately 85 to 90 percent of DFSPs are low grade. Despite its rare metastasis rate (<5%), all DFSP variants have a tendency to show local recurrence [4] [5]. Most commonly occurring in adult patients in their thirties, DFSP accounts for approximately 1% - 6%

of soft tissue sarcoma cancers [5] [6] [7]. More than 90% of DFSP tumors have rearrangement of chromosome 17 and 22 that fuses collagen gene COL1A1 with platelet-derived growth factor (PDGF), leading to transcriptional up-regulation of PDGF and thus established a self-stimulatory growth signal [7]. Immunoreactivity for CD34 in DFSP was described in 1993 and remains to be the main immunohistochemical marker for diagnosing DFSP [7]. Due to its slow growing nature similar to nodules that appear as hypertrophic scars or benign soft tissue tumors without any definite symptoms, it is difficult to diagnose pre-operatively. The preferred initial treatment for a localized dermatofibrosarcoma protuberans (DFSP) is resection with pathologically negative margins. However, extensive excision may not be feasible due to either anatomic limitation or cosmetic concerns, especially if found in the head and neck region. A case requiring an additional surgery and adjuvant radiotherapy, due to difficulty of an accurate pre-operative diagnosis and tumor location, is reported here.

2. Case Report

A 68-year-old Taiwanese man visited our clinic due to noticing a left posterior neck mass for 6 months. The patient reported progressive growth of the tumor in size during the preceding six months. The patient denied any recent weight loss, fever, night sweats or chills. On physical examination, the mass measured about 2.0×2.0 cm over the patient's left posterior neck, and it was firm, painless, with no sign of localized heat or erythematous change, and relatively movable. The patient's neck was supple, with no palpable cervical lymphadenopathies. The patient denied past history of malignancy. A neck sonography exam was performed, and a well-defined, heterogeneous subcutaneous tumor was noted, measuring 1.58×0.92 cm, with posterior enhancement (**Figure 1**). Under the impression of a posterior neck lipoma or sebaceous cyst, the patient underwent excisional biopsy under local anesthesia on an outpatient basis. Gross tumor was completely removed, but an infiltrative nature was noted around the border without a well-defined margin. Upon histological examination, infiltrating and hypercellular tumor composed of interlacing fascicles of hyperchromatic atypical spindle cells arranged in vague storiform fashion was described (**Figure 2**), as well as abnormal mitotic figures average 4 per 10 HPFs noted. Special stain showed tumor cells are immunoreactive against antibody to CD34 (**Figure 3**). Based on the histological and immunohistochemical findings, the diagnosis of dermatofibrosarcoma was made. The patient underwent an additional wide excision under general anesthesia. Due to lack of gross tumor, the wide excision was focused to encompass the whole previous surgical site with resection margins checked intra-operatively via frozen section pathologic review. Although the histopathology report revealed free margins and only few atypical spindle cells present in the specimen, the surgeon felt that resection of an adequate margin could not be achieved, especially the deep margin, due to the location of the tumor being at the neck. The patient underwent adjuvant local radiotherapy treatment with a total radiation dose of 66 Gy in 33 fractions. The patient cur-

rently has no signs of local recurrence 12 months after surgery, and is under regular follow-up with sonography exam (**Figure 4**).

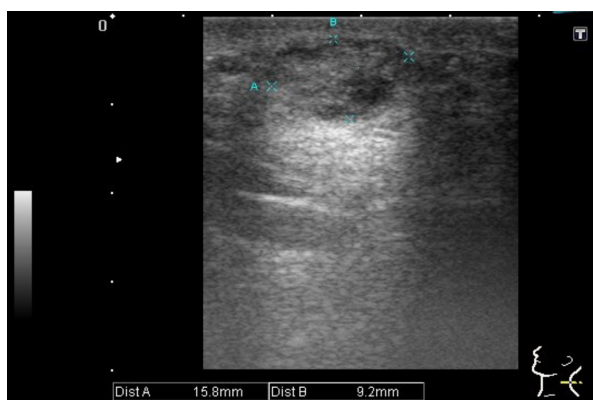


Figure 1. Pre-operative sonography of the posterior neck lesion showing a relatively well-defined heterogeneous nodular lesion with acoustic shadow.

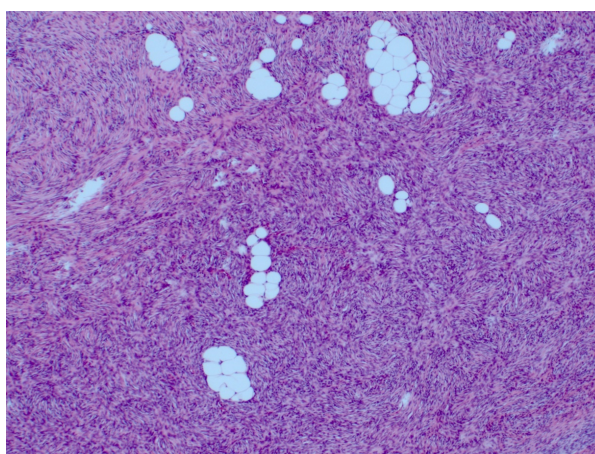


Figure 2. Histological examination: infiltrating and hypercellular tumor composed of interlacing fascicles of hyperchromatic atypical spindle cells arranged in vague storiform fashion.

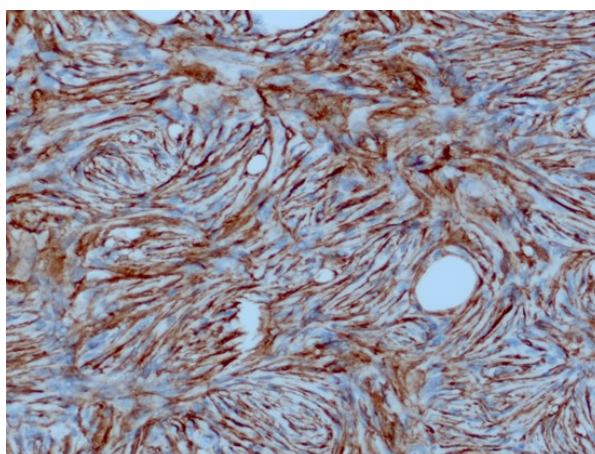


Figure 3. Special staining showed the tumor is immunoreactive against antibody to CD34.

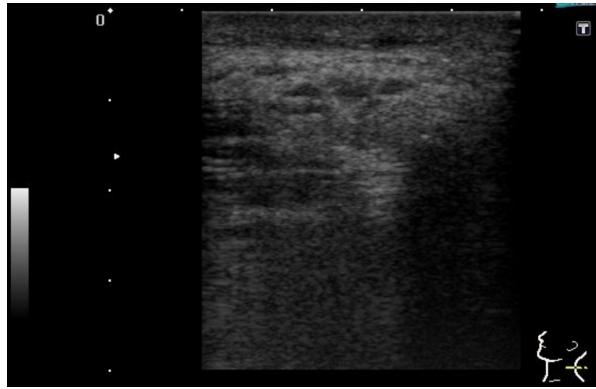


Figure 4. Post-operative sonography examination done 12 months after surgery showing no tumor recurrence.

3. Discussion

In the early stages, it is difficult to differentiate DFSP from other benign subcutaneous lesions such as lipomas, epidermal cysts, keloids, or dermatofibroma without undergoing surgical excision and histology examination. DFSP often presents as a minor skin firm area. It commonly resembles a bruise or pimple, and is a slow growing tumor. The most common location of DFSP is the trunk (42% - 72%) followed by proximal extremities (20% - 30%), with head and neck region being the least (10% - 16%) [8].

With sonography, DFSPs have been found to be around (67%) or ovoid (33%) subcutaneous lesion that are mostly well defined (89%) [9]. Appearance of DFSP usually involves a mildly lobulated border and a heterogeneously hypoechoic matrix. Posterior enhancement can also usually be noted [9]. Vascularity of DFSP, which is a marker of malignancy, varies as well. Since lipomas may also present with similar features, a distinction is not always possible. Hence, an initial preoperative impression of a benign lesion such as sebaceous cyst or lipoma was made for our case, which led to additional surgery and adjuvant treatment. Several authors have reported benefits of utilizing magnetic resonance imaging (MRI) preoperatively to better evaluate tumor extent [10]. In cases of large tumors (>4 cm in diameter), MRI is able to aid clinicians in distinguishing the relationship of the tumor with surrounding structures [11].

Definite diagnosis of DFSP requires histopathological examination. In order to ensure adequate tissue for examination, biopsy should be performed with a core-needle or surgical incision [12]. Microscopically, DFSP infiltrates the dermis diffusely, while often sparing the epidermis and skin. Kim *et al.* described seven histological DFSP subtypes, with “classic” DFSP comprising about 90% [13]. The “classic” subtype of DFSP has histological characteristics of monomorphic fusiform cells (spindle cells) and a large elongated nucleus and a low mitotic index. These spindle cells are irregularly organized with a storiform arrangement [13]. Due to its tendency to invade surrounding tissue from a central lesion, DFSP has projections that show a honeycomb (30%) or multilayered (70%) subcutaneous pattern [14] [15]. DFSP typically shows positive staining for vi-

mentin, CD34, apolipoprotein D, and nestin on immunohistochemistry [16].

DFSP is a low-grade malignancy, but has been reported a local recurrence rate up to 60% that may be attributed to incomplete excision due to poor circumscription and irregular boundaries [17]. Wide excision with 2 - 4 cm safe margin remains the cornerstone of the treatment of DFSP [12] [18] [19]. Although a sufficient excision margin is the key to reduce the local recurrence rate, it is often limited by the anatomical site, and the lesion typically infiltrates well beyond its grossly visible margin into the surrounding tissue. The use of Mohs micrographic surgery with incremental excision until normal tissue is obtained, as documented by repeat frozen sections, has been favored as the treatment of choice by some authors [20] [21].

Given that metastases to lymph nodes are extremely rare, there is no role for prophylactic regional node dissection. Radiotherapy (RT) is a noninvasive adjuvant treatment that can improve local control in patients, especially those having close or positive margins and re-excision not feasible. Several studies have suggested post-operative adjuvant RT reduced recurrence rate. A retrospective study of 184 patients reported a significant 5-year disease-free survival improvement in patients receiving adjuvant RT compared with patients that did not (88.1% vs 56.2%, $P = 0.044$) [22]. In our case, re-excision surgery was done after the diagnosis was made in an effort to achieve free and adequate resection margins. Although free surgical margin was reported, a wide safe margin was not feasible due to the location of the tumor, and adjuvant RT was utilized to improve local control. For DFSP patients with inoperable or metastatic diseases, Imatinib therapy is currently the standard of care. This targeted therapy may also potentially facilitate resection or decrease possible disfigurement when used in a neoadjuvant fashion [23].

4. Conclusion

DFSP is a rare locally aggressive malignant soft tissue sarcoma that originates from the dermis layer of the skin. Diagnostic delay or even misdiagnosis is not uncommon due to its indolent nature. DFSP is diagnosed by histopathology examination; therefore a biopsy or at least a core-needle biopsy is required in order to ensure adequate tissue specimen is obtained. Wide excision with negative margins is the treatment of choice. For patients with close or positive surgical margins, RT is a noninvasive adjuvant treatment that improves local control.

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Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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