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Late Local Recurrence of Dermatofibrosarcoma Protuberans of the Scapular Region: A Case Report and Review of the Literature

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Abstract | **Case Report**

Background : Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing cutaneous sarcoma with a high risk of local recurrence if incompletely excised. Although its metastatic potential is low, late local recurrences may occur, underscoring the need for long-term surveillance. ***Case presentation :*** We report the case of an 81-year-old woman who developed a late local recurrence of DFSP in the left scapular region, 18 years after initial excision. Clinical examination revealed a polylobulated cutaneous mass without lymphadenopathy. Histopathology demonstrated a storiform spindle-cell proliferation infiltrating the dermis and subcutaneous tissue, with diffuse CD34 positivity. Surgical excision revealed tumor persistence at the inferior lateral margin. The patient was referred for re-excision and reconstructive surgery. ***Discussion :*** This case illustrates the potential for very late recurrence of DFSP, even decades after primary treatment. Complete surgical excision with wide margins or Mohs micrographic surgery remains the cornerstone of management. Radiotherapy may be considered in unresectable cases or when re-excision is not feasible. Targeted therapy with imatinib represents an option for advanced or metastatic disease harboring the COL1A1–PDGFB fusion gene. ***Conclusion :*** DFSP requires lifelong follow-up due to its potential for late recurrence. Achieving negative surgical margins is critical to prevent relapse, and a multidisciplinary approach is essential in recurrent or complex cases.

Keywords: Dermatofibrosarcoma protuberans, DFSP, Sarcoma, Local recurrence.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP), first described by Darier and Ferrand in 1924 [1], is a rare, slow-growing cutaneous sarcoma accounting for less than 0.1% of all malignant neoplasms and approximately 1% of soft tissue sarcomas [2]. DFSP typically arises in the trunk and proximal extremities of young to middle-aged adults, but it can occur at any age [3].

Histologically, DFSP is characterized by storiform spindle-cell proliferation with strong and diffuse CD34 expression. The hallmark molecular alteration is the COL1A1–PDGFB fusion gene resulting from t(17;22) (q22;q13), which promotes constitutive activation of PDGFB signaling [4].

The mainstay of treatment is wide local excision (WLE) or Mohs micrographic surgery, as positive margins are strongly associated with local recurrence [5]. Although DFSP has a low metastatic potential, its recurrence rate can exceed 50% in cases of inadequate surgical margins [6].

We report the case of an elderly patient with a late local recurrence of DFSP of the scapular region, 18 years after initial excision, underscoring the need for long-term follow-up and meticulous margin control.

CASE PRESENTATION

An 81-year-old woman, with no significant medical history, was first diagnosed in 2007 with a cutaneous nodule in the left scapular region. The lesion was excised, and histopathology confirmed a dermatofibrosarcoma protuberans. The patient remained disease-free until early 2025, when she developed a progressively enlarging, non-ulcerated cutaneous lesion over the same site.

On admission, clinical examination revealed a polylobulated mass in the left scapular region, and no palpable lymphadenopathy was detected.

A biposy of the recurrent lesion was performed, and histopathological and immunohistochemical analyses confirmed the diagnosis of dermatofibrosarcoma protuberans.

The patient underwent surgical excision.

Histopathological examination of the surgical specimen revealed a mesenchymal spindle-cell proliferation with storiform architecture, infiltrating the dermis and hypodermis, isolating islands of mature adipocytes. Nuclear atypia was minimal, and mitotic activity was low. Immunohistochemistry demonstrated strong CD34 positivity (Figure 1).

Margins were microscopically positive at the inferior lateral limit, with other margins ranging from 0.8 cm to 1.5 cm of healthy tissue.

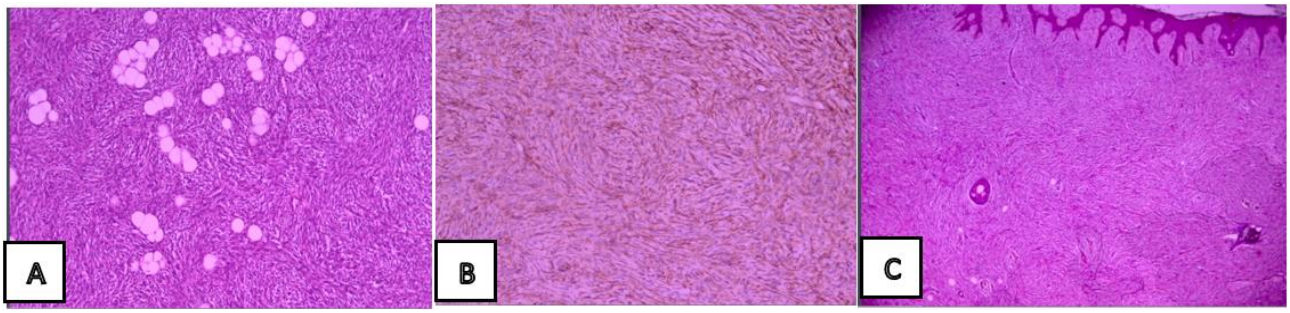

Figure 1:
A. Hematoxylin and eosin (H&E) staining, low magnification: dermal proliferation of spindle-shaped cells. The overlying epidermis appears papillomatous and non-neoplastic.
B. High magnification: spindle cells infiltrating and entrapping the adipose tissue.
C. CD34 immunohistochemistry: spindle cells showing diffuse cytoplasmic positivity.

The patient was referred for surgical re-excision and cutaneous coverage by the plastic surgery team.

DISCUSSION

DFSP is a rare, slow-growing sarcoma of the skin and subcutaneous tissue, with locally aggressive behavior and a high risk of recurrence if incompletely excised [2,5,6]. The recurrence rate varies from 20% to 50%, depending on surgical margins [7].

Our patient illustrates a very late local recurrence, 18 years after initial excision. Such delayed recurrences have been reported, confirming the importance of lifelong clinical and radiological surveillance [8].

The main prognostic factor remains the status of surgical margins [5,6]. Wide local excision with margins of at least 2–3 cm or Mohs micrographic surgery significantly reduces recurrence rates [9]. In our case, the persistence of tumor at the lateral inferior margin highlights the technical challenges of complete excision in the scapular region.

Radiotherapy may be considered in cases of unresectable tumors or positive margins when re-excision is not feasible [10,11]. Targeted therapy with imatinib mesylate, a PDGFRB inhibitor, has shown efficacy in unresectable, recurrent, or metastatic DFSP harboring the COL1A1–PDGFB fusion gene [12].

This case underlines three key aspects:

1. DFSP can recur many years after primary excision, necessitating lifelong follow-up.
2. Achieving clear surgical margins is crucial to prevent recurrence.
3. Multidisciplinary management (surgery, radiotherapy, targeted therapy) is essential in recurrent or unresectable cases.

CONCLUSION

Dermatofibrosarcoma protuberans is a rare cutaneous sarcoma with a high propensity for local recurrence. This case emphasizes the importance of achieving wide excision margins and maintaining long-term follow-up, as late recurrences may occur even decades after initial treatment.

REFERENCES

1. Darier J, Ferrand J. Dermatofibrosarcome progressif et récidivant ou fibrosarcome de la peau. Annales de Dermatologie et de Syphiligraphie. 1924 ; 5:545–562.
2. Llombart B, Serra-Guillén C, Monteagudo C, *et al.,* Dermatofibrosarcoma protuberans: a comprehensive review and update on diagnosis and management. Semin Diagn Pathol. 2013; 30(1):13–28.
3. McKee PH, Calonje E, Granter SR. Pathology of the skin. 3rd ed. Elsevier Mosby; 2005.

4. Simon MP, Navarro M, Roux D, *et al.,* Deregulation of the platelet-derived growth factor B-chain gene via fusion with collagen gene COL1A1 in dermatofibrosarcoma protuberans and giant-cell fibroblastoma. Nat Genet. 1997;15(1):95–98.
5. Gloster HM. Dermatofibrosarcoma protuberans. J Am Acad Dermatol. 1996;35(3):355–374.
6. Stivala A, Lombardo GA, Pompili G, *et al.,* Dermatofibrosarcoma protuberans: wide local excision vs. Mohs micrographic surgery. Cancer Treat Rev. 2012;38(5):423–431.
7. Bowne WB, Antonescu CR, Leung DH, *et al.,* Dermatofibrosarcoma protuberans: a clinicopathologic analysis of patients treated and followed at a single institution. Cancer. 2000;88(12):2711–2720.
8. Snow SN, Gordon EM, Larson PO, *et al.,* Dermatofibrosarcoma protuberans: a report on 29 patients treated by Mohs surgery with long-term follow-up and review of the literature. Cancer. 2004;101(1):28–38.
9. Farma JM, Ammori JB, Zager JS, *et al.,* Dermatofibrosarcoma protuberans: how wide should we resect? Ann Surg Oncol. 2010;17(8):2112–2118.
10. Judson I, *et al., Guidelines for the management of soft tissue sarcomas.* Sarcoma. 2016; 2016:3586185.
11. Ugurel S, *et al., Local treatment modalities in dermatofibrosarcoma protuberans.* Cancers (Basel). 2025; 17(3):534.
12. Rutkowski P, Debiec-Rychter M, Nowecki ZI, *et al.,* Treatment of advanced dermatofibrosarcoma protuberans with imatinib mesylate: results from a multicenter study. J Clin Oncol. 2010;28(10):1772–1779.