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Case Report

An evolving supraclavicular plaque: diagnostic challenges in dermatofibrosarcoma protuberans with atrophic and myxoid features

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Abstract

Dermatofibrosarcoma protuberans is an uncommon locally aggressive mesenchymal neoplasm that classically presents with a proliferation of monomorphic spindled cells with thin nuclei and scant cytoplasm. We report a man in his twenties who presented for an unrelated skin concern and was incidentally noted to have a large, atrophic scar-like depression of the left supraclavicular neck and shoulder. The plaque was photographed, and the patient was instructed to follow up in one year, and careful photo comparison revealed the lesion had subtly enlarged and darkened in color. Accordingly, a punch biopsy revealed features of myxoid dermatofibrosarcoma protuberans, a dermal proliferation of spindled cells with mucinous degeneration and more cellular areas with extensive nuclear palisading resembling the Verocay bodies of a neurofibroma. Clinicians should be aware of the broad clinicopathologic spectrum of DFSP to ensure timely diagnosis and effective treatment. Myxoid dermatofibrosarcoma protuberans is a rare and clinically challenging diagnosis, as the mucinous areas can impart a bluish hue. Dermatofibrosarcoma protuberans can also be atrophic, with the loss of the normal dermal thickness corresponding to a clinical appearance easily mistaken for anetoderma or atrophic scar. Palisaded nuclei resembling the Verocay bodies of schwannoma are sometimes seen and can further obfuscate the diagnosis.

Introduction

Dermatofibrosarcoma protuberans (DFSP) is an uncommon, locally aggressive tumor, and recurrence is a frequent complication after excision. Mohs surgery has been shown to significantly enhance rate of clearance; however, effective Mohs surgery relies on recognition of the full histopathologic spectrum of DFSP to ensure margin control. Herein, we describe a challenging case of DFSP containing multiple unusual histologic subtypes intercalating with areas of conventional DFSP that may be easily confused. Overall, clinicians should be aware of the broad clinicopathologic spectrum of DFSP to promote timely diagnosis and effective treatment.

Case Synopsis

An otherwise healthy man in his twenties was noted to have an atrophic scar-like depression of the left supraclavicular neck and shoulder ([Figure 1](#)). Upon initial evaluation, the patient reported that this lesion was unchanged for over ten years and attributed it to a previously ruptured cyst. At one year follow-up, photo comparison revealed the plaque had enlarged and darkened in color ([Figure 2](#)). Physical examination revealed a 5x3cm atrophic scar-like depression studded with bluish translucent papules. A punch biopsy was obtained for histopathological analysis.

The punch biopsy revealed a dense proliferation of spindle cells in a storiform configuration in the more cellular upper part of the lesion, with extensive myxoid areas to the bluish papules ([Figure 3A-3B](#)). In addition, there were focal areas of nuclear palisading resembling the Verocay bodies of a schwannoma ([Figure 3C](#)). No additional stains were performed. A diagnosis of myxoid dermatofibrosarcoma protuberans was rendered and the patient was referred to Mohs micrographic surgery.

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Figure 1. *Myxoid dermatofibrosarcoma protuberans:* clinical image at initial visit.



Figure 2. *Myxoid dermatofibrosarcoma protuberans:* clinical image at follow-up visit of 5x3cm atrophic scar-like depression studded with bluish translucent papules.

Additionally, preoperative MRI of the face and neck revealed that the tumor was superficial without adenopathy. MRI brain revealed no evidence of brain involvement or metastasis. A debulking prior to Mohs surgery showed giant cells in the myxoid areas, similar to those seen in giant cell fibroblastoma, a variant of DFSP (**Figure 3D**). The final Mohs defect measured 12.5x7cm and was repaired with a split thickness skin graft. One month after excision, the patient's wound was healing appropriately without evidence of infection or tumor recurrence.

Case Discussion

Dermatofibrosarcoma protuberans (DFSP) is an uncommon locally aggressive mesenchymal neoplasm that classically presents with a proliferation of monomorphic spindled cells with thin nuclei and scant cytoplasm. In the plaque stage, these cells are oriented parallel to the epidermal surface. In nodules, they assume a storiform pattern.¹ The vast majority of DFSP, approximately 90% of

cases, are thought to be triggered by a translocation between chromosomes 17 and 22.² This t(17;22) translocation allows fusion between the *COL1A1* and *PDGFB* genes (and rarely *PDGFD*), the first of which leads to excessive expression of exon 2 of *PDGFB* under the control of *COL1A1* promoter.² Given the largely monogenic pathogenesis of DFSP, it is not surprising that most DFSP initially share characteristic clinical and histologic features (ie, diffuse infiltration of fat in a lace-like or storiform pattern by fairly uniform, spindled cells) and indeed such "conventional" features of DFSP were well-demonstrated in this case. Furthermore, risk factors for the development of DFSP include scar formation, such as in the setting of surgical procedures or tattoo.³ Patel et al hypothesize that chronic inflammation may drive malignant transformation of scars in DFSP. In our case, there was no history of surgical procedure or tattoo to the affected area. However, this patient's reported history of cyst rupture suggests another mechanism of cutaneous injury, inflammation, and scar formation that may pose a risk of DFSP development.

As with many DFSPs, this tumor had grown without intervention for many years before the initial biopsy. Tumors that have been present for longer may be more likely to accrue additional mutations and have foci with unusual features such as myxoid degeneration and Verocay-like nuclear palisading.⁴ These seemingly unrelated histologic findings may delay diagnosis and consequently delay treatment for this locally aggressive neoplasm.⁵ Other histologic subtypes not present in this case but of clinical importance include Bednar tumor (pigmented DFSP), which can be mistaken for blue nevus or hemosiderotic dermatofibroma, and fibrosarcomatous DFSP, which displays a hypercellular 'herringbone' pattern, named for the V-shaped arrangement of cells. Fibrosarcomatous DFSP is thought to be more aggressive than other types of DFSP and some clinicians recommend obtaining imaging when this histologic pattern is seen.¹ Metastases have been reported in fibrosarcomatous DFSP in up to 18% of cases.⁶ In our patient, imaging, including MRI of the face, neck and brain, was obtained to assess for depth of infiltration given its high-risk location. The tumor presented with superficial depth of involvement without evidence of lymph node invasion. The Mohs surgeon obtained clear margins, and in light of these reassuring findings, no additional imaging was performed.

In this case, clinical appearance lends to a broad differential diagnosis, including atrophic dermatofibroma, plaque morphea, and scrofuloderma. Given the initial appearance of the lesion with features of a scar-like depression, atrophic dermatofibroma was a primary consideration. Although the loss of collagen can be appreciated on histology, the additional features of spindled cells with mucinous degeneration and extensive nuclear palisading differentiate atrophic dermatofibroma from our case.⁷ ⁸ The clinical presentation may also be mistaken for atrophic plaque morphea, which is characterized by asymmetric indurated plaques distributed along the trunk,

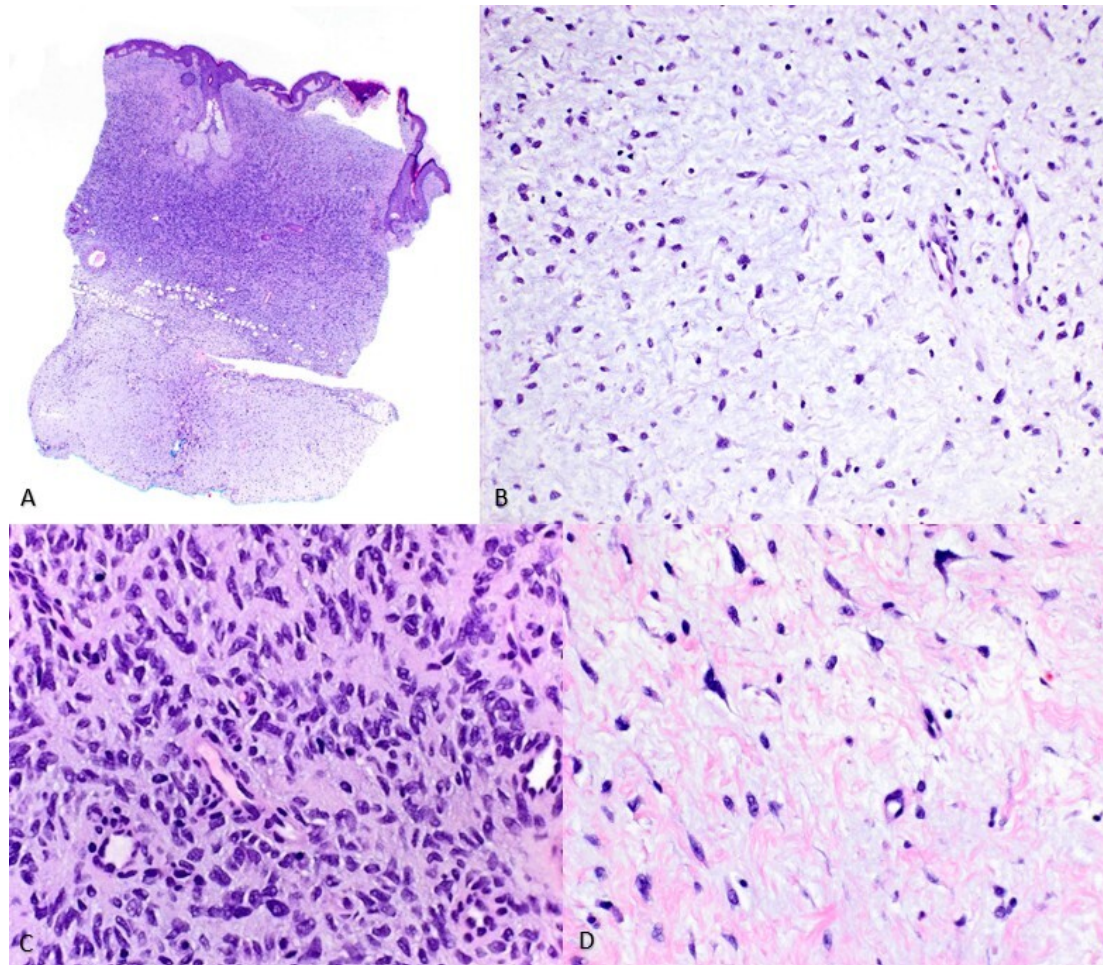


Figure 3. **A)** Low power magnification of hematoxylin and eosin-stained specimen from the debulking specimen obtained prior to Mohs micrographic surgery of featuring a residual spindle cell neoplasm that extends into the subcutis, with both hypercellular and pauci-cellular areas, with a poorly circumscribed configuration. **B)** High power magnification from the debulking specimen featuring a moderately cellular area with myxoid stroma and spindled cells with small nuclei. A few multinucleated cells are present. **C)** High power magnification from the initial punch biopsy, featuring small, irregularly shaped nuclei that form parallel arrays simulating Verocay bodies. **D)** A myxoid area from the debulking done prior to Mohs surgery, with giant cells resembling those of giant cell fibroblastoma.

neck, and back.⁹ Early plaques of morphea are often erythematous and become white and firmer over time.⁹ Over the course of the disease, these plaques may soften again, and the skin may become atrophic, hypo- or hyperpigmented.¹⁰ Histological findings of atrophic plaques in morphea include thickened collagen bundles with loss of the artifactual clefts normally found in between them and a dearth of blood vessels, which were not observed in this case.⁹ Scrofuloderma, a form of cutaneous tuberculosis resulting from contiguous spread of a tuberculous foci, could also be considered as an entity in the differential diagnosis.¹¹ These lesions commonly involve the neck or groin and initially present as painless, firm subcutaneous nodules. Over time, these nodules may gradually enlarge into ulcerated plaques and form sinus tracts, which may retract and become atrophic upon healing.¹² On histology, a pattern of caseating granulomatous inflammation is observed in the lower dermis, which were not observed in this case.¹²

Among the numerous variants of DFSP, myxoid DFSP remains a rare variant of DFSP, with limited case studies largely informing our current understanding. In our case of myxoid DFSP, this lesion developed on the supraclavicular fossa at the site of a reported prior scar, clinically evolving after a long period of quiescence. It was successfully removed with Mohs surgical excision without evidence of invasion. In the available literature, myxoid DFSP typically presents as a slow-growing firm mass.⁵ It most commonly involves extremities, followed by the head and neck, including the supraclavicular fossa.⁵ Although trauma and scar have been reported as a possible predisposing factor for the development of typical DFSP, it remains unclear if these factors precipitate myxoid DFSP.¹³

Conclusion

Overall, clinicians should be aware of the broad clinico-pathologic spectrum of DFSP to promote timely diagnosis

and effective treatment. Atrophic DFSP is a rare and clinically challenging diagnosis, as the loss of dermal thickness leads to a clinical appearance that is easily mistaken for atrophic scar, morphea, and lipoatrophy.¹⁴ Myxoid stroma in DFSP can cause a blue cast clinically, which can be confusing. Verocay bodies are sometimes seen microscopically and can further obfuscate the diagnosis. Mohs surgery has been shown to significantly enhance rate of clearance¹; however, effective Mohs surgery relies on

recognition of the full histopathologic spectrum of DFSP to ensure margin control and prevent recurrence.⁴

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Potential conflicts of interest

AH is a consultant to CSL Behring and an investigator for Priovant and AstraZeneca.

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