

Congenital Atrophic Dermatofibrosarcoma Protuberans: A Case Report and Review of the Literature

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Abstract: Dermatofibrosarcoma protuberans (DFSP) is a rare mesenchymal tumor of intermediate malignant potential. The neoplasm is locally aggressive with a high rate of recurrence. It typically presents in adults. Atrophic congenital DFSP is extremely rare. The few reported cases have presented as a morphea-like plaque that persists for years, before progressing into a nodular form. To our knowledge, congenital atrophic DFSP has been only reported fourteen times, and of those, only nine were confirmed by molecular studies. Herein we report a congenital case of atrophic DFSP, which initially presented as a bruise-like atrophic plaque on the dorsal forearm, initially mistaken for child abuse. The clinical appearance, histopathology, and molecular features of this rare form of DFSP are reviewed. Our case highlights the importance of early detection and adequate sampling of congenital DFSP; early treatment allows for treating small lesions without large, disfiguring, and potentially disabling excisions.

Keywords: DFSP; congenital; atrophic; dermatofibrosarcoma



Citation: Salem, I.; Bradley, K.; Mann, J.A.; Shin, J.H.; LeBoeuf, M.; Sriharan, A. Congenital Atrophic Dermatofibrosarcoma Protuberans: A Case Report and Review of the Literature. *Dermato* **2023**, *3*, 97–108. <https://doi.org/10.3390/dermato3020008>

Academic Editor: Chalid Assaf

Received: 11 January 2023

Revised: 7 April 2023

Accepted: 11 April 2023

Published: 16 April 2023



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1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is an uncommon soft tissue cancer with intermediate malignant potential, characterized by progressive local growth into muscle and fascia [1,2]. There is a tendency for recurrence, especially in the first three years post-excision [3]. Metastasis is extremely rare. Lesions with fibrosarcomatous transformation on histology and those with multiple recurrences are more likely to metastasize [3,4]. DFSP is typically a malignancy of adulthood. Recently, however, its prevalence in childhood was found to be more frequent than previously thought [5]. Juvenile DFSP is often misdiagnosed as more common pediatric diseases such as vascular anomalies, bruises, deep penetrating nevi or morphea [6]. The frequent initial misdiagnosis often results in delay in management, which can necessitate large excisions, complicated by significant morbidity and mortality [6]. Herein, we report a case of a ten-year-old female presenting with an atrophic bruise-like plaque on her left forearm, first noticed at birth. After excluding possible child abuse at the age of one, a preliminary clinical diagnosis of morphea was made. However, an initial punch biopsy revealed a fibrohistiocytic proliferation. A wedge biopsy was subsequently obtained to further assess the lesion. The morphology, expression of CD34, and lack of factor XIIIa or S100 expression, were indicative of DFSP. The detection of the COL1A1-PDGFB gene fusion, by two different orthogonal techniques, confirmed the diagnosis. Magnetic resonance imaging (MRI) revealed sparing of the muscle and fascia.

The patient underwent a complete local excision with a skin graft, without additional chemotherapy or radiation, as recommended by the sarcoma tumor board. This report aims to shed light on this rare entity and emphasizes the importance of maintaining a high index of suspicion of DFSP in similar presentations. It also acknowledges the value of a multidisciplinary approach to managing such unusual cases.

2. Case Report

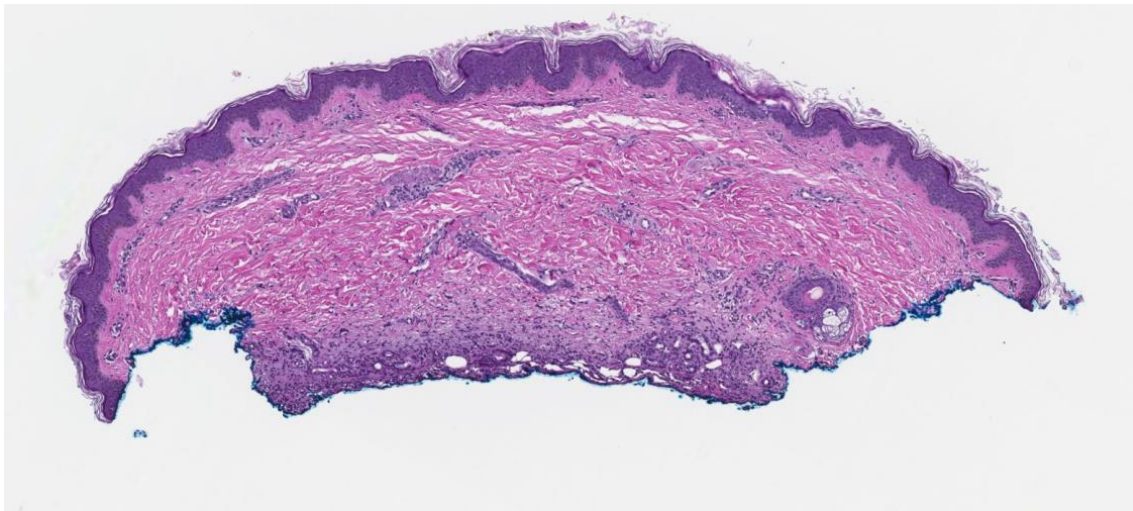
A 10-year-old girl presented to the dermatology clinic with a bruise-like scleroatrophic plaque on her left elbow, first noticed at birth but presented to the clinic when she was one year old. Since the initial observation, the lesion had evolved from a dark blue macule to a lighter, depressed scar-like atrophic plaque which gradually enlarged to wrap around her entire arm circumferentially. The lesion initially looked like a bruise, prompting the Division for Children, Youth, and Families (DCYF) to investigate child abuse. However, after excluding this possibility, a diagnosis was never pursued. Nine years later, the patient was seen at the clinic, at which time a preliminary clinical differential diagnosis of morphea versus arterial venous malformation (AVM) was thought likely. The patient stated that sometimes the area was a little painful and intermittently looks a little puffy with exercise. Results from the initial punch biopsy, however, revealed an atypical CD34+ dermal spindle cell proliferation, concerning for DFSP. The differential diagnosis included CD34 dendrocytic hamartoma. More extensive sampling was suggested. A subsequent physical exam revealed a faint ecchymotic band surrounding the lesion, raising consideration of giant cell fibroblastoma (GCF), a histologic variant of DFSP (Figure 1). Due to the concern for DFSP and related entities, a wedge biopsy was pursued by a plastic surgeon. The biopsy revealed a poorly circumscribed mass in the deep dermis and subcutaneous fat, with a largely plaque-like pattern of growth consisting of numerous spindled cells (Figure 2A). In some areas, there was honeycomb-like involvement of the subcutaneous fat. The spindled cells largely spared the adnexa. The spindled lesional cells were without significant pleomorphism and had little discernible cytoplasm. The nuclei showed finely dispersed chromatin and rare discernible nucleoli (Figure 2B). These findings were most consistent with DFSP.



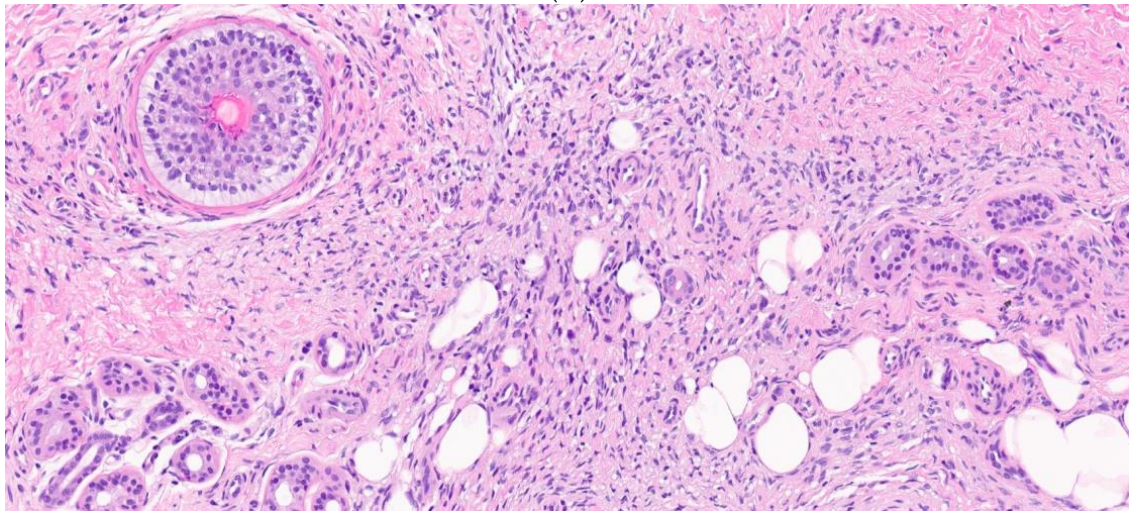
Figure 1. Dermatofibrosarcoma protuberans on left distal forearm. A bruise-like atrophic plaque measuring 6.5 cm × 2 cm spanned the circumference of the left distal forearm.

Immunohistochemical studies revealed that the lesional cells had a strong, diffuse expression of CD34 (Figure 3), without significant expression of factor XIIIa or S100. Addi-

tionally, TRK was partially expressed in some regions. Fluorescence in situ hybridization (FISH) testing by a breakapart probe revealed rearrangement of the PDGFB locus. Next-generation sequencing using the Archer FusionPlex Sarcoma Kit (Illumina^R, San Diego, CA, USA) [7] showed evidence of a COL1A1-PDGFB gene fusion. Together, the morphologic, immunohistochemical, and molecular findings were diagnostic of DFSP. MRI, pursued to assess the extent of the tumor, was reassuring for the absence of spread to skeletal musculature, bone, or bone marrow.



(A)



(B)

Figure 2. Dermatofibrosarcoma protuberans. (A) H&E stain, 10 \times : biopsy from the distal left arm revealed a poorly circumscribed proliferation in the deep dermis and subcutaneous fat in a largely plaque-like pattern of growth. (B) H&E stain, 100 \times : the proliferation consisted of numerous spindled cells. In some areas, there was honeycomb-like involvement of the subcutaneous fat. The spindled cells largely spared the adnexa. The lesional cells were without significant nuclear pleomorphism.

Subsequently, the sarcoma tumor board recommended surgical excision without adjuvant or neoadjuvant treatment. Taking into consideration patient, tumor, procedure, and logistic factors, a mutual decision was made to pursue wide local excision over micrographic options. After reviewing the potential adverse effects, including disfiguring scarring, loss of limb function, tumor recurrence, and the possible need for multiple surgeries, the parents agreed to wide excision followed by a split-thickness skin graft. The patient then underwent two excisions, with 9 days in between. The resected specimen in

the first surgery was approximately 11 cm × 7 cm. The tumor measured approximately 7 cm in its greatest dimension. The wound was covered with an equal size split-thickness bilayer matrix meshed bovine graft. Subsequent microscopic examination of the margins, however, revealed a residual tumor, prompting a second excision, a week later. A total of 5 × 2 cm of fascia and muscle were resected, resulting in a 10 × 11 cm defect (Figure 4). A wound VAC measuring 8 × 10 cm was placed overlying the split-thickness graft. The patient remains without adverse events at her 6-month post-surgery visit.

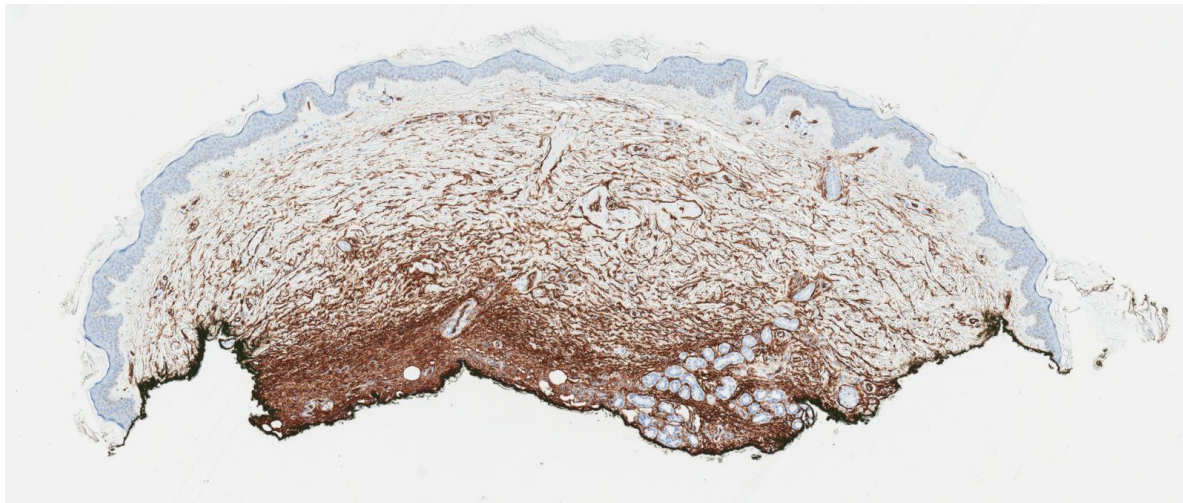


Figure 3. CD34, 10×: the lesional cells had strong, diffuse expression of CD34.



Figure 4. Post-surgical defect. A skin defect measuring 10 × 11 cm and extending to the muscle following wide surgical excision.

3. Discussion

Dermatofibrosarcoma protuberans (DFSP) is a rare cutaneous soft tissue tumor with an estimated incidence of 1–5 cases per one million individuals per year [8]. DFSP is fibrohistiocytic in origin, usually involving the dermis and subcutaneous fat. If left untreated, the neoplasm can locally invade the fascia, muscle, periosteum, and bone and, in

advanced stages, metastasize to distant organs, including the brain, lung, lymph nodes, and viscera [9]. Post-excision recurrence rates are high, especially for those on the head and neck (75%) and trunk (21%) [10,11]. The tumor typically affects adults in their second and fifth decades [12]. More than 30 years after its first description in 1924, DFSP began to be reported in children [13]. Pediatric DFSP is now more frequently reported than in the past, representing 6% of all cases [14]. Congenital DFSP, specifically, is still extremely rare. For example, a review of the literature by Valdivielso-Ramos et al. in 2012 identified 200 cases of juvenile DFSP, only 34 of which were congenital [15].

Atrophic DFSP is an asymptomatic morphea-like plaque that can persist for years [16]. The first report of this variant was in 1985 [17]. In the 32 years following, fewer than 50 additional cases were described, most of whom were adults. Some have proposed that the atrophic variant may represent an early phase of DFSP. In that model, the depressed or flat lesions would gradually enlarge to form indurated reddish-blue or violaceous nodules. DFSPs are typically localized on the trunk, the proximal extremities, and the head and neck, in 90% of cases [11,18,19].

While some congenital DFSPs have been reported, only a small number of congenital atrophic DFSPs have been documented (Table 1) [11,18,20–25]. Therefore, congenital atrophic DFSP is very difficult to diagnose. Three presentations are reported for this variant; a morphea-like, anetoderma-like depressed sclerotic, or soft plaque [26]. The present case is compatible with the first type. This atrophic form can be easily mistaken for vascular malformation, morphea, tufted angioma, bruise, or melanocytic nevus, adding to the challenge of timely detection.

For a review of all published cases of congenital atrophic DFSP, a literature review was performed in PubMed with input from Google Scholar using Rayyan application [27]. The search terms used were a combination of the words “congenital” or “juvenile”, “atrophic”, “morphea-like”, or “bruise-like”, with “dermatofibrosarcoma protuberans” or “DFSP”. Records were excluded based on the language, scope, accessibility, and ages of the patients described. Full-text articles were assessed for eligibility, and eight studies (Figure 5) were found to have described novel reports of congenital atrophic DFSP with a total of fourteen cases (Table 1). Nine were initially misdiagnosed with an average of nine years before the detection of DFSP. These lesions evolved to the protuberant phase after a quiescent period ranging from two to ten years (Table 1).

The table compares the current case to the previously reported 14 cases of congenital atrophic DFSP. It highlights the differences in the age of presentation and diagnosis as well as the range of time between lesion presentation and development of the protuberant variant of DFSP. Six cases described by Maire et al. did not describe age of presentation, but the authors noted a time delay ranging from 5 ½ months to 15 years between onset of lesion and proper diagnosis [25]. Lesions were located on the trunk and extremities except for two lesions on the head and neck. Ten cases were positive for the translocation t (17; 22) or the fusion gene including the current case. All cases were treated with wide excision with the exception of two cases where Mohs surgery was pursued.

Molecular studies have demonstrated the presence of supernumerary ring chromosomes derived from chromosomes 17 and 22 or chromosomal translocation t (17; 22) (q22; q13) in over 90% of DFSPs. These cytogenetic abnormalities result in the fusion of collagen type 1-alpha 1 (COL1A1 at 17q22) and platelet-derived growth factor beta (PDGFB at 22q13) genes. As a result, the PDGFB gene is overexpressed under the influence of the COL1A1 promoter, resulting in constant activation of the PDGF receptor β protein-tyrosine kinase [28]. The COL1A1-PDGFB fusion gene is highly specific and sensitive to DFSP. Unlike the adult variant, this cytogenetic abnormality has not been commonly documented in childhood or congenital presentations, possibly due to under-reporting. Additionally, while the aberrant fusion gene is found in more than 90% of protuberant DFSP cases, the first atrophic DFSP confirmed by the COL1A1-PDGFB fusion gene was only reported in 2007 [16,25]. Despite the importance of this fusion gene for tumor growth in adults, not all published pediatric cases had gene fusion testing performed [20,22–24].

Table 1. Comparison of current case to the reported cases of congenital atrophic DFSP.

Reference	Age of Presentation	Age of Diagnosis	Quiescent Period to Protuberant Phase	Location	Misdiagnosis	Translocation T (17; 22)/ COL1A1-PDGFB Fusion Gene	Treatment
Salem et al.	One year *	10 years	Remained atrophic	Forearm	Bruise, morphea	Positive	Wide excision
Makino et al. [18]	Birth	19 years	10 years	Anterior chest	N/A	Positive	Wide excision
Marini et al. [20]	Birth	16 years	7 years	Anterior leg	Congenital fibroma	N/A	Mohs surgery
Han et al. [21]	Birth	6 years	4 years	Posterior neck	Dermatitis	Negative	Wide excision
Feramisco et al. [11]	7 m *	7 m	Remained atrophic	Left inguinal	N/A	Positive	Mohs surgery
Weinstein et al. [22]	6 years *	14 years	6 years	Back	Vascular malformation, Fibrous histiocytoma	N/A	Wide excision
Martin et al. [23,24]	Birth	13 years	7 years	Calf	Hematoma	Positive	Wide excision
Weinstein et al. [22]	6 m *	1 year	6 m	Right thigh	Aplasia cutis	N/A	Wide excision
Martin et al. [23,24]	Birth	3 years	2 years	Periumbilical	Morphea	N/A	Wide excision
Maire et al. [25]	N/A	3 years	N/A	Lumbar	“Difficult to Characterize”	Positive	Wide excision
Maire et al. [25]	N/A	11 years	N/A	Lumbar	N/A	Positive	Wide excision
Maire et al. [25]	2 years *	7 years	N/A	Occipital	Aplasia cutis, Fibrous hamartoma, Infantile fibromatosis	Positive	Wide excision
Maire et al. [25]	N/A	10 years	N/A	Foot	Difficult to characterize	Positive	Wide excision
Maire et al. [25]	15 years *	17 years	N/A	Trunk	Fibromatosis, Xanthomatous hamartoma, Aplasia cutis, Infantile fibrosarcoma	Positive	Wide excision
Maire et al. [25]	2 years *	3 years	N/A	Thorax	Fibrosarcoma, Angioma, Mastocytoma	N/A	Wide excision

* Patients who had the lesions at birth but did not present until later in life.

PRISMA flow diagram

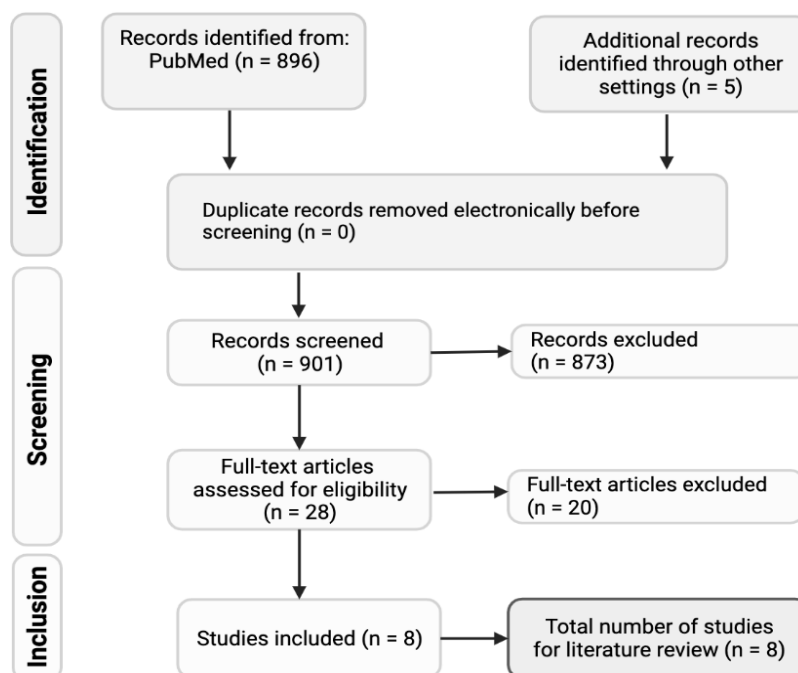


Figure 5. Prism chart of congenital atrophic DFSP review of literature. Adapted from “PRISMA flow diagram for systematic reviews”, by BioRender.com (2023). Retrieved from <https://app.biorender.com/biorender-templates> (accessed on 19 July 2018).

Clinically DFSP can be mistaken for vascular neoplasms including tufted angioma (TA) [29]. A distinction from DFSP can be made morphologically, where TA typically shows numerous capillary tufts scattered in a cannonball distribution and, unlike DFSP, exhibits a less prominent spindled component [30].

One of the important histological differentials in our case was medallion-like dermal dendrocyte hamartoma (ML-DDH). ML-DDH is clinically almost indistinguishable from the atrophic variant of DFSP. ML-DDH also overlaps microscopically with the atrophic form of DFSP: both lack the hypercellularity or storiform pattern of conventional DFSPs [31,32]. Instead, atrophic DFSP has a plexiform architecture and a loose angiomatous stroma [32]. Morphologic features that can help distinguish atrophic DFSP from ML-DDH include a deeper more infiltrative architecture in the former, as opposed to superficial band-like architecture in the latter [18,32]. Additionally, ML-DDH often shows vertically oriented fibroblasts with associated dilated venules on a bed of horizontally oriented fibroblasts. In contrast, atrophic DFSP is characterized by horizontally arranged cellular spindle-cell tracts and fascicles, along with occasional loosely aggregated small fibroblasts within a slightly myxoid stroma. Immunohistochemistry is often not helpful in the distinction, as both express CD34, infrequently express factor XIIIa, and do not typically stain positive for S100. Importantly, molecular testing for the COL1A1-PDGFB gene fusion is typically decisive [33–35]. The evolution of ML-DDH differs substantially from that of DFSP, where its size increases proportionally to the body’s growth and is therefore perceived as stable, as opposed to the progressive nature of DFSP. As such, ML-DDH requires only simple excision and not mutilating surgeries as with DFSP [31].

A Bednar tumor (BT) and Giant cell fibrolastoma (GCF) are two histological variants of DFSP [35]. Both share the morphological findings of dermal or subcutaneous location, myxoid changes, and sparing of adnexa and prominent vasculature. However, the presence of solid areas with stromal giant cells, onion skin-like chronic inflammation, pseudo vascular spaces lined by giant cells, consistent hemorrhage, and the absence of a storiform

pattern are more characteristic of GCF [36,37]. BT is a pigmented variant of DFSP. The presence of melanin-containing dendritic cells is the only feature distinguishing BT from DFSP [35].

The first line treatment for localized DFSP is surgical resection with negative margins, as simple excision has a 50% risk of recurrence [36]. The excision can be performed by micrographic surgery including Mohs and slow Mohs techniques or wide local excision. In our case, these techniques were discussed, but not pursued. The discussion at a multi-disciplinary tumor board, involving plastic surgery, Mohs surgery, pediatric dermatology, and dermatopathology, arrived at the following reasons to pursue wide local excision: (1) The patient's young age would require doing these interventions under general anesthesia. (2) Mohs may incur a long waiting time for tissue processing of a large excision which would have resulted in prolonged exposure to anesthesia in this pediatric patient. (3) Delayed wound closure, in the case of staged excision/"slow Mohs", would have been difficult for the young child to tolerate. (4) Much of the literature reporting outcomes of these techniques in DFSPs is based on data from older patients. In such patients, DFSP has often been present for multiple decades and has a significant risk of subclinical spread. In our case, this risk is deemed to be small. (5) Due to the low likelihood of subclinical spread, removing the tumor with the required margins would give the plastic surgeon a high likelihood of clearance with a linear closure. (6) The absence of tissue rearrangement would mean that, even if there was a positive margin, simply submitting a clearly oriented excision to pathology would allow for later identification and removal of residual tumor. A description of three surgical options and their advantages and disadvantages are summarized in (Table 2) [38–41].

The recurrence rate is different based on the selected surgical modality. Lower rates were reported with MMS compared to wide local excision. Half of the cases that do recur are presented in the first year post-surgery. An additional 30% recur in the two years after that. Recurrent neoplasms have a propensity for spreading to muscle and bone and carry a higher risk of distant metastasis [36]. Therefore, postoperative follow-up at three to six months intervals is indicated for at least three years, followed by annual checks for life [20]. Our patient did not show any signs of recurrence or adverse effects at her 6-month follow-up visit. Adjuvant treatment with radiotherapy is used to control residual lesions or as a complimentary treatment in incomplete excisions. Chemotherapy is recommended for metastases [20].

Timely diagnosis of congenital DFSP is critical for treatment. Early excision, in which relatively smaller tumors are treated, minimizes the risk of extensive surgical scarring, disfigurement, and functional disability. Our patient underwent a wide excision with a 10 × 11 cm defect that required a split-thickness skin graft; had the diagnosis been made earlier in the 9-year course of the disease, a less radical treatment option might have sufficed.

Table 2. Comparison of the three surgical options used to treat congenital atrophic DFSP. The table describes the three surgical options used to treat congenital atrophic DFSP and summarizes their advantages and disadvantages.

	Wide Local Excision	Mohs Excision	Staged Excision “Slow Mohs”
Description	Excision with 2–4 cm margins followed by closure of the defect	<ul style="list-style-type: none"> - Real-time analysis of frozen horizontal tissue sections to examine all margins, by the dermatologic surgeon. - This process is repeated until no further residual tumor is seen 	<ul style="list-style-type: none"> - En bloc excision of the tumor with a margin of normal-looking tissue in a staged approach. - The tissue is fixed in formalin and embedded in paraffin - The wound is then temporized with dressings while histopathologic analysis is achieved - Secondary operation to either remove the incompletely excised tumor, (if the margin is positive) or reconstruct the defect, (if the margin is negative, the defect is reconstructed).
Advantage	<ul style="list-style-type: none"> - Faster than MMS * - Closure of the defect is comparatively less complex - Relatively lower cost 	<ul style="list-style-type: none"> - Spares uninvolved tissue to the greatest extent possible - Better cosmetic appearance - Low tumor recurrence rate 	<ul style="list-style-type: none"> - Narrower surgical margins - The specimens are analyzed off-site by a dermatopathologist using conventional sections rather than frozen tissue. Less operative time - Shorter exposure to anesthesia
Disadvantage	<ul style="list-style-type: none"> - More disfiguring - Higher reported rate of tumor recurrence compared to MMS* surgeries - Requires general anesthesia 	<ul style="list-style-type: none"> - Requires considerable training and a specialized ancillary team - Requires specialized equipment to process and examine the specimens near the operation room - Time consuming - Prolonged exposure to general anesthesia in those who cannot maintain immobility (e.g., children) - Frozen sections can be more difficult to interpret histologically due to crystallization and other artifacts 	<ul style="list-style-type: none"> - Risks associated with delayed wound closure - Problems with compliance with dressings while awaiting pathology results.

MMS *: Mohs Micrographic surgery.

4. Conclusions

Congenital atrophic DFSP is an extremely uncommon intermediate-grade neoplasm. Only eight cases have been reported in the literature. Some experts have proposed that it may be an early stage of the nodular form, with a variable quiescent period. While often not performed in congenital cases, the molecular identification of the COL1A1-PDGFB fusion gene is an important diagnostic feature for DFSP. Early detection and surgical management are crucial to avoiding large excisions with their subsequent disability and disfigurement. Micrographic surgical techniques are associated with lower recurrence rates and better cosmetic outcomes and are currently recommended over wide local excision. However, the surgical modality of choice should be decided on a case-by-case basis taking into consideration patient, tumor, procedure, and logistic factors.

Author Contributions: Conceptualization: I.S., A.S. and K.B.; methodology, I.S., A.S. and K.B.; validation, A.S., J.A.M., J.H.S. and M.L.; formal analysis, K.B.; investigation, I.S., A.S., J.A.M., J.H.S. and M.L.; resources, I.S., A.S., J.A.M., J.H.S., K.B. and M.L.; data curation, I.S., A.S., M.L. and K.B.; writing—original draft preparation, I.S., A.S., J.A.M. and J.H.S.; writing—review and editing, A.S., K.B., J.A.M., J.H.S. and M.L.; visualization, I.S., A.S. and K.B.; supervision, A.S.; project administration, I.S.; funding acquisition, I.S. All authors have read and agreed to the published version of the manuscript.

Funding: This research received no external funding.

Informed Consent Statement: Informed consent was obtained from all subjects involved in the study.

Data Availability Statement: Data supporting reported results can be found in references.

Conflicts of Interest: The authors declare no conflict of interest.

References

1. Brooks, J.; Ramsey, M.L. Dermatofibrosarcoma Protuberans. 2018. Available online: <https://europepmc.org/article/NBK/nbk513305> (accessed on 19 July 2018).
2. Van Lee, C.; Kan, W.C.; Gran, S.; Mooyaart, A.; Mureau, M.; Williams, H.; Matin, R.; Van Den Bos, R.; Hollestein, L. Dermatofibrosarcoma protuberans re-excision and recurrence rates in the Netherlands Between 1989 and 2016. *Acta Derm.-Venereol.* **2019**, *99*, 1160–1165. [[CrossRef](#)] [[PubMed](#)]
3. Badhey, A.K.; Tikhtman, R.; Tang, A.L. Management of dermatofibrosarcoma protuberans. *Curr. Opin. Otolaryngol. Head Neck Surg.* **2021**, *29*, 278–282. [[CrossRef](#)] [[PubMed](#)]
4. Chen, Y.; Shi, Y.Z.; Feng, X.H.; Wang, X.T.; He, X.L.; Zhao, M. Novel TNC-PDGFD fusion in fibrosarcomatous dermatofibrosarcoma protuberans: A case report. *Diagn. Pathol.* **2021**, *16*, 1–6. [[CrossRef](#)]
5. Reddy, C.; Hayward, P.; Thompson, P.; Kan, A. Dermatofibrosarcoma protuberans in children. *J. Plast. Reconstr. Aesthetic Surg.* **2009**, *62*, 819–823. [[CrossRef](#)]
6. Posso-De Los Rios, C.J.; Lara-Corrales, I.; Ho, N. Dermatofibrosarcoma protuberans in pediatric patients: A report of 17 cases. *J. Cutan. Med. Surg.* **2014**, *18*, 180–185. [[CrossRef](#)] [[PubMed](#)]
7. Lam, S.W.; Cleton-Jansen, A.M.; Cleven, A.H.; Ruano, D.; van Wezel, T.; Szuhai, K.; Bovée, J.V. Molecular analysis of gene fusions in bone and soft tissue tumors by anchored multiplex PCR-based targeted next-generation sequencing. *J. Mol. Diagn.* **2018**, *20*, 653–663. [[CrossRef](#)]
8. Zhou, Y.; Chin, J.; Strutin, M.D.; Lomiguen, C.M. Unmasking dermatofibrosarcoma protuberans: Case report of an atypical presentation complicated by post-surgical excision. *Int. J. Surg. Case Rep.* **2020**, *69*, 101–104. [[CrossRef](#)]
9. Souiki, T.; Belhaj, A.; Ait Abderrhim, A.; Alami, B.; Tahiri, L.; Chbani, L.; Ibn Majdoub, K.; Toughrai, I.; Mazaz, K. Dermatofibrosarcoma protuberans of the anterior abdominal wall: Case report and literature review. *J. Surg. Case Rep.* **2022**, *2022*, rjac272. [[CrossRef](#)]
10. Bakry, O.; Attia, A. Atrophic dermatofibrosarcoma protuberans. *J. Dermatol. Case Rep.* **2012**, *6*, 14. [[CrossRef](#)]
11. Feramisco, J.; Larsen, F.; Weitzul, S.; Cockerell, C.; Ghali, F. Congenital atrophic dermatofibrosarcoma protuberans in a 7-month-old boy treated with Mohs micrographic surgery. *Pediatr. Dermatol.* **2008**, *25*, 455–459. [[CrossRef](#)]
12. Llombart, B.; Serra, C.; Requena, C.; Alsina, M.; Morgado-Carrasco, D.; Través, V.; Sanmartín, O. Guidelines for diagnosis and treatment of cutaneous sarcomas: Dermatofibrosarcoma protuberans. *Actas Dermo-Sifiliográficas* **2018**, *109*, 868–877. [[CrossRef](#)] [[PubMed](#)]
13. Rao, C.; Shetty K, J.; Prasad, H.L.K. An uncommon malignancy of the breast; dermatofibrosarcoma protruberans—A case report. *Indian J. Surg. Oncol.* **2012**, *3*, 242–244. [[CrossRef](#)]

14. Reilly, D.J.; Loo, Y.L.; Alexander, W.M.; Wilks, D.J.; MacGregor, D.; Coombs, C.J. Diagnostic and Management Considerations in Pediatric Dermatofibrosarcoma Protuberans. *Ann. Plast. Surg.* **2022**, *88*, 319–322. [[CrossRef](#)] [[PubMed](#)]
15. Valdivielso-Ramos, M.; Hernanz, J.M. Dermatofibrosarcoma protuberans en la infancia. *Actas Dermo-Sifiliogr.* **2012**, *103*, 863–873. [[CrossRef](#)] [[PubMed](#)]
16. Liu, Y.; Zhang, B.; Han, X.; Ma, L. Pediatric atrophic dermatofibrosarcoma protuberans. *Pediatr. Investig.* **2017**, *1*, 50. [[CrossRef](#)] [[PubMed](#)]
17. Akay, B.N.; Unlu, E.; Erdem, C.; Heper, A.O. Dermatoscopic findings of atrophic dermatofibrosarcoma protuberans. *Dermatol. Pract. Concept.* **2015**, *5*, 71. [[CrossRef](#)] [[PubMed](#)]
18. Makino, M.; Sasaoka, S.; Nakanishi, G.; Makino, E.; Fujimoto, W. Congenital atrophic dermatofibrosarcoma protuberans detected by COL1A1-PDGFB rearrangement. *Diagn. Pathol.* **2016**, *11*, 1–6. [[CrossRef](#)] [[PubMed](#)]
19. Hanabusa, M.; Kamo, R.; Harada, T.; Ishii, M. Dermatofibrosarcoma protuberans with atrophic appearance at early stage of the tumor. *J. Dermatol.* **2007**, *34*, 336–339. [[CrossRef](#)] [[PubMed](#)]
20. Marini, M.; Saponaro, A.; Magariños, G.; De Baldrich, A.; Lynch, P.; Remorino, L. Congenital atrophic dermatofibrosarcoma protuberans. *Int. J. Dermatol.* **2001**, *40*, 448–450. [[CrossRef](#)]
21. Han, H.H.; Lim, S.Y.; Park, Y.M.; Rhie, J.W. Congenital dermatofibrosarcoma protuberans: A case report and literature review. *Ann. Dermatol.* **2015**, *27*, 597–600. [[CrossRef](#)]
22. Weinstein, J.M.; Drolet, B.A.; Esterly, N.B.; Rogers, M.; Bauer, B.S.; Wagner, A.M.; Mancini, A.J. Congenital dermatofibrosarcoma protuberans: Variability in presentation. *Arch. Dermatol.* **2003**, *139*, 207–211. [[CrossRef](#)]
23. Martin, L.; Combemale, P.; Dupin, M.; Chouvet, B.; Kanitakis, J.; Bouyssou-Gauthier, M.L.; Dubreuil, G.; Claudy, A.; Grimand, P.G. The atrophic variant of dermatofibrosarcoma protuberans in childhood: A report of six cases. *Br. J. Dermatol.* **1998**, *139*, 719–725. [[PubMed](#)]
24. Martin, L.; Piette, F.; Blanc, P.; Mortier, L.; Avril, M.F.; Delaunay, M.M.; Dréno, B.; Granel, F.; Mantoux, F.; Aubin, F.; et al. Clinical variants of the preprotuberant stage of dermatofibrosarcoma protuberans. *Br. J. Dermatol.* **2005**, *153*, 932–936. [[CrossRef](#)] [[PubMed](#)]
25. Maire, G.; Fraitag, S.; Galmiche, L.; Keslair, F.; Ebran, N.; Terrier-Lacombe, M.J.; de Prost, Y.; Pedeutour, F. A clinical, histologic, and molecular study of 9 cases of congenital dermatofibrosarcoma protuberans. *Arch. Dermatol.* **2007**, *143*, 203–210. [[CrossRef](#)] [[PubMed](#)]
26. Xu, S.; Zhao, L.; Wang, J. Atrophic dermatofibrosarcoma protuberans: A clinicopathological study of 16 cases. *Pathology* **2019**, *51*, 615–620. [[CrossRef](#)]
27. Ouzzani, M.; Hammady, H.; Fedorowicz, Z.; Elmagarmid, A. Rayyan — a web and mobile app for systematic reviews. *Syst. Rev.* **2016**, *5*, 210. [[CrossRef](#)]
28. Hao, X.; Billings, S.D.; Wu, F.; Stultz, T.W.; Procop, G.W.; Mirkin, G.; Vidimos, A.T. Dermatofibrosarcoma protuberans: Update on the diagnosis and treatment. *J. Clin. Med.* **2020**, *9*, 1752. [[CrossRef](#)] [[PubMed](#)]
29. Ghosh, S.K.; Bandyopadhyay, D.; Ghosh, A.; Biswas, S.K.; Barma, K.D. Acquired multifocal tufted angiomas in an immunocompetent young adult. *Indian J. Dermatol.* **2011**, *56*, 412. [[CrossRef](#)]
30. Jandali, S.; Kirschner, R.E. Congenital leukemia cutis presenting as multiple violaceous lesions in a newborn. *Ann. Plast. Surg.* **2011**, *66*, 310–312. [[CrossRef](#)]
31. Tantcheva-Poor, I.; Marathovouniotis, N.; Kutzner, H.; Mentzel, T. Vascular congenital dermatofibrosarcoma protuberans: A new histological variant of dermatofibrosarcoma protuberans. *Am. J. Dermatopathol.* **2012**, *34*, e46–e49. [[CrossRef](#)]
32. Marque, M.; Bessis, D.; Pedeutour, F.; Viseux, V.; Guillot, B.; Fraitag-Spinner, S. Medallion-like dermal dendrocyte hamartoma: The main diagnostic pitfall is congenital atrophic dermatofibrosarcoma. *Br. J. Dermatol.* **2008**, *160*, 190–193. [[CrossRef](#)] [[PubMed](#)]
33. Bai, J.; Liu, B.; Liu, T.; Qiao, J.; Fang, H. Atrophic Pigmented Dermatofibrosarcoma Protuberans: A Case Report and Literature Review. *Front. Oncol.* **2021**, *11*, 669754. [[CrossRef](#)]
34. Rodríguez-Jurado, R.; Palacios, C.; Durán-McKinster, C.; Mercadillo, P.; Orozco-Covarrubias, L.; del Mar Saez-de-Ocariz, M.; Ruiz-Maldonado, R. Medallion-like dermal dendrocyte hamartoma: A new clinically and histopathologically distinct lesion. *J. Am. Acad. Dermatol.* **2004**, *51*, 359–363. [[CrossRef](#)] [[PubMed](#)]
35. Shah, K.N.; Anderson, E.; Junkins-Hopkins, J.; James, W.D. Medallion-like dermal dendrocyte hamartoma. *Pediatr. Dermatol.* **2007**, *24*, 632–636. [[CrossRef](#)] [[PubMed](#)]
36. Sirvent, N.; Maire, G.; Pedeutour, F. Genetics of dermatofibrosarcoma protuberans family of tumors: From ring chromosomes to tyrosine kinase inhibitor treatment. *Genes Chromosomes Cancer* **2003**, *37*, 1–19. [[CrossRef](#)] [[PubMed](#)]
37. Kadam, S.S.; Kadam, T. Recurrent Dermatofibrosarcoma Protruberance over Shoulder: An Unresolved Problem. *Int. Res. J. Oncol.* **2021**, *4*, 11–20.
38. Sleiwah, A.; Wright, T.C.; Chapman, T.; Dangoor, A.; Maggiani, F.; Clancy, R. Dermatofibrosarcoma protuberans in children. *Curr. Treat. Options Oncol.* **2022**, *23*, 843–854. [[CrossRef](#)] [[PubMed](#)]
39. Chicaud, M.; Frassati-Biaggi, A.; Kaltenbach, S.; Karanian, M.; Orbach, D.; Fraitag, S. Dermatofibrosarcoma protuberans, fibrosarcomatous variant: A rare tumor in children. *Pediatr. Dermatol.* **2021**, *38*, 217–222. [[CrossRef](#)] [[PubMed](#)]

40. Macarenco, R.S.; Zamolyi, R.; Franco, M.F.; Nascimento, A.G.; Abott, J.J.; Wang, X.; Erickson-Johnson, M.R.; Oliveira, A.M. Genomic gains of COL1A1-PDFGB occur in the histologic evolution of giant cell fibroblastoma into dermatofibrosarcoma protuberans. *Genes Chromosomes Cancer* **2008**, *47*, 260–265. [[CrossRef](#)] [[PubMed](#)]
41. Singh, A.P.; Gupta, A.K.; Ansari, M.; Mehra, S.K.; Barolia, D.K. Very rare childhood tumor: Giant cell fibroblastoma. *Med. J. Dr. DY Patil Univ.* **2018**, *11*, 452. [[CrossRef](#)]

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