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Case Report

Additional Wide Resection of Infantile Dermatofibrosarcoma Protuberans after Unplanned Excision: A Case Report

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Keywords
Infantile dermatofibrosarcoma protuberans · Unplanned excision · Skin tumor · Surgical margin · Pediatric sarcoma

Abstract
Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive intermediate soft tissue neoplasm that occurs in the dermis. DFSP generally occurs in young to middle-aged adults and rarely in infancy. Because of its extreme rarity, DFSP is difficult to diagnose and treat, especially when it occurs in infancy. In this paper, we reported a case of infantile DFSP in which we performed additional wide resection with a 3-cm horizontal margin for a mass that had previously undergone unplanned excision. No tumor recurrence has been seen for 3 years postoperatively. We suggest that the possibility of DFSP should always be considered when an enlarging superficial mass is identified on the trunk, even in an infant. Additionally, radical local treatment is as important for DFSP in infancy as it is for DFSP in adults, even after unplanned excision.

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive intermediate fibroblastic tumor [1]. Because DFSP occurs most often in young adults [2], few reports describe the details of the clinical course of infantile cases [1]. Because of the paucity of reported cases,

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there is no established surgical margin for infantile DFSP, especially after unplanned previous excision. We reported a successful case of infantile DFSP who underwent additional wide resection after unplanned excision and achieved a good oncological outcome. We also reviewed the associated literature. The CARE Checklist has been completed by the authors for this case report and attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000530639).

Case Presentation

The patient was a 15-month-old girl who presented with a history of excision of a lump on her back at a nearby hospital. Her parents first noticed the superficial elastic-soft lump when she was 8 months old (Fig. 1a). The mass measured approximately 4 cm in diameter and was considered a benign lesion, such as myxoma, and was observed without intervention at the nearby hospital for 5 months. However, the mass increased in size during observation. When the patient was 13 months of age, the mass was described as elastic-hard and generally mobile but firmly fixed to the overlying skin, measuring 5.5 cm in diameter. Blood examination results were within the normal limit. Although the MRI findings strongly suggested a malignant tumor in a retrospective review (Fig. 1b–d), the mass was interpreted as a benign lesion and was excised unplanned at the age of 13 months (Fig. 1e, f). Because the pathological diagnosis suggested a malignant tumor, the patient was referred to our hospital for further evaluation and treatment at the age of 15 months. Precise pathological examination revealed spindle cell proliferation and that the mass was positive for CD34 by immunohistochemistry and positive for PDGFB rearrangement by fluorescence in situ hybridization, indicating the final diagnosis of DFSP (Fig. 1g–j). The surgical margin of the previous excision was positive. Upon referral, physical examination revealed a scar on the patient's back measuring 7 cm in length without underlying palpable nodules (Fig. 2a). MRI suggested a residual tumor on gadolinium enhancement (Fig. 2b), and computed tomography revealed no metastatic lesions. At the age of 16 months, 3 months after the unplanned excision, additional wide resection was performed with a 3-cm horizontal margin and with resection of the underlying fascia to achieve a clear vertical margin. Primary closure was achieved without requiring a secondary graft or flap coverage for the massive skin defect measuring 15 × 11 cm (Fig. 3a–c). The pathological findings revealed the presence of residual tumor cells but the absence of tumor cells at the surgical margin (Fig. 3d–g). Through physical examination and imaging test, we confirmed that the patient has been tumor-free for 3 years since the additional resection.

Discussion

DFSP is a rare soft tissue neoplasm frequently originating from the dermis on the trunk [1] that accounts for approximately 0.1% of all cutaneous malignant tumors [3]. DFSP occurs most often in young and middle-aged adults [2], and in infancy in exceptionally rare cases [1]. To the best of our knowledge, 52 cases of congenital and infantile DFSP have been reported since 2000 (Table 1). Among the reported cases, the median time to diagnosis was 6 years (range: 3 months to 29 years), and 74% (37/50) of the cases required more than 1 year for diagnosis. This clearly shows that diagnosis of DFSP in infants is difficult and can take a long time. Regarding the imaging findings, the signal intensity of DFSP on MRI is frequently low on T1-weighted images and high on T2-weighted images, and contrast enhancement effects are present [4]. The MRI findings in our case were compatible with DFSP; however, the MRI findings were not specific, and she was not diagnosed with DFSP at the nearby hospital. In our

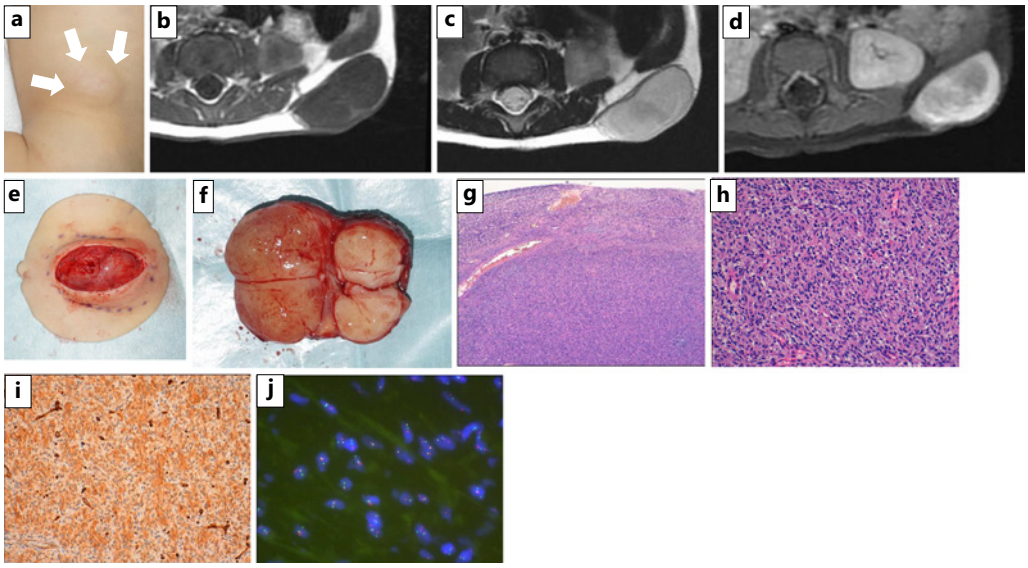

Fig. 1. Image and operative findings and pathological findings at a nearby hospital. **a** Tumor appearance (white arrows). The tumor was a superficial, elastic-soft lump without a notable change in skin color. **b–d** MRI findings of the primary tumor. The tumor was a relatively well-defined lesion in the subcutaneous layer, measuring 47 mm in diameter, with homogenously low signal on T1-weighted images (**b**), homogenously high signal on T2-weighted images (**c**), and homogenously high signal on T1-weighted gadolinium-enhanced images (**d**). **e**, **f** Operative findings at the nearby hospital. The tumor was marginally excised upon initial surgery. The tumor was not excised with the overlying skin and was macroscopically exposed through the tumor capsule. **g**, **h** Hematoxylin and eosin (HE) staining showed spindle cell proliferation. The margin was positive microscopically. Low-power field **(g)** and high-power field **(h)**. **i** An immunohistochemical study showed the tumor cells were positive for CD34. **j** Fluorescent in situ hybridization (FISH) showed positive for PDGFB rearrangement. The red spots indicated the presence of the *COL1A1* gene. The green spots showed the presence of the *PDGFB* gene. The chromosome that was labeled with yellow spots was the one where rearrangement was present.

case, the mass had been observed untreated as a benign tumor at the nearby hospital for 7 months while the tumor had increased in size. When an enlarging superficial mass is observed, even in an infant, malignant tumor should be suspected and a biopsy should be performed.

DFSP in adults shows a strong tendency for local recurrence [5]. Thus, appropriate local treatment is essential for DFSP to minimize the possibility of local recurrence. Mohs surgery has been reported as useful local treatment, especially for those occurring on the face or neck, where these tumors are difficult to resect with adequate wide surgical margins while preserving the surrounding tissues [6], however it has several disadvantages, namely, the extensive time required, depending on the size of the tumor, and the requirement for frequent local anesthesia [7]. In contrast, in a recent retrospective study of 34 patients who underwent wide resection, the 5-year disease-free survival rate was 100% and the local recurrence rate was 0%, indicating that wide resection is highly effective [8]. The efficacy and roles of radiotherapy and chemotherapy in resectable DFSP are controversial [5, 6]. Therefore, wide resection is considered the most effective local treatment for resectable DFSP.

The best way to achieve adequate surgical margins for adult DFSP during wide resection has been debated. In a single-center study of 33 adult patients with DFSP who underwent wide resection with various surgical margins, 3 patients with histological margins of 2 cm or

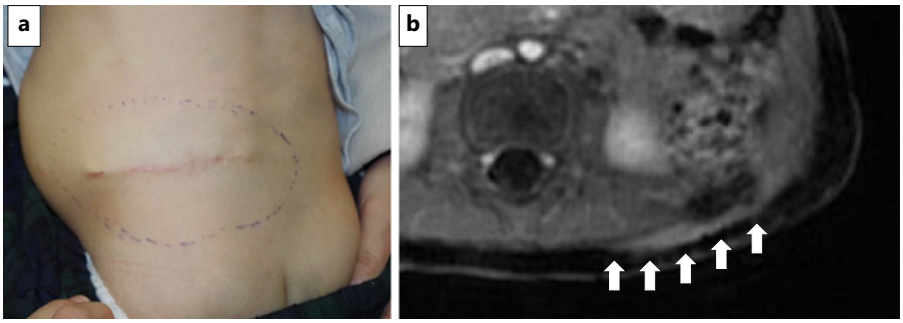

Fig. 2. Findings upon referral to our hospital. **a** Physical examination at presentation to our hospital revealed she had a scar on her back measuring 7 cm in length without any palpable nodules beneath. **b** MRI findings after primary surgery. T1-weighted images with gadolinium enhancement suggested a residual lesion at the primary site (white arrows).

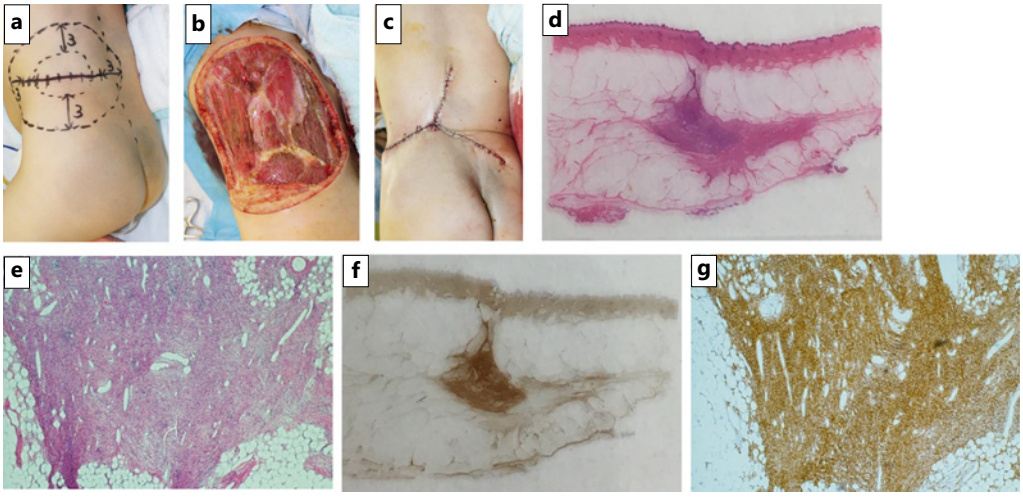

Fig. 3. Operative and pathological findings. **a** Preoperative planning. We planned to perform additional wide resection 3 months following the unplanned excision with a 3-cm margin in the horizontal direction and resected the underlying fascia for the margin in the vertical direction. **b** Massive skin defect measuring 15 × 11 cm in diameter after additional wide resection of the lesion. **c** A three-point anchoring suture was done for primary closure without skin graft or flap coverage. **d**, **e** HE staining of the specimen at low-power field (**d**) and at high-power field (**e**) revealed the presence of spindle cell proliferation. There was no fascial invasion in the deep direction, and the horizontal margin was negative at 14 mm. **f**, **g** Immunohistochemical study at low-power field (**f**) and at high-power field (**g**) showed the tumor cells were positive for CD34, CD68, and PDGFβ, which indicated DFSP (data not shown).

less experienced recurrence [9], and the authors suggested that a histological margin of 2 cm or more was necessary. In accordance with these findings, we used a horizontal margin of 2 cm or more from the enhanced area on MRI as a criterion for wide resection in the initial treatment of DFSP.

For sarcomas in general, 5-year disease-free survival worsens when unplanned excision is performed and residual tumor is present [10]. After unplanned resection, the reported survival rate is 68.8% with a residual lesion and 92% if the lesion is completely removed [10]. Therefore, when residual disease is suspected after unplanned surgery, it is important to plan an additional wide resection with sufficient surgical margins. However, setting an appropriate

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Table 1. Clinical features of 52 cases of DFSP that were reported after the year 2000

Case number	Author	Published year	Location	Age at onset	Age at diagnosis	Treatment	Surgical margin	Recurrence	Follow-up duration
1	Checketts	2000	Trunk	Congenital	5 years	Wide resection	2 cm	None	9 months
2	Marini	2001	Lower limb	Congenital	16 years	Mohs surgery	NA	None	9 months
3	Weinstein	2003	Head	Congenital	8 months	Wide resection	3 cm	None	1 year
4			Lower limb	Congenital	3 years 6 months	Wide resection	1 cm	None	2 years
5			Trunk	Congenital	14 years	Wide resection	NA	None	6 years
6			Neck	Congenital	1 year 9 months	Wide resection	NA	Recurrence	3 years 6 months
7			Trunk	Congenital	5 months	Wide resection	NA	None	3 years
8			Lower limb	Congenital	1 year	Wide resection	NA	NA	NA
9	Terrier-Lacombe	2003	Trunk	Congenital	NA	Wide resection	3 cm	None	3 years
10			Lower limb	Congenital	NA	Wide resection	NA	NA	NA
11	Price	2005	Lower limb	Congenital	1 year 6 months	Imatinib	–	NA	NA
12	Gu	2005	Trunk	Congenital	29 years	Wide resection	NA	None	1 year
13	Thornton	2005	Trunk	Congenital	15 years	Mohs surgery	2.5 cm	None	10 years
14			Trunk	Congenital	3 years	Mohs surgery	1.5 cm	None	2 years 5 months
15			Trunk	Congenital	16 years	Mohs surgery	2 cm	None	1 year 3 months
16			Upper limb	Congenital	13 years	Mohs surgery	2 cm	None	1 year
17			Trunk	Congenital	2 years	Mohs surgery	2 cm	None	9 months
18	Fidalgo	2006	Upper limb	Congenital	5 years	Wide resection	2 cm	None	3 years
19	Musiesa	2007	Lower limb	Congenital	10 years	Wide resection	NA	NA	NA
20	Stebut	2007	Trunk	Congenital	3 years	Wide resection	2 cm	None	NA
21	Marie	2007	Trunk	Congenital	6 months	Wide resection	NA	None	7 months
22			Trunk	Congenital	3 years	Wide resection	NA	NA	NA
23			Trunk	Congenital	11 years	Wide resection	NA	NA	NA

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Table 1 (continued)

Case number	Author	Published year	Location	Age at onset	Age at diagnosis	Treatment	Surgical margin	Recurrence	Follow-up duration
24			Head	Congenital	2 years	Excision	NA	NA	NA
25			Lower limb	Congenital	10 years	Wide resection	NA	NA	NA
26			Lower limb	Congenital	3 years	None	NA	NA	NA
27			Trunk	Congenital	15 years	Excision	NA	NA	NA
28			Neck	Congenital	2 years	Wide resection	3 cm	NA	NA
29			Trunk	Congenital	10 months	Limited excision	NA	None	6 months
30	Toper	2008	Trunk	Congenital	26 years	Wide resection	3 cm	None	3 years
31	Jafarian	2008	Lower limb	Congenital	14 years	Wide resection	NA	None	NA
32			Lower limb	Congenital	3 Months	Wide resection	NA	None	NA
33			Trunk	Congenital	3 Months	Wide resection	NA	None	NA
34			Trunk	3 months	7 years	Wide resection	NA	None	NA
35	Lee	2008	Trunk	Congenital	9 months	Wide resection	NA	None	6 months
36	Gerlini	2008	Lower limb	Congenital	12 years	Wide resection	2 cm	None	4 years
37	Feramisco	2008	Trunk	Congenital	7 months	Mohs surgery	NA	None	11 months
38	Marque	2008	Upper limb	2 months	1 year 6 months	Wide resection	NA	None	NA
39			Lower limb	Congenital	8 years	Wide resection	3.5 cm	None	NA
40	Moumine	2008	Head	Congenital	2 years	Wide resection	3 cm	None	5 years
41	Reddy	2009	Trunk	Congenital	5 years	Wide resection	3 cm	None	1 year 6 months
42	Kim	2009	Trunk	Congenital	12 years	Wide resection	NA	None	8 months
43	Love	2009	Trunk	Congenital	10 months	Mohs surgery	1 cm	None	1 year
44	Suzuki	2011	Trunk	Congenital	8 months	Mohs surgery + imatinib	NA	None	5 years
45	Blaser	2011	Upper limb	Congenital	4 months	Mohs surgery	NA	NA	NA
46	Goyal	2012	Head	6 months	NA	NA	NA	NA	NA

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Table 1 (continued)

Case number	Author	Published year	Location	Age at onset	Age at diagnosis	Treatment	Surgical margin	Recurrence	Follow-up duration
47	Morais	2012	Trunk	Congenital	5 years	Mohs surgery	NA	None	NA
48	Tantcheva-Poor	2012	Trunk	Congenital	11 months	Wide resection	3 cm	None	1 year 6 months
49	Iwata	2013	Trunk	Congenital	11 years	Wide resection	NA	NA	NA
50	Han	2015	Neck	Congenital	6 years	Wide resection	3 cm	None	4 years
51	Gualdi	2015	Lower limb	5 months	5 years	Wide resection	3 cm	None	2 years
52	Caroppo	2020	Trunk	Congenital	2 years	Wide resection	3 cm	None	1 year 6 months

NA, not available.

surgical margin for lesions after unplanned excision is even more difficult than determining the margin size for the primary tumors. In a retrospective review of 35 adult cases of DFSP at a single institution, including patients who developed local recurrence after unplanned excision, no recurrence was reported in 28 patients who underwent additional wide resection, with a mean histological margin of 2.1 cm. However, three of the remaining 7 patients who underwent marginal or intralesional resection developed recurrent tumors [11]. Another single-center retrospective review of 35 adult cases of DFSP who underwent wide resection, including a local recurrence group, reported that the mean histological margin in the recurrence-free cases was 2.5 cm for the primary cases and 3.3 cm for the recurrent cases [7]. Regarding pediatric cases after unplanned excision, the evidence is so limited that the pediatric patients should be treated as for adult cases. In previous reports, no local recurrence was reported after wide

resection; however, the details of the surgical margins were unclear (Table 1). Therefore, in our case, the margin was set at 3 cm in accordance with the recommendation for adult cases. The skin coverage in our case was concerning after the wide 3 cm resection; however, fortunately, a three-point suture closure was successfully achieved without a skin graft or local skin flap.

In conclusion, malignant tumors, as well as DFSP, should be suspected and confirmed by a biopsy when encountering an enlarging superficial tumor of the trunk in infants. In the current case, after unplanned resection, the margin during the additional wide excision was set at 3 cm, and the patient's postoperative course has been uneventful to date, without recurrence.

Statement of Ethics

This study protocol was reviewed and approved by the Keio University School of Medicine IRB, approval number 20160298. Written informed consent was obtained from the parent of the patient for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Tomoharu Tanaka, Robert Nakayama, Tomoaki Mori, Naofumi Asano, Noriko Aramaki Hattori, Masaya Nakamura, and Morio Matsumoto performed the collection and assembly of the data. Tomoharu Tanaka performed the data analysis and prepared the initial draft of the manuscript. Tomoharu Tanaka and Robert Nakayama contributed to the writing of the manuscript. Robert Nakayama provided final approval of the manuscript. All authors have read and approved the final manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material. Further inquiries can be directed to the corresponding author.

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