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CASE REPORT

Dermatofibrosarcoma Protuberans in a 12-Year-Old Child: A Rare Case

Eva Krishna Sutedja[1], Endang Sutedja[1], Kartika Ruchiatan[1], Yogi Faldian[1], Yuri Yogya[1], Risa Miliawati Nurul Hidayah[1], Rafithia Anandita[1], Yohana Azhar[2], Anglita Yantisetiasti[3], Bethy Suryawathy Hernowo[3], Yovan Rivanzah[1]

[1]Department of Dermatology and Venereology, Faculty of Medicine, Universitas Padjadjaran - Dr. Hasan Sadikin Hospital, Bandung, West Java, Indonesia; [2]Department of Surgery, Faculty of Medicine, Universitas Padjadjaran – Dr. Hasan Sadikin General Hospital, Bandung, Indonesia; [3]Department of Pathological Anatomy, Faculty of Medicine, Universitas Padjadjaran – Dr. Hasan Sadikin General Hospital, Bandung, Indonesia

Correspondence: Eva Krishna Sutedja, Department of Dermatology and Venereology, Faculty of Medicine, Universitas Padjadjaran - Dr. Hasan Sadikin Hospital, Jl. Pasteur 38, Bandung, West Java, 40161, Indonesia, Tel +628122014300 ext. 3449, Fax +62222032426, Email eva.krishna@unpad.ac.id

Abstract: Dermatofibrosarcoma protuberans (DFSP) is an uncommon soft tissue tumor involving the dermis and subcutaneous fat that rarely occurs in children, manifested as a slowly growing firm plaque on the trunk. A 12-year-old girl patient presented with dark patch on the nasal root after finishing 25 sessions of radiotherapy. Initially, patient came to Oncology Surgery Clinic at Hasan Sadikin General Hospital Bandung with the chief complaint of a large exophytic mass located in the nasal area, which was neither itchy nor painful. A large, firm, painless mass with no sign of localized heat or redness was found on physical examination. There were no palpable cervical or axillary lymph nodes. Wide local excision and frontal flap procedure were performed by Oncology Surgery Department leaving a pedicle with 2×1.5×1 cm on size was observed. Upon histopathological examination, tumor mass was found in the subepithelium and consisted of oval to spindle-shaped cells that were hyperplastic, compacted, diffuse, forming fasciculus, whorled, and cartwheel. Cell nuclei were pleomorphic (oval to wavy), hyperchromatic, with clear nucleolus, and occasion mitotic figures. Hyalinisation was seen between the tumor masses. On immunohistochemical stains, there were diffuse positivity for epithelial membrane antigen (EMA) and vimentin. Based on the histological and immunohistochemical findings, the diagnosis of stage II DFSP was made. Until now, there is no established algorithm for treatment of DFSP. Wide local excision and radiotherapy for 25 sessions was performed on this patient, resulting in complete tumor mass removal. After three months of observation, the second surgery was done to remove a pedicle; however, there is no recurrence of tumor growth. Despite its rarity, DFSP should be considered as a differential diagnosis to avoid underdiagnosis or misdiagnosis.

Keywords: dermatofibrosarcoma protuberans, radiotherapy, wide local excision

Introduction

Dermatofibrosarcoma protuberans (DFSP) is an uncommon soft tissue tumor that involves the dermis, subcutaneous fat, and in rare cases, muscle and fascia.[1,2] It accounts for 1–6% of all soft tissue sarcomas and 18% of all cutaneous soft tissue sarcomas. DFSP in children, although rare, is reported in some countries, including congenital presentations.[2] According to Ramos and Hernanz in 2012,[3] only 6% of these tumors present in children, while according Loeb et al in 2008,[4] the prevalence of DFSP before 20 years of age is 1.0 per million. It typically presents as a slowly growing,[5] firm plaque on the trunk of young adults.[2,3,5]

The cause is not clearly understood yet, however, some studies have implicated a chromosomal translocation that results in a fusion protein COL1A1-PDGFB that promotes tumor growth through the overproduction of platelet-derived growth factor (PDGF).[1,6] Diagnosis is made through skin biopsy. It is considered an intermediate-grade malignancy with a low likelihood of metastasis but a high local recurrence rate.[2,5] Molecular detection of the gene rearrangement or fusion transcripts is helpful for the diagnosis of patients with atypical morphology and for screening candidates for targeted therapy with tyrosine kinase inhibitors.[5]

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The standard treatment is complete surgical excision, including wide local excision (WLE) with tumor-free margins, Mohs micrographic surgery (MMS) and amputation.[5] Given its propensity for a subclinical extension, the optimal treatment modality is MMS, a surgical technique that allows complete margin assessment and tissue preservation. The goal of the therapy for DFSP is to reach a wide and clear resection margin of 2–3 cm to reduce the local recurrence rate.[7,8] Unresectable DFSPs are treated with radiation therapy and/or targeted therapy. The chemotherapeutic agent imatinib mesylate is currently Food and Drug Administration (FDA)-approved for adults with unresectable, recurrent, or metastatic DFSP.[1,2,9,10]

The overall prognosis is good with a 10-year survival rate of 99.1%. However, there is still a possibility of complication with metastasis being the main one. Other complications including post-surgical scarring and cosmetic defect.[2] Hereby, we present a rare case of DFSP in children with clinical manifestation of tumor mass located in nasal root, which is an uncommon predilection of DFSP.

Case Illustration

A 12-year-old girl consulted to the Dermatology and Venereology Department due to the presence of dark patch on the nasal root after finishing 25 sessions of radiotherapy (Figure 1). The patient was initially presented to the Oncology Surgery Clinic at Hasan Sadikin General Hospital Bandung with the chief complaint of a large exophytic mass located in the nasal area, which was neither itchy nor painful (Figure 2A). The skin lesion first appeared 5 years ago as a small, irregularly shaped, pus-filled bump on nasal root that was painful. The lesion was thought to be a boil, therefore her teacher manipulated the lesion until it bled. Since then, the lesion has progressively increased in size. The patient denied any recent weight loss, fever, night sweats or chills. Previous medical history was insignificant. Family history of malignancy was positive.

In August 2023, the patient underwent surgical excision combined with frontal flap performed by the Department of Surgery, Oncology Surgery Division at Hasan Sadikin General Hospital Bandung (Figure 2B–D). A mass measuring 4×1.5×0.5 cm was excised with diagnostic frozen section examination showing suspected malignant

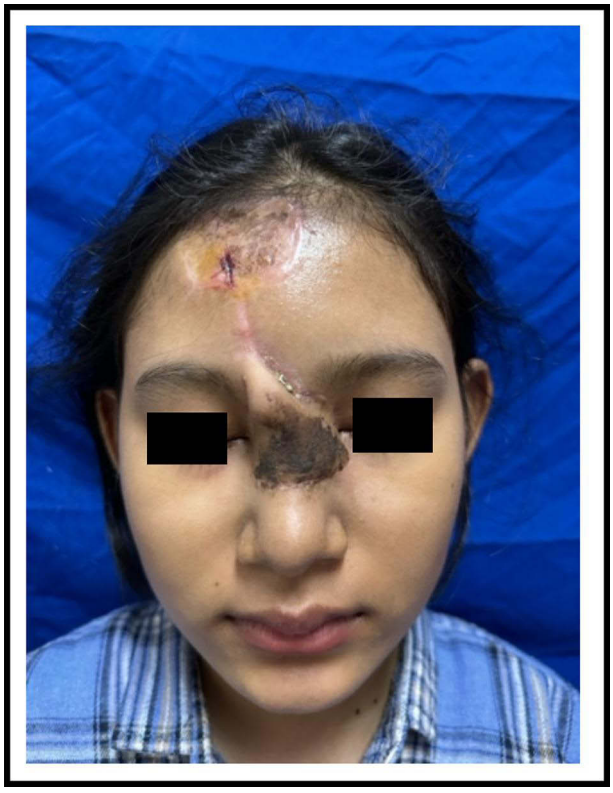

Figure 1 Lesion when first consulted to Dermatology and Venereology Department (A).

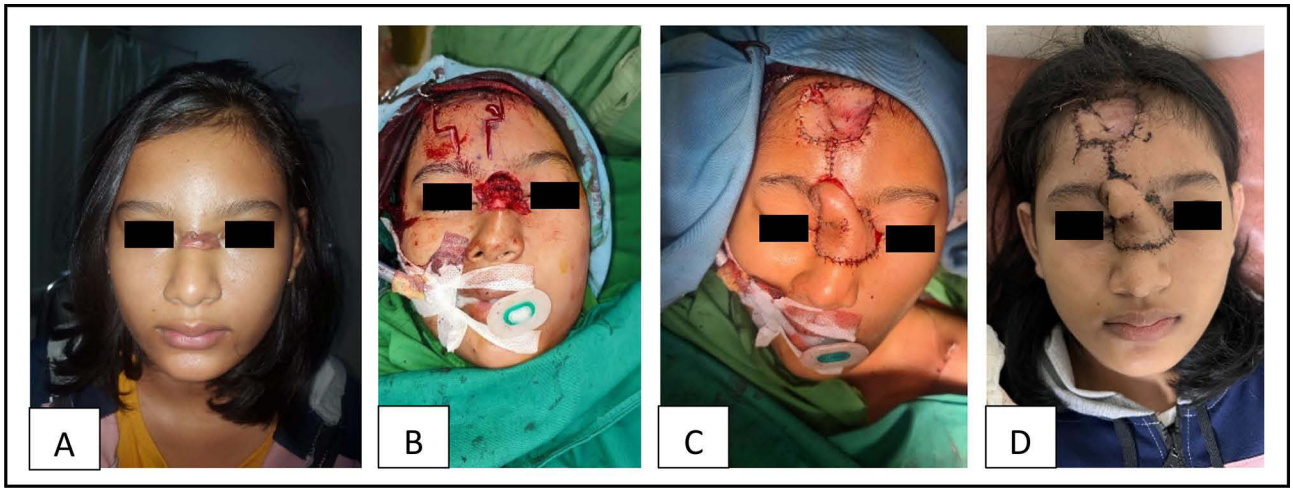

Figure 2 Lesion during the first visit to the Oncology Surgery Department (**A**), lesion during wide excision operation (**B** and **C**), and postoperative lesion (**D**).

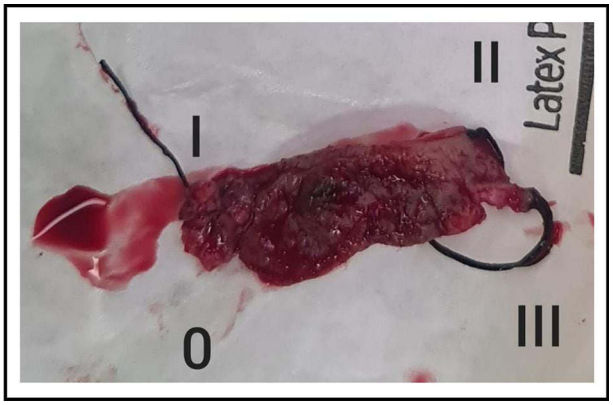

Figure 3 Intraoperative lesion.

lesion (Figure 3). A pedicle was observed after the surgery. The safety margin could not be achieved because the lesion was too close to ocular structures. Therefore, adjunctive radiotherapies were performed for 25 times and afterwards the dark patch appeared on the nasal root. At follow-up 3 months after surgery, a pedicle structure was observed on the nasal root area. A second surgery was performed by Department of Surgery, Oncology Surgery Division, to excise the pedicle (Figure 4A and B).

On physical examination, a large, firm, painless mass was found with no sign of localized heat or redness. There were no palpable cervical or axillary lymph nodes. A pedicle in the size of 2×1.5×1 cm was observed. Upon histopathological examination, tumor mass was found in the subepithelium and consisted of oval to spindle-shaped cells that were hyperplastic, compacted, diffuse, forming fasciculus, whorled, and cartwheel. Cell nuclei were pleomorphic (oval to wavy), hyperchromatic, with clear nucleolus, and occasion mitotic figures. Hyalinisation was seen between the tumor masses (Figure 5A–D). On immunohistochemical stains, there were diffuse positivity for EMA (Figure 6A) and vimentin (Figure 6B). There was no positivity for cluster of differentiation (CD)34 (Figure 6C) and cytokeratins (CK) (Figure 6D). Based on the histological and immunohistochemical findings, the diagnosis of stage II DFSP was made.

The patient's postoperative course after the pedicle excision was uneventful, and she was discharged on the second postoperative day (Figure 7A–C).

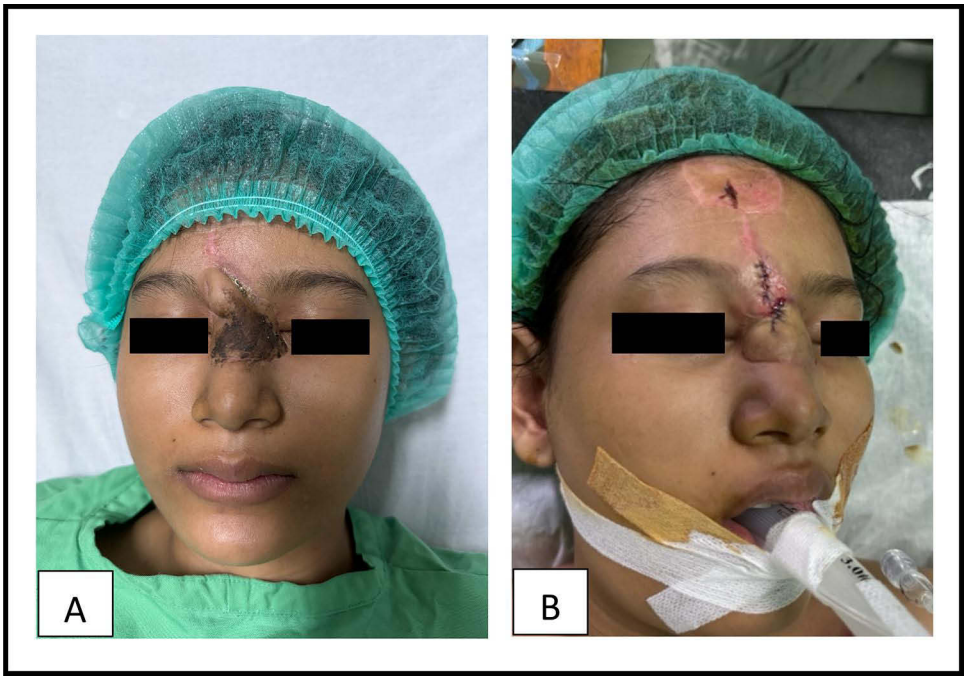

Figure 4 Lesion before pedicle excision procedure (**A**) and lesion after pedicle excision procedure (**B**).

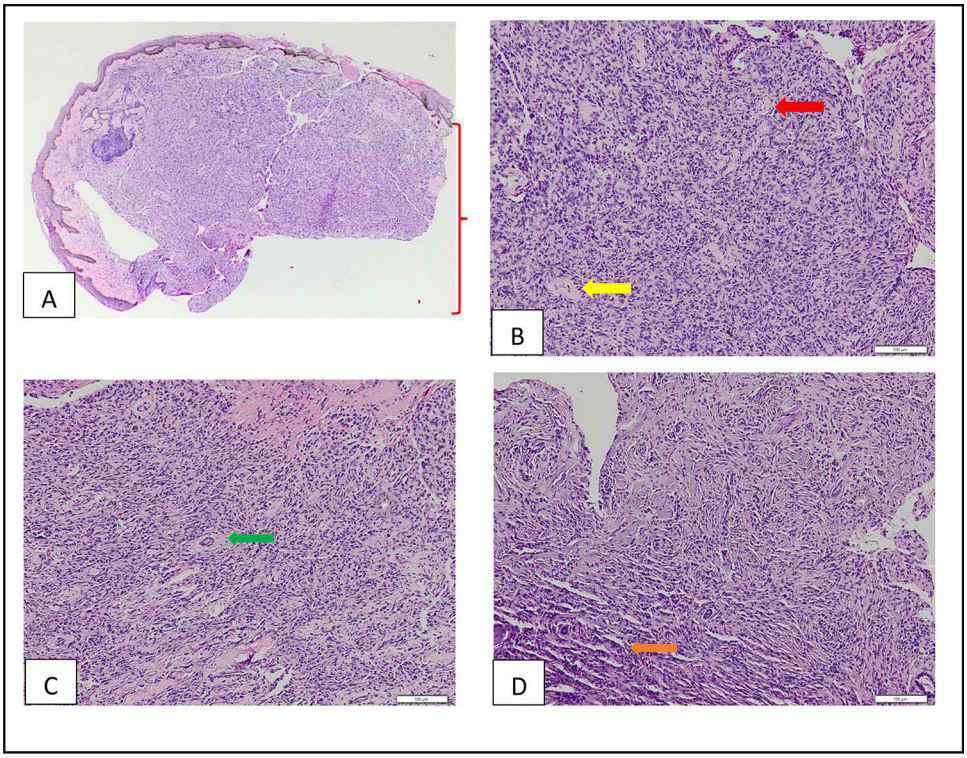

Figure 5 Histopathologic examination of DFSP consist tumor cell (**A**-red line), hyalinization (**B**-yellow arrow), cartwheel (**B**-red arrow), whorled pattern (**C**-green arrow), and fasciculus pattern (**D**-Orange arrow).

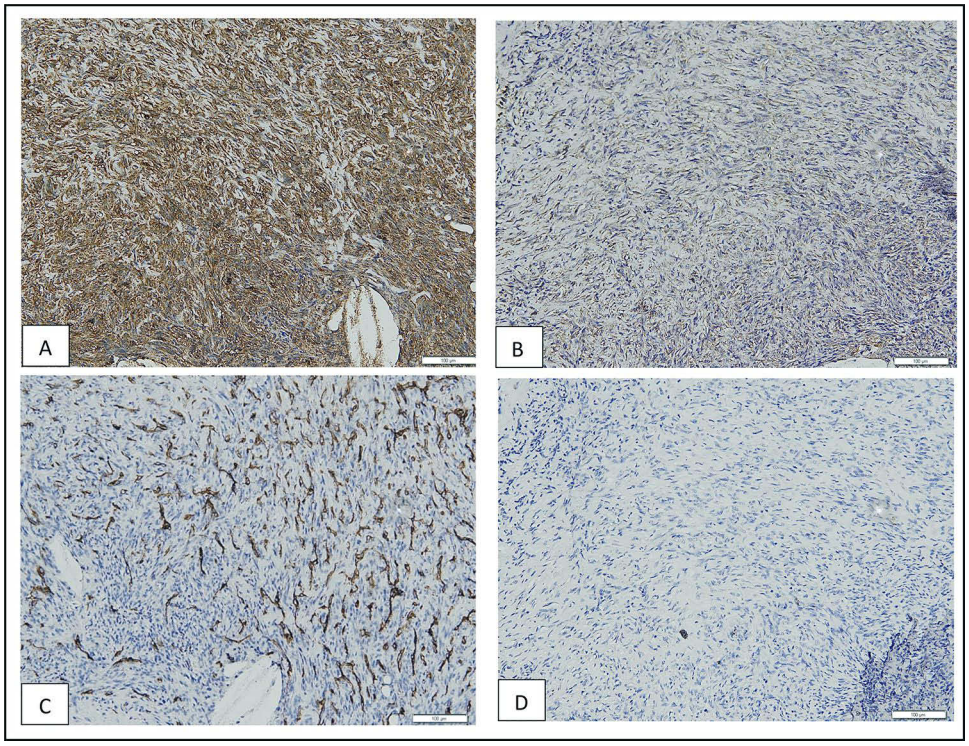

Figure 6 Immunohistochemistry staining EMA strongly positive (**A**), vimentin positive in tumor cell (**B**), CD34 negative (**C**), and CK negative in tumor cell (**D**).

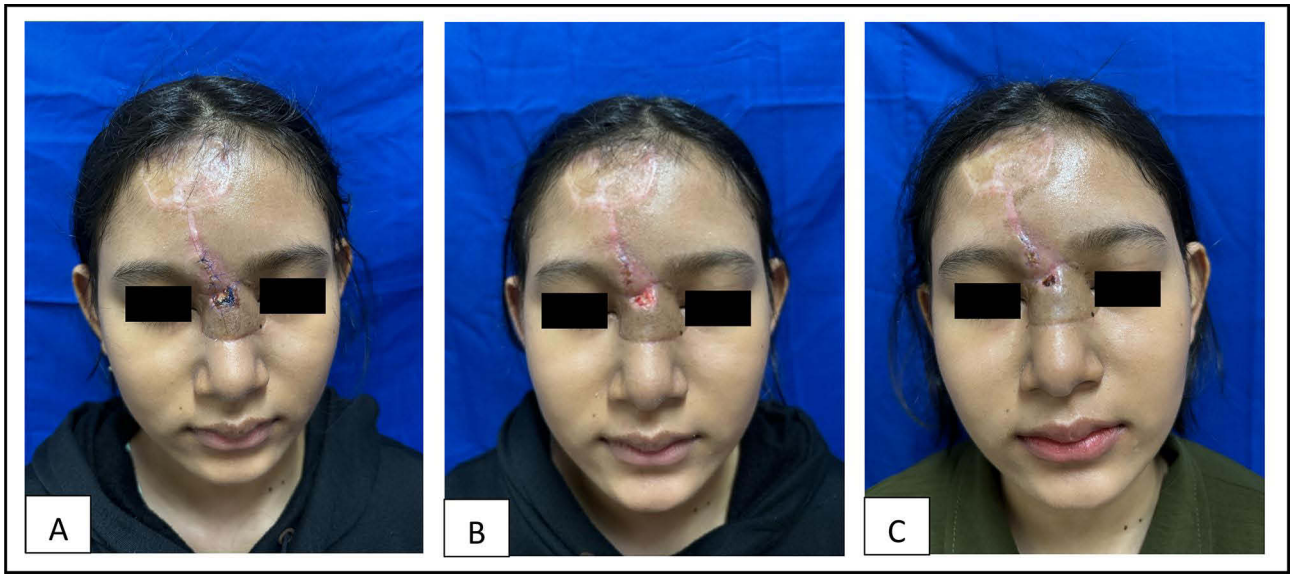

Figure 7 Post procedure lesion day 11 (**A**), day 18 (**B**), and day 23 (**C**).

Discussion

Dermatofibrosarcoma protuberans is a rare tumor affecting the dermis, subcutaneous fat, and occasionally muscle and fascia.[1,2] Typically, it presents as a slow growing, firm plaque on the trunk of young adults.[2,9] Incidence of this tumor is approximately 4.2 per 1 million in the United States.[1,2] DFSP occurs most often in middle-aged adults, although it has been reported in all age groups, including congenital presentations.[2,5] Fibrosarcomatous (FS) change is observed on histopathology in 3 to 20% of patients with DFSP.[7] FS is more often encountered in patient with large sized tumors

which is more malignant.[1] Furthermore, tumors with this feature have approximately double the chance of distant metastatic spread as well as a lower overall survival.[7]

The cause of DFSP is not clearly understood yet, however, some studies have implicated translocation of chromosomes 17 and 22 [t(17:22)(q22:q13)] is seen in over 90% of DFSP. The translocation then results in a fusion protein COL1A1-PDGFB that promotes tumor growth through the overproduction of PDGF, continuous autocrine activation of platelet-derived growth factor receptor-beta (PDGFRb), cellular proliferation, and tumor formation.[1,2,6]

Dermatofibrosarcoma protuberans initially manifests as an asymptomatic plaque, initially skin-colored in early stage and later transitioning to red-brown induration, eventually giving rise to multiple elevated violaceous to red-brown nodules in protuberant stage.[2,5] Progressing slowly, these nodules can grow to several centimeters in diameter.[2] DFSP is predominantly found on the trunk (50%), followed by the extremities (35%), and head and neck (15%). DFSP exhibits a characteristic predilection for the shoulder and pelvic regions. Although typically extending into subcutaneous fat, it rarely involves fascia, muscle, or bone unless it persists or recurs over time. If left untreated, the tumors can locally invade more deeply into the fascia, muscle, periosteum, bone, and occasionally metastasize to other organs in advanced stages.[11]

The clinical presentation of DFSP in childhood is similar to that in adults and varies with the stage of growth. In the early stages, lesions typically appear as single papules or plaques, with deep-seated nodules being less common.[12] One of the most consistent features of this tumor is induration.[3] The lesion usually moves freely over deep tissue structures until the late stages of the disease, when it begins to invade the underlying structures.[13] The overlying skin can appear erythematous, brownish, violaceous, or flesh-colored. A bluish tinge may be the initial manifestation of the tumor, potentially leading to an incorrect initial diagnosis of a vascular lesion.[14] Generally, these lesions are asymptomatic and measure between 1 and 5 cm at the time of diagnosis. They tend to grow progressively.[15]

There are various histologic variants of DFSP have been identified, including atrophic and FS.[16] The atrophic variant presents as a violaceous plaque, resembling morphea or scar. In its early stages, the tumor may resemble a keloid or dermatofibroma, leading to frequent misdiagnoses. With increased size, some may ulcerate and become painful. DFSP displays relentless growth with asymmetrical, root-like projections, not discernible in clinical examinations, leading to common local recurrences after excision. Distant metastasis is infrequent, occurring in only 1% to 4% of cases, usually following multiple local recurrences, with the lung being the primary site of metastasis via hematogenous spread. Regional lymph node involvement is rare. The FS variant of DFSP possesses a higher risk of local recurrence (14 to 52%) and distant metastases (8 to 29%). It is considered an intermediate-grade malignancy with a low likelihood of metastasis, but a high local recurrence rate.[2]

Diagnosis is made through skin biopsy, preferably an incisional or excisional biopsy. A thorough history and physical exam, including lymph node examination, should be done. Some supporting examinations, such as chest X-ray and preoperative magnetic resonance imaging (MRI), are sometimes performed to evaluate for any metastatic disease and to define tumor extension before surgery.[17–20] DFSP typically exhibits poor circumscription, commonly affecting both the dermis and subcutaneous tissues, with rare cases limited solely to the dermis. Positioned as an intermediate tumor between a benign dermatofibroma and a fibrosarcoma, DFSP carries the potential for distant metastasis and aggressive local invasion. While the overlying epidermis lacks atypical histological features, the skin may present a macroscopic appearance that is greyish or lightly pigmented.[2]

Histopathological stand point of DFSP is uniform spindle cell fascicles exhibiting a storiform pattern, along with several variants and robust, widespread CD34 immunoreactivity. Nonetheless, its spindle cell morphology and CD34 immunostaining pattern exhibit similarities with other both benign and malignant lesions, requiring careful differentiation. On an ultrastructural level, DFSP is characterized by stellate or spindle cells featuring long, slender, ramified cell processes connected by primitive junctions reminiscent of dermal dendrocytes.[5,21] However, areas of FS change may show reduction or loss of CD34 expression immunohistochemically.[22]

There is no standard staging system of DFSP available up to today.[23] Hao et al[5] then proposed a modified staging system of DFSP based on European consensus-based interdisciplinary guideline, the progression of DFSPs' tumorigenesis and clinical presentation, as shown in Table 1. This staging system is useful for treatment.[5]

Table 1 Staging System of DFSP

Stage	Criteria
Stage I	Non-protuberant lesions including atrophic or sclerotic plaque, macula or small nodules
Stage II	Protuberant primary tumor
Stage IIA	Superficial tumor: without invasion of the underlying fascia
Stage IIB	Deep tumor: either superficial to the fascia with infiltrating the fascia or occurred beneath the superficial fascia
Stage III	Lymph node metastasis
Stage IV	Distant metastasis to other organs

Notes: Reprinted from Hao X, Billings SD, Wu F, Stultz TW, Procop GW, Mirkin G, Vidimos AT. Dermatofibrosarcoma protuberans: Update on the diagnosis and treatment. *Journal of Clinical Medicine*. 2020; 9(6):1752. Creative Commons.[5]

The standard treatment is complete surgical excision, including wide local excision (WLE) with tumor-free margins, Mohs micrographic surgery (MMS) and rarely amputation.[5] Given its propensity for a subclinical extension, the optimal treatment modality is MMS, a surgical technique that allows complete margin assessment and tissue preservation. The goal of the therapy for DFSP is to reach a wide and clear resection margin of 2–3 cm to reduce the local recurrence rate.[7,8] Wide undermining following surgical excision of DFSP is discouraged due to the risk of tumor seeding and difficulties in interpreting subsequent reexcisions.[24]

Multiple studies have shown that MMS significantly reduces the risk of recurrence of DFSP, compared with WLE.[25,26] Notably, DFSP treated with WLE has a recurrence rate of around 7.3%, in contrast to the 1% recurrence rate seen with MMS.[26] Collaborative care, especially for infiltrative DFSP, enhances treatment outcomes. NCCN guidelines recommend lateral margins of 2–4 cm for WLE and excision of investing fascia for infiltrating tumors.[5] The choice of surgical procedure depends on tumor depth, location, and patient factors.[5,27]

Adjuvant radiation therapy is effective in controlling tumor growth and reducing recurrence, with high disease-free survival rates reported in studies.[28–32] The recommended radiation dose ranges from 60 Gy for indeterminate or microscopic positive margins to up to 70 Gy for macroscopic positive margins or primary gross tumors. The radiation field should extend 3–5 cm beyond surgical margins or the primary tumor boundary, with an individual dose administered at 2 Gy daily, 5 times weekly.[23,25,26,33–36]

The molecular identification of gene rearrangements or fusion transcripts proves advantageous not only for diagnosing cases lacking typical morphology but also for screening patients eligible for imatinib mesylate (IM) use.[5,37] The chemotherapeutic agent imatinib mesylate is currently FDA-approved for adults with unresectable, recurrent, or metastatic DFPS.[1,2,9,10] Imatinib mesylate inhibits the constitutively activated PDGF receptor β in DFSP, demonstrating growth-inhibitory effects in studies.[5] IM may also modulate immune responses, exerting direct antitumor effects on the PDGFR pathway and indirect effects via T-cell-mediated immune modulation. Effective in localized and metastatic DFSP cases, IM showed similar responses with daily doses of 400 mg or 800 mg.[38] Preoperative IM therapy resulted in median tumor volume shrinkages (20%–31.5%), making unresectable tumors resectable.[39–42] However, approximately 10% of DFSPs do not respond to IM, and some develop rapid secondary resistance. Surgical excision during the "shrinkage window" is crucial.[5] Resistance mechanisms are unclear, but low PDGFR phosphorylation in resistant tumors may be a factor. In cases of IM resistance, alternatives like sunitinib, sorafenib, and pazopanib can be considered, showing effectiveness in resistant DFSP patients.[43,44]

The overall prognosis is good with a 10-year survival rate of 99.1%. Given the infrequent occurrence of metastasis, the predominant concern lies in the morbidity resulting from local recurrence. Individuals aged over 50 meet an increased risk of local recurrence. On average, patients diagnosed with metastatic disease survive approximately two years. Risk factors associated with elevated mortality rates include a high mitotic index, heightened cellularity, belonging to the black race, male gender, and having the tumor located on the head, neck, or limb. However, there is still a possibility of complication with metastasize being the main one.[2] Other complications center around post-surgical scarring and cosmesis.[2]

Conclusion

Dermatofibrosarcoma protuberans is an uncommon soft tissue tumor involving the dermis and subcutaneous fat that rarely occurs in children, usually manifested as a slowly growing firm plaque on the trunk. However, in this case, the tumor mass was located in nasal root which is an uncommon predilection for DFSP. Until now, there is no established algorithm for treatment of DSFP. Nonetheless, the standard treatment includes complete surgical excision, such as WLE with tumor-free margins, MMS, and amputation. As a clinician, we must consider the diagnosis of DFSP because it tends to be underdiagnosed or misdiagnosed.

Ethic Statement

The publications of images were included in the patient's consent for publication of the case. Institutional approval has been obtained to publish the case details.

Consent Statement

The authors certify that they have obtained all appropriate patient consent forms from the parents of the patient as the patient is under the age of 18. The parents of the patient signed a consent form for the publication of the case details and images.

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Disclosure

The authors report no conflicts of interest in this work.

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