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CASE REPORT

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# Bifocal metachronous dermato fibrosarcoma protuberans in children: A case report

**Wajdi Arfa**[1,2] | **Leila Bouhajja**[2,3] | **Mohamed Ghammem**[1] | **Mohamed Amri**[1,2] | **Faten Farah**[2,3] | **Zied Jlalia**[1,2] | **Mourad Jenzri**[1,2]

[1]Mohammed Kassab National Institute of Orthopaedics, Pediatric orthopedic department Kssar Said, La Mannouba, Tunisia

[2]University of Tunis El Manar Faculty of Medicine of Tunis, Tunis, Tunisia

[3]Mohammed Kassab National Institute of Orthopaedics, Anatompathology department Kssar Said, La Mannouba, Tunisia

**Correspondence**
Wajdi Arfa, Mohammed Kassab National Institute of Orthopaedics, Pediatric orthopedic department Kssar Said, La Mannouba, Tunisia.
Email: wajdi.arfa@fmt.utm.tn

**Key Clinical Message**

Dermato fibrosarcoma protuberans presents challenges in diagnosis and treatment, especially in children. Awareness of its aggressive local recurrence and its potential for multifocal presentation is crucial. Wide surgical resection with adequate margins remains the basis of management, in association with regular follow-up of affected patients.

**Abstract**

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive cutaneous sarcoma, particularly uncommon in children. We report the case of a 13-year-old boy who initially presented with a firm mass on the left foot, later diagnosed as DFSP following histological and immunohistochemical analysis. The tumor was surgically excised with a wide margin, and a skin graft was used for coverage. Despite an initial favorable outcome, a new DFSP lesion developed at the proximal left thigh 1 year later, requiring further wide surgical excision and coverage with a tensor fascia lata flap. Both sites remained free of recurrence at one-year follow-up. This case underscores the importance of long-term vigilance in managing DFSP due to the potential risk of recurrence and multifocal involvement.

**KEYWORDS**

children, dermatofibrosarcoma, recurrence, surgical oncology

## 1 | INTRODUCTION

Dermato fibrosarcoma protuberans (DFSP) or Darier and Ferrand dermato fibrosarcoma is a rare malignant cutaneous mesenchymal tumor. It accounts for 1.8% of soft tissues sarcomas.[1] It is annual incidence is 4.2 per million.[2] It is known for its local aggressivity and high potential for recurrence after treatment.[3] Its occurrence in children is rare.[4] Multifocal forms are rarely reported in the literature.[5,6] In this case report, we described the evolution and the management of metachronous bifocal DFSP in children.

## 2 | CASE HISTORY

A 13-year-old boy with no previous medical history, living in a rural region consulted for a swelling of the left

foot that had been evolving for 2 years and had increased in size. There was no history of local trauma or fever. Physical examination revealed a firm oblong swelling on the dorsal side of the left foot measuring 18 × 15 cm, fixed to the cutaneous plane and non-painful (Figure 1A). The mobility of the toes was preserved.

## 3 | METHODS

X-ray of the foot found no bone abnormalities. Magnetic resonance imaging (MRI) showed a superficial, well-limited mass with a T1 hyposignal and heterogeneous T2 hypersignal, intensely enhanced by injection of Gadolinium (Figure 2). A surgical biopsy of the mass was performed. Histological examination revealed spindle cells proliferation arranged in a storiform pattern (Figure 3A), involving the dermis and infiltrating the hypodermis (Figure 3B). The spindle cells have abundant, eosinophilic cytoplasm with ovoid to elongated nuclei and rare mitotic activity (Figure 3C). Some diagnoses were suggested: DFSP, synovial sarcoma, muscular tumors, nervous tumors and melanoma. Immunohistochemistry showed that the spindle cells were diffusely immunoreactive for CD34 (Figure 3D). However, the tumor cells were negative for synovial sarcoma markers (CD99, CK, EMA, BCL2), muscular markers (Desmin, Caldesmon, Smooth Muscle Actin), nervous markers (PS100) and melanoma's marker (SOX10). The Ki-67 proliferative index was low. The molecular biology study showed the absence of the synovial sarcoma's specific translocation (t(X;18) (p11;q11)). So, we have eliminated the diagnosis of synovial sarcoma but the cytogenetic study of DFSP wasn't done. The diagnosis of classic DFSP was retained. The pigmentation or a sarcomatous transformation were not observed.

There were no similar family cases. A wide resection was performed, removing the tumor (Figure 4), the adjacent skin with a 3 cm margin and the dorsal cortices of the first three metatarsals. Axial pinning of the first 3 toes was performed to prevent claw deformity given that their extensors had been removed with the tumor (Figure 1B). A skin graft was used to cover the loss of substance.

## 4 | CONCLUSION AND RESULTS

The follow-up at 1 year showed a healing of the skin without local recurrence (Figure 1C). However, the patient developed a large, firm swelling at the proximal left thigh that had been evolving for 3 months (Figure 1D). MRI revealed a subcutaneous tumor, in contact with the femoral pedicle with similar signal as the initial foot tumor (Figure 5). Ultrasound-guided biopsy revealed a second metachronous DFSP localization. A wide resection was performed, removing the tumor and the surrounding skin with a 3 cm margin. At depth, the tumor was separated from the femoral pedicle through a cleavage plane removing the adjacent part of the femoral sheath. The skin loss in front of the femoral pedicle was covered by a tensor fascia lata flap (Figure 1E). At 1 year follow-up, neither recurrence at both localizations nor metastasis were noted (Figure 1C,F).

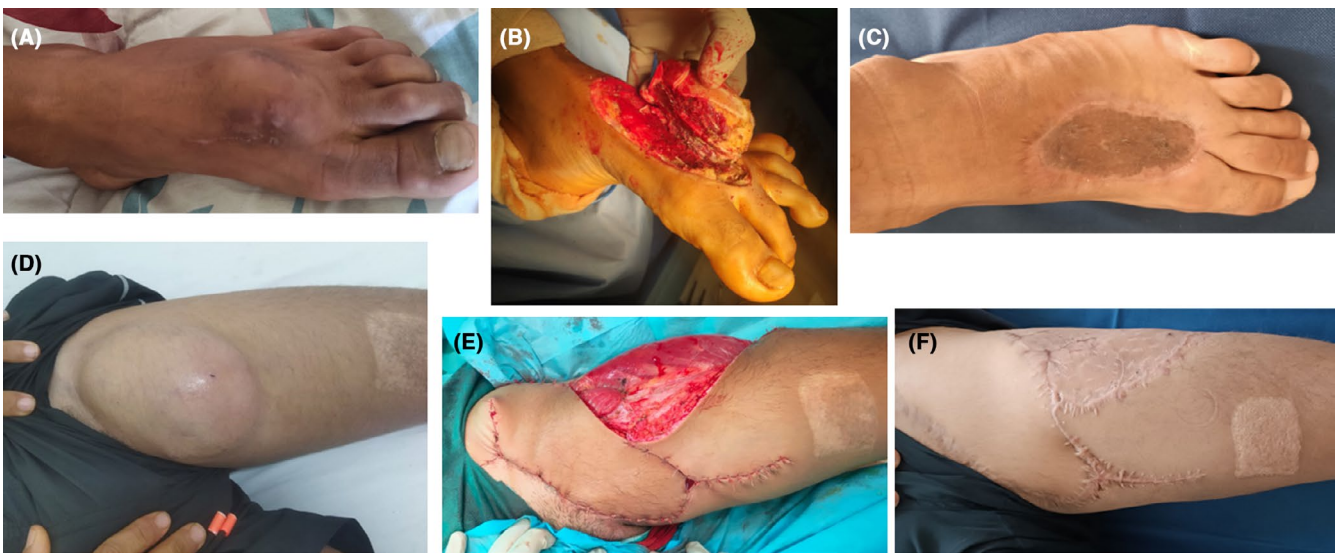


**FIGURE 1** Clinical images: (A) Appearance of the left foot tumor (B) Intraoperative view of the left foot tumor (C) One-year postoperative view of the left foot (D) Proximal left crural tumor (E) Coverage of the left crural tumor crater by the tensor fascia lata flap (per operative view) (F) One-year postoperative view of the proximal left crural region.

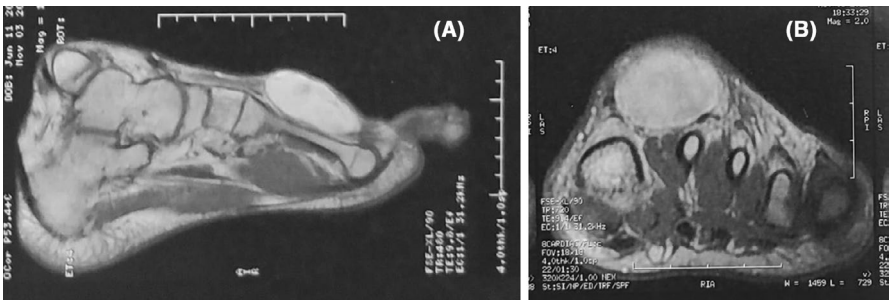


**FIGURE 2** MRI of the foot [(A): T2 sagittal section (B): T2 axial section] showing a well-limited superficial mass on the dorsal side of the left foot centered on the first intermetatarsal space, intensely enhanced after injection of gadolinium.

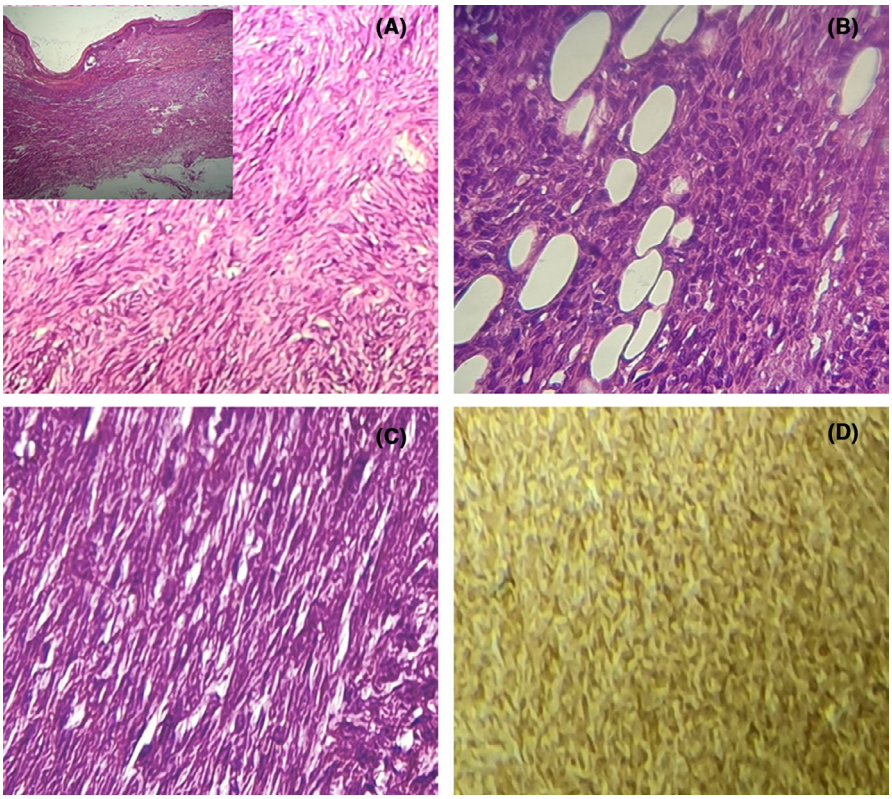


**FIGURE 3** (A) Spindle cells with a storiform patten (H&E; 100x) involving the dermis (inset, H&E; 40x) (B) Uniform and medium-sized spindle cells infiltrate the adipose tissue of the hypodermis (H&E; 400 x) (C) Spindle cells with eosinophilic cytoplasm and elongated nuclei (H&E; 400x) (D) Diffuse and strong cytoplasmic immunostaining in spindle cells for CD34 (CD34; 200x).

## 5 | DISCUSSION

Dermato fibrosarcoma protuberans (DFSP) or Darier and Ferrand dermato fibrosarcoma is a rare cutaneous mesenchymal tumor classified by the WHO as a low-grade sarcoma, locally aggressive but with low metastatic potential.[7] The average age of onset in the literature varies between 20 and 50 years.[8] Its occurrence in children is rare.[4] Its incidence is estimated at 1 per million in subjects under 20 years old.[9] DFSP can manifest in early infancy, including in newborns, but it is more commonly diagnosed in older children and adolescents.[10] Most frequent localizations for this tumor are the trunk, proximal extremities, the head and the neck region.[11] Clinically, DFSP often presents as an induration that progressively spreads to form a firm, uni or multinodular mass in few months.[11] The surrounding skin may be normal, or ulcerated.[11] Tumors can reach enormous dimensions, up to 25 cm in diameter.[12] In children, DFSP often presents as a slow-growing, indurated nodule or plaque, frequently misdiagnosed as benign lesions such as hemangiomas or hypertrophic scars.[10] Histologically, DFSP presents as a storiform proliferation of spindle cells in the deep dermis and infiltrates the hypodermis with a characteristic "honeycomb" pattern.[11] DFSP rarely involves the fascia, muscle, or bone. The epidermis overlying DFSP is thin and it is separated from the tumor by a Grenz zone. Spindle cells have elongated nuclei with little pleomorphism, and mitotic activity is variable.[1,13] Immunohistochemical examination shows a diffuse staining of tumor cells for CD 34, and negative for the following markers: PS 100, SMA, Desmin, keratins and Melan A.[14,15] On cytogenetic study, 90% of DFSPs express the specific translocation t (17;22) (q22;q13) resulting in COL1A1–PDGFB fusion.[16] The presence of this translocation is specific to DFSP

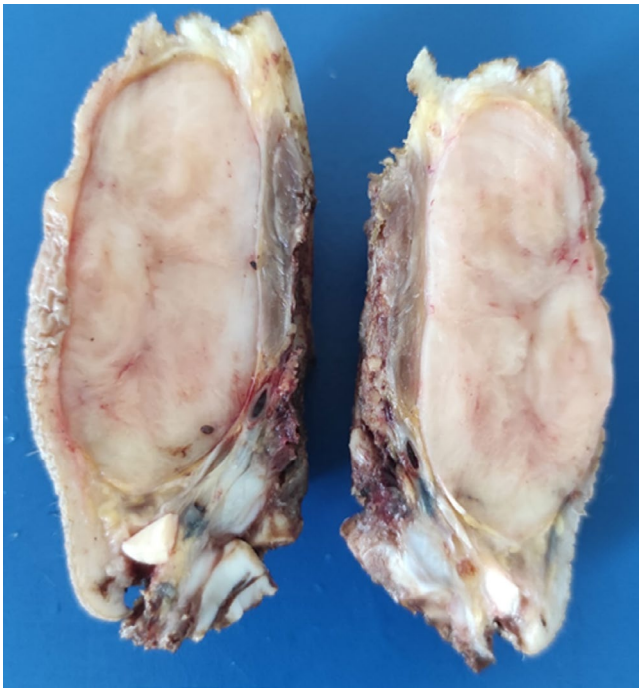

FIGURE 4 Longitudinal section of the foot tumor showing a well-circumscribed nodule with a grayish appearance and firm consistency.

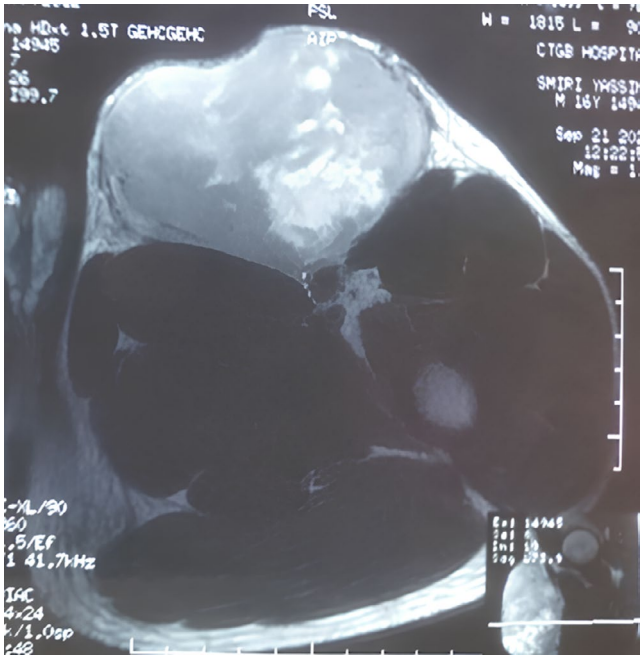

FIGURE 5 Axial section of the MRI of the left thigh, showing a well-limited subcutaneous mass on the anterior side of the proximal thigh, intensely enhanced after gadolinium injection. It displaces the anterior thigh muscles and comes into contact with the femoral vascular pedicle.

and helpful for the diagnosis.[16] The most common differential diagnoses of DFSP are dermatofibroma, benign fibrous histiocytoma, neurofibroma, solitary fibrous tumor, synovial sarcoma, melanoma, peripheral nerve sheath tumor, sarcomatoid carcinoma, and spindle cell lipoma, hemangioma (in children), giant cell fibroblastoma, pilomatrixoma.[17] The most important variants of DFSP include the Bednar tumor which contain melanin in dendritic cells and myxoid DFSP. The fibrosarcomatous transformation is present in 10–15% of all DFSPs.[18] Multifocal forms, as in our case are exceptional.[5,6] Deo et al. reported a case of bifocal DFSP: abdominal and mandibular localization.[19] This suggests that a complete physical examination should be carried out in patients with DFSP to detect other potential localizations. The particularity of this tumor is its deep cell extensions into the hypodermis, beyond the palpable and/or visible limits of the tumor, which explains the frequency of recurrence despite wide resection.[20] In children it can be more aggressive due to its potential for growth within developing tissues, despite its low metastatic potential.[10] The reference treatment is wide surgical resection with a safety margin that varies between 3 and 5 cm depending on the study, with sacrifice of a healthy anatomic barrier at depth.[21,22] In our case, to manage the resection margins in depth, the dorsal cortices of the metatarsals were resected for the foot tumor, and the adjacent portion of the femoral sheath was excised for the thigh tumor. The covering of skin loss caused by resection may be managed by reconstructive plastic surgery, through skin grafting or flaps. Mohs micrographic surgery (MMS) is an alternative to wide local excision, particularly for tumors located in delicate areas such as the head, the neck or orifices.[23] It was not used in our case, as the affected areas were the foot and thigh. It involves the resection of a thin margin of tissue around and deep inside the clinical tumor, which is examined extemporaneously. The process is repeated until the tumor presents healthy margins.[20] MMS thus enables precise microscopic control of the entire tumor margin, while preserving as much healthy tissue as possible.[24] Due to the rarity of cases in children, there are no pediatric-specific treatment guidelines, and the treatment is generally modeled after adult protocols.[25] The rate of local recurrence after wide resection reached 63.6% in the literature.[1] The risk of local recurrence (occurs in 20–50% of cases) depends on the quality of excision and the state of the margins. Re-excision is recommended if the surgical margin is positive.[26] The percentage of patients who develop metastases varies between 0 and 9.1%, with predominance of pulmonary metastases.[1] As regards adjuvant treatment, imatinib, a protein tyrosine kinase inhibitor, has been approved for adults with unresectable, recurrent and/or metastatic DFSP presenting the translocation t(17, 22) (q22,q13).[27] Post-operative radiotherapy may

be recommended in the event of local recurrence, or in the case of localization making wide excision surgery impossible or mutilating.[28]

DFSP is characterized by its slow local evolution and local recurrence with rare metastases. Multifocal localizations can occur. Wide surgical resection is the rule. Skin loss after resection can be managed using reconstructive surgery techniques.

## AUTHOR CONTRIBUTIONS

**Wajdi Arfa:** Writing – original draft. **Leila Bouhajja:** Writing – original draft. **Mohamed Ghammem:** Data curation. **Mohamed Amri:** Conceptualization. **Faten Farah:** Validation. **Zied Jlalia:** Conceptualization. **Mourad Jenzri:** Supervision; validation.

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## CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest.

## DATA AVAILABILITY STATEMENT

Raw data that support the findings of this case report are available from the corresponding author, upon a reasonable request.

## ETHICS STATEMENT

Participation was voluntary, confidential and not remunerated.

## CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

## ORCID

*Wajdi Arfa* https://orcid.org/0009-0005-0047-6660

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