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CASE REPORT
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A Rare Case of Dermatofibrosarcoma in a Pediatric Patient

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Summary: Dermatofibrosarcoma protuberans (DFSP) is a rare malignant fibroblastic tumor. DFSP has an insidious onset, slow growth, and heterogeneous presentation that can create a delay in diagnosis and increase morbidity. In this case report, we present a child with DFSP that presented as a large, slow-growing mass over the dorsum of the left foot. She underwent successful surgical excision with no functional sequelae. *(Plast Reconstr Surg Glob Open 2024; 12:e5546; doi: 10.1097/GOX.0000000000005546; Published online 23 January 2024.)*

Dermatofibrosarcoma protuberans (DFSP) is a rare malignant tumor of fibroblast origin. The reported incidence of DFSP in children is less than one case per million per year, typically occurring in adolescents and individuals of African descent.[1–3] This sarcoma often presents as a slow-growing, firm, and painful mass on the trunk or extremity, but the location and clinical manifestations can be variable.[4] There is a paucity in the literature regarding the presentation, diagnosis, and treatment of DFSP in the pediatric population. We present a case of a child with DFSP of the foot that had an atypical and delayed presentation.

CASE SUMMARY

An 11-year-old girl was presented to our clinic with a large, slow-growing mass on the dorsum of her left foot, which was first noticed 4.5 years previously. The family attributed this to intermittent trauma to this area during gymnastics classes and did not seek care for more than 2 years until the size of the mass began causing some difficulty with shoe wear and activities. At that time, the patient was seen in the emergency department and had a 4.5 cm × 4.0 cm firm, raised mass over the dorsal left foot. She had no functional deficit, was ambulatory, and had little discomfort related to the lesion. An ultrasound at that time noted a superficial, vascularized soft tissue mass. She was referred to a specialty clinic but was lost to follow-up.

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Two years later, physical examination at our clinic revealed a much larger mass, measuring 7×9 cm, with no overlying skin changes. The patient denied any pain, impaired mobility, or any issues with footwear, except that she could not secure her shoelaces on that side due to the size of the mass. Sensation and function of the foot and toes were intact (Fig. 1). Magnetic resonance imaging (MRI) revealed a T1 hypointense and T2 hyperintense 7.5×3.3×6 cm subcutaneous lesion with no underlying bone edema or invasion to adjacent tissues (Fig. 2). Differential diagnoses included a myofibroblastic, fibrohistiocytic, or synovial tumor.

Surgical excision was performed under tourniquet control and through a dorsal incision directly over the mass. The lesion was immediately below the skin. It was encapsulated and surprisingly easy to separate from the surrounding subcutaneous tissue using finger dissection alone. (**See figure, Supplemental Digital Content 1**, which displays the intraoperative image showing complete removal of an encapsulated DFSP, **http://links.lww.com/PRSGO/D33**.) The entire mass (12×8 cm) was removed and sent for histopathology. Significant skin redundancy required contouring during closure, and the incision was dressed with a compressive dressing and a short leg non–weight-bearing cast, which was removed 3 weeks later. There were no complications during or immediately after surgery (Fig. 3).

Histopathology revealed a cellular low-grade spindle cell proliferation involving the dermis and subcutaneous fat. The tumor exhibited spindled cells in a whirled pattern with some mitotic activity and scattered dendritic melanocytic cells (Fig. 4). Immunohistochemistry stained positive for vimentin and CD34. The stains were negative for desmin, migenin, pankeratin, and S100 protein.

Disclosure statements are at the end of this article, following the correspondence information.

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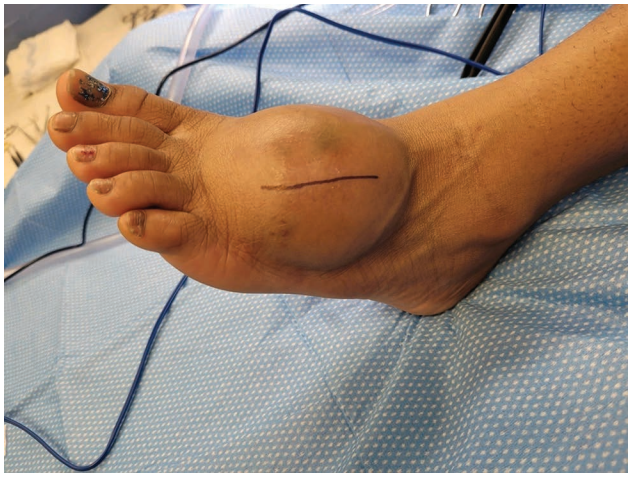

Fig. 1. Intraoperative image of dorsal foot mass with markings for planned incision.

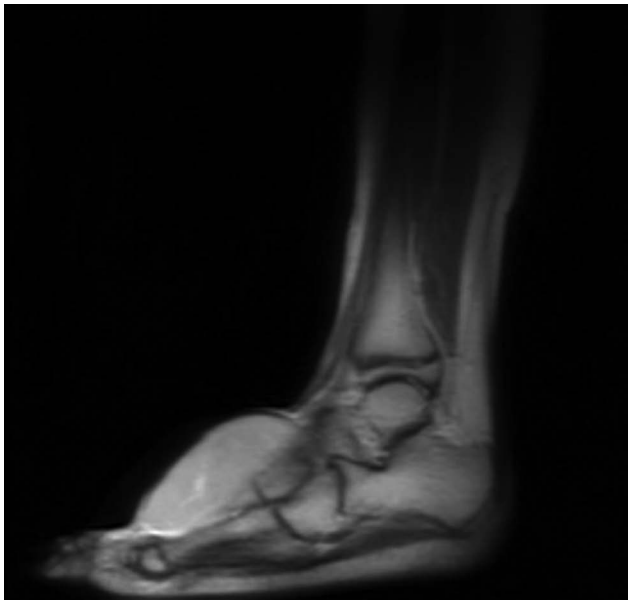

Fig. 2. MRI showing well-defined T2 hyperintense lesion of the dorsal left foot.

Pathology concluded the tumor was consistent with a pigmented-type DFSP. The patient was referred to oncology for subsequent management, and radiographic screening found no evidence of metastases. Oncology recommendations included routine monitoring with serial MRI every 3 months for 1 year, but adjuvant radiation therapy was not considered necessary because the resection was considered complete, and the margins were negative. At 6-month postoperative follow-up, the wound was healed, and the patient was ambulatory and without pain or any functional deficit. There is no evidence of recurrence.

DISCUSSION

First described in 1924, DFSP has been the subject of various case reports and series, the collective experience

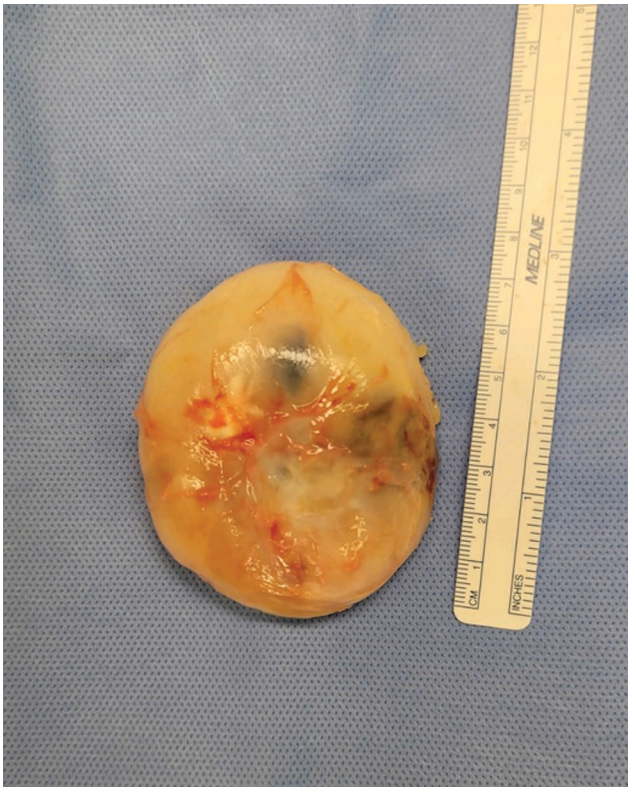

Fig. 3. Surgical specimen (8 cm × 5 cm).

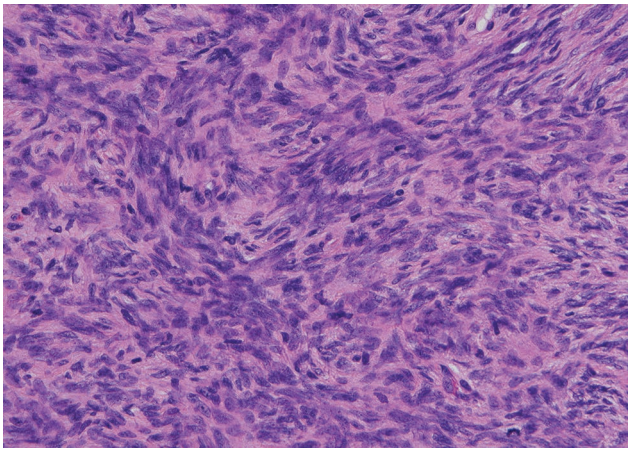

Fig. 4. Histopathology HE stained sections showing a cellular low-grade spindle cell proliferation involving the dermis and subcutaneous fat. The tumor shows spindled cells in a whorling pattern with some mitotic activity and scattered dendritic melanocytic cells. The latter feature defines this as a pigmented DFSP.

of which emphasizes the variable presentation of this entity. This unusual sarcoma is so rare in pediatric patients that delays in accurate and timely diagnosis of DFSP ranging from months up to decades have been reported.[4,5] DFSP is commonly misdiagnosed as dermatofibromas, fibromatosis, malignant fibrous histiocytomas, or vascular malformations, and most commonly occurs on the trunk or extremities as in our patient.[3,4,6] These lesions can be painful/tender to touch, but most patients report little or

no or symptoms or functional limitations. It is noteworthy that our patient continued her athletic and recreational activities despite this lesion without discomfort. Overlying skin changes such as bruising, red plaques, and gray papules have also been described in the literature, although these were not observed in our patient.

Available literature on pediatric DFSP is limited to small cohort studies. Reilly et al[7] found that just over half of the patients with DFSP presented with localized tumors in the lower limbs (53%). All patients underwent surgical excision, and none required adjuvant treatment. CD34 expression was positive in 85% of the tumors. Two of the three patients evaluated for genetic mutations had a collagen type 1A1 and platelet-derived growth factor β fusion gene.[7] Posso-De Los Rios et al[4] reported the anatomic distribution of the tumor in 17 patients. Location included the trunk (47%), extremities (41%), and other sites such as genitalia and scalp (12%).[4] DFSP can be locally invasive, but metastasis is rare. In this case, our oncology service tumor board felt that the presence of an intact and rather thick capsule, lack of adherence to any surrounding tissue, and the significant functional effect and morbidity that would result from a wide local excision was sufficient to pursue a more tempered approach to subsequent management. Standard therapy consists of tumor resection, ensuring negative margins by either Mohs micrographic surgery or wide local excision. In rare cases, patients may require adjuvant radiation and/or imatinib therapy. Recurrence rates vary and are poorly described in the pediatric population, but there is a lower recurrence rate than in adults.[1,8]

The etiology of DFSP is diverse. Translocation of chromosomes 17 and 22 is implicated in DFSP, as it leads to a fusion of collagen type 1A1 and platelet-derived growth factor β genes.[9] Nevertheless, there is an established association with trauma that may explain its predilection for the extremities.[10] In our patient, MRI findings confirmed the tumor was well circumscribed and did not invade surrounding tissues. Given the paucity of outcomes literature on DFSP in pediatric patients, our patient will undergo long-term monitoring for recurrence.

CONCLUSIONS

DFSP is a rare malignant tumor with a variable presentation and should be considered in the differential diagnosis when encountering a growing mass in a child. Early recognition, MRI evaluation, and complete surgical excision can limit local tissue invasion and morbidity.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

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