Dermatofibrosarcoma Protuberans in a 9-Year-Old Child

Abstract

Dermatofibrosarcoma protuberans (DFSP) is an uncommon soft tissue neoplasm of low-to-intermediate grade malignant potential. Childhood onset of DFSP is rare. It is most commonly seen on the trunk and proximal extremities. In children, a high index of suspicion is necessary to avoid delays in diagnosis that can lead to further morbidity. Here, we report a case of DFSP in a 9-year-old female child. Excision biopsy of lesion was performed with 1 cm margin. After confirmation of the diagnosis by histopathology, the patient was observed for recurrence, but there was no recurrence after 1 and half years of follow up.

Keywords: *Cartwheel pattern, children, dermatofibrosarcoma protuberans, Mohs micrographic surgery*

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Introduction

Dermatofibrosarcoma protuberans (DFSP) is an infiltrative low-to-intermediate grade neoplasm originating from the dermis with a limited potential for metastasis, but with a high rate of recurrence. It is commonly seen on the trunk and proximal extremities. Arrangement of tumor cells in a cartwheel pattern is histologically characteristic. It is usually seen in adults but can occur in infancy and childhood. Because of the rarity of the tumor, diagnosis of DFSP in children is quite difficult. The treatment of this tumor is adequate surgical excision and carries excellent prognosis.

Case Report

A 9-year-old female child, born of nonconsanguineous marriage, presented with red, raised, gradually progressive lesion on the left shoulder since 5 years. She complained of pain on palpation of the lesion. There was no history of trauma prior to the onset of lesion or bleeding from the lesion. There was no history of weight loss, fever, chills, or night sweats. There was no evidence of regional lymphadenopathy. On cutaneous examination, single dark red tender 4 × 3 cm firm nodule with smooth telangiectatic surface was present on the left shoulder [Figure 1]. Differential diagnosis of appendageal tumor, pyogenic granuloma, dermatofibroma, and dermatofibrosarcoma protuberans was considered. Excision biopsy showed evidence of orthokeratotic stratum corneum overlying acanthotic epidermis with lymphohistiocytic infiltrate in the upper dermis. There was diffuse infiltration of mid and reticular dermis, with spindle-shaped cells arranged in a storiform pattern, extending into the subcutaneous tissue giving honeycomb appearance suggesting the diagnosis of DFSP [Figure 2a and b]. The mitoses were less than 5 per 10 high power fields which differentiated it from dermatofibroma [Figure 3]. Patient was advised regular follow-up for the next 5 years. There was no evidence of local recurrence even after 1 and half years of follow up.

Discussion

DFSP is a relatively rare slow growing soft tissue infiltrative tumor of intermediate-to-low grade malignancy with a limited potential for metastasis, but with a high rate of recurrence.[1] DFSP was originally described as progressive and recurring dermatofibroma, hypertrophic morphea, and sarcomatous tumors resembling keloid and fibrosarcoma of the skin. It was first described by Darier and Ferrand however Hoffman officially coined the term dermatofibrosarcoma protuberans.[2] It accounts for 0.1% of all malignancies and 2–5% of soft-tissue tumors with an annual incidence of

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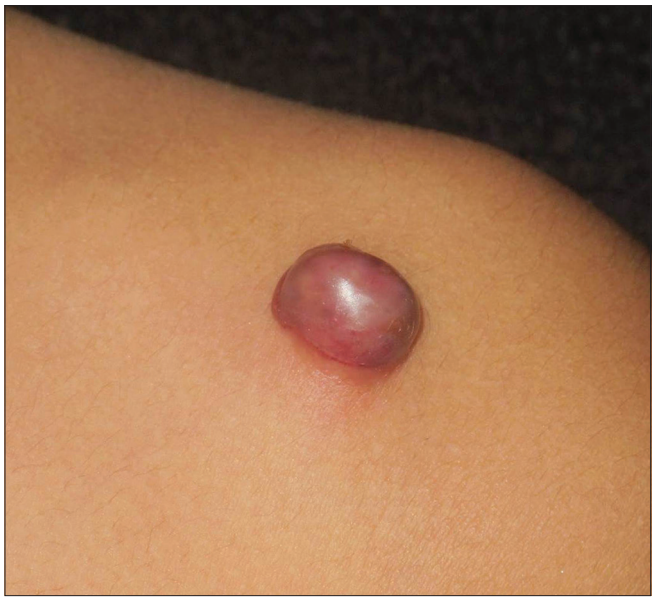

Figure 1: Single dark red tender 4 × 3 cm firm swelling with smooth telangiectatic surface present on the left shoulder

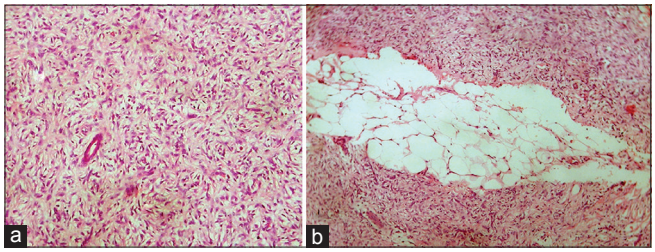

Figure 2: (a and b) Histopathology revealing spindle-shaped cells in storiform pattern and lymphohistiocytic infiltrate in dermis extending into the subcutaneous tissue giving honey comb appearance (H and E, a: ×400 and b: ×100)

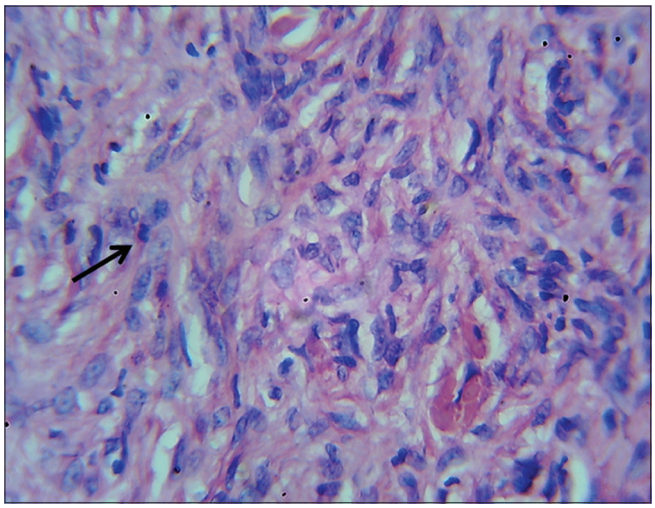

Figure 3: Histopathology revealing mitoses less than 5 per 10 high power fields which differentiated from dermatofibroma (H and E, ×400)

0.8–4.5 per million in adults.[3] The incidence in children is not known, however, it is lower than that in adults, and therefore, a high degree of clinical suspicion is required for diagnosis.

The exact pathogenesis is unknown, however, recent advances show tumour cells carry abnormal chromosomes within the tumour cells—t (17;22)(q22;q13)—resulting in the fusion gene COL1A1 (collagen type 1α 1 gene) – PDGF β (platelet derived growth factor β). This encodes a protein that causes the tumour to grow.[4]

The clinical, histological, and immunohistochemical features in DFSP in children are similar to those observed in adults. Only 6% of tumors are found in children under 16 years of age or at birth with female or no sex predilection in children.[5] Cutaneous lesions mostly occur on the trunk (40–60%) and proximal limbs (20–30%), and rarely on the head and neck (10–16%).[6] In children, it has greater propensity to appear on the legs and acral regions.[7] Congenital forms are frequently located on the trunk and proximal aspect of the limbs. The tumor has a low chance of metastasis, either to regional lymph nodes or distantly, however, is aggressive locally. A local recurrence rate of DFSP is reported to be 60%.[8] Clinical follow-up is required every 6–12 months, particularly in the first 3 years after surgery, and should include palpation of the surgical scar and regional lymph nodes.[9]

The tumor most commonly presents as a slow-growing, asymptomatic, skin-colored, indurated, firm plaque that eventually develops violaceous to red-brown nodules of varying size from one to several centimeters in diameter.[10] The tumor is often covered by a brown-yellow, red-tinged, sclerodermiform or telangiectatic, and atrophic skin. It is often mistaken for keloid, sclerosing basal cell carcinoma, morphoea, anetoderma, or scar.

There are four early clinical variants of DFSP namely confluent nodular lesions forming a sclerotic plaque, keloid-like sclerotic plaque, tumor-like, and atrophic plaque form.[11]

In congenital DFSP, erythematous-violaceous nodular plaque is the most common presentation. Histologically, DFSP has a monomorphous appearance of spindle cells with elongated nuclei and scanty pale cytoplasm primarily in the dermis arranged in storiform or whorled pattern with irregular infiltration of the fibrous septae of subcutaneous fat forming pseudoseptate or lace-like pattern, also known as honeycomb appearance. The epidermis is usually spared, but can be hyperplastic. Pleomorphism is minimal or absent.

Histological variants of DFSP include Bednar tumors (pigmented DFSP), myxoid tumors, giant cell fibroblastoma (typically affects children and adolescents, and is characterized by giant cells in the tumor), neuroid, fibrosarcomatous, myoid, and granular cell types depending upon the tissue admixed in DFSP.

The immunostaining pattern of DFSP is CD34 positive and factor XIIIa negative, which helps in differentiating DFSP from other conditions. Dermatofibromas are CD34 negative and factor XIIIa positive.

The behavior of this tumor in children is usually less aggressive. Recurrences and metastasis rates are lower than those described for adults. Complications such as bleeding, ulceration, and pain may arise at this stage. The tumor often invades deep structures such as the fascia, muscle, or bone.

The DFSP in pediatric age group should be differentiated from other fibrohistiocytic tumors such as dermatofibroma, leiomyoma, neurofibroma, dermatomyofibroma, infantile myofibromatosis, and fibrous hamartoma of infancy.

Wide local excision with clear margins is necessary because DFSP often exhibits extensive infiltration beyond gross margins.[12] Mohs micrographic surgery improves outcome and reduces recurrence rates. Other treatment modalities include radiotherapy,[13] imatinib, and sorafenib.

We report this case for the rare presentation of DFSP in children, and its rarity can often lead to misdiagnosis. Wide local excision is necessary because DFSP often exhibits extensive infiltration beyond gross margins. In our patient, DFSP was not the most probable diagnosis, hence excision biopsy was performed with 1 cm margin, however, surprisingly there is no recurrence of tumor even after 1 and half years of follow up.

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Conflicts of interest

There are no conflicts of interest.

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