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## A rare case of dermatofibrosarcoma protuberance in an adolescent female

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### Abstract

DFSP is a rare, locally aggressive, cutaneous soft tissue sarcoma, that arises from the dermis. It is seen mostly in adult males, and complete surgical excision is the standard treatment. The present case describes a young adolescent female presenting with a 6-month history of an asymptomatic small, cutaneous lesion in the right thigh. Biopsy revealed the feature of dermatofibrosarcoma protuberance (DFSP), and complete surgical excision was performed utilizing Mohs micrographic surgery (MMS) technique. This case report and literature review highlight the need for awareness of such cases which will assist in understanding and managing these tumors.

**Keywords:** Soft tissue neoplasm, dermatofibrosarcoma protuberance, spindle cell tumor, Grenz zone.

### Introduction

Dermatofibrosarcoma protuberance (DFSP) is a rare, indolent, cutaneous soft tissue sarcoma first described by Taylor in 1890<sup>[1]</sup>. The exact etiology of DFSP is unknown. It is characterized by a translocation between chromosomes 17 and 22 [t(17;22) (q22;q13)] resulting in the overexpression of platelet-derived growth factor receptor  $\beta$  (PDGFRB).<sup>2</sup> It is most often seen in adults in their thirties with slight male predominance, in the region of the trunk and proximal extremities however it can involve almost every body parts<sup>[3]</sup>. DFSP evolves as painless, skin-colored, pinkish or violet-red plaques, with surrounding telangiectatic skin. At later stages as size increases, the lesion can change into nodular, protuberant with invasion and fixation to deeper layers in particularly in advance and recurrent cases however metastasis is very rare<sup>[4,5]</sup>. Diagnostic evaluation of DFSP comprises of physical examination to assess tumor extent and mobility, imaging studies with magnetic resonance imaging (MRI) for ascertaining deeper invasion and computed tomography (CT) only in rare cases of bone involvement<sup>[6]</sup>. Biopsy with the histological examination is the definitive diagnostic method that demonstrates characteristic histological appearance with spindle cell proliferation in a typical pattern<sup>[4]</sup>. The immunostaining pattern is generally CD34 positivity<sup>[7]</sup>. Complete surgical excision with 2-3 cm of healthy margin all around is the standard treatment. DFSP occurring in an adolescent female is rarely reported. The purpose of this case report is to present the unusual example of an adolescent female patient presenting with DFSP and discuss the appropriate management options.

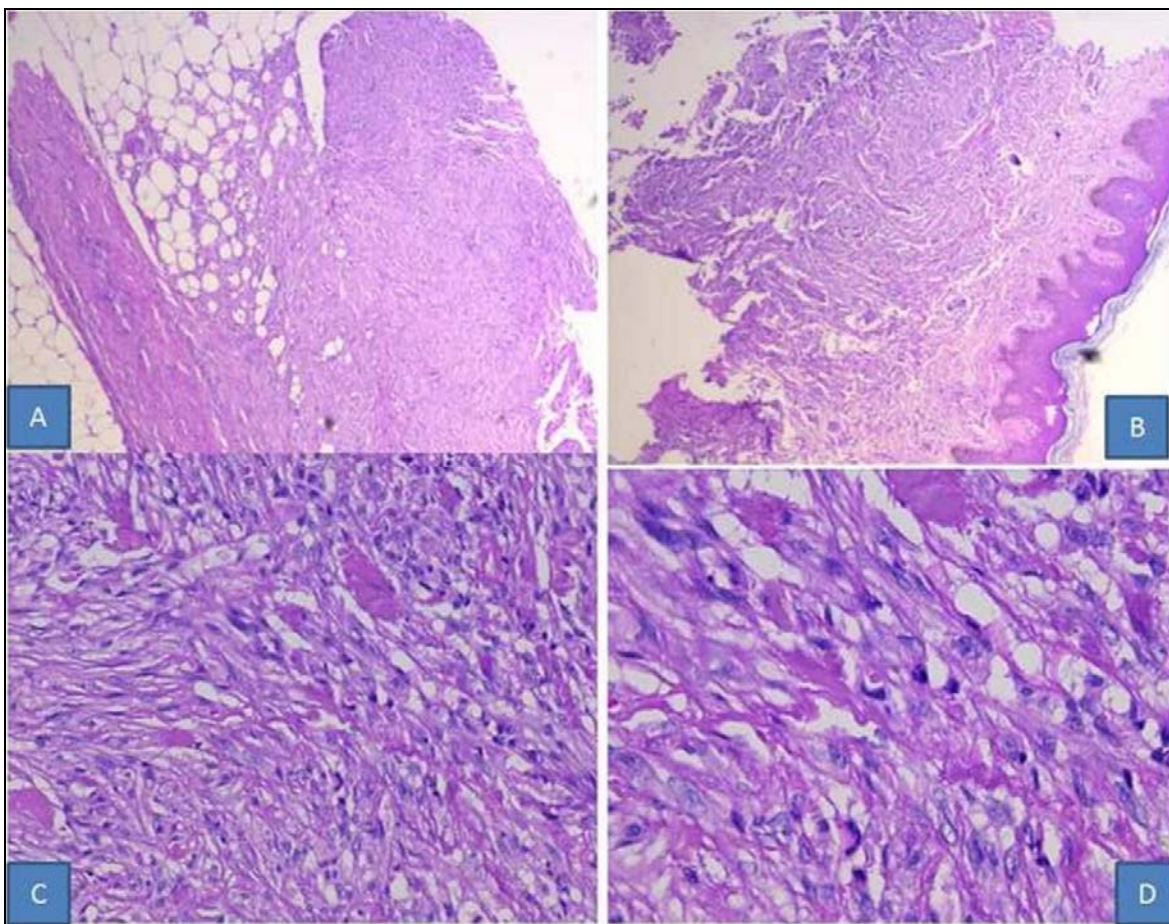
### Case presentation

This a case of a 14-year-old Indian female, who presented to our institution with a 6-month history of an insidious onset, painless, slowly growing, reddish-brown nodular lesion in the right thigh. She denied any history of associated constitutional symptom, antecedent trauma or similar lesion elsewhere in her body. Her local physical examination demonstrated a round, indurated nontender, plaque measuring 2x2 cm size, with superimposed protuberant nodules at the center (Figure 1). The lesion was relatively freely mobile over the underlying tissues but appeared tethered to the overlying skin. However, the dimple sign was positive. No palpable lymphadenopathy was noted in the inguinal area. The systemic examination was unremarkable. The reports of laboratory investigations were unremarkable. Fine needle aspiration cytology (FNAC) of the lesion showed nonspecific cells and was reported inconclusive. An Incisional biopsy was done which showed characteristic tumor cells infiltrating the whole dermis and upper subcutaneous adipose tissue, entrapping the adnexae and sparing a sub-epidermal Grenz

zone (Figure 2B). The tumor cells were of spindle-shaped, arranged in the storiform pattern and fascicles, with intervening collagenous tissue and slit-like thin-walled blood vessels. Individual tumor cells showed plump to spindle-shaped nuclei and moderate eosinophilic cytoplasm (Figures 2A, B, C, and D). The findings were consistent with the diagnosis of DFSP. Immunohistochemistry (IHC) markers like BCL-2 and CD34 were found positive (score 1+) in the tumor cells, while CD99 (MIC2) was non-immunoreactive thus confirming the diagnosis of DFSP. Complete surgical excision of the plaque was performed utilizing Mohs micrographic surgical technique (MMS). The final resection margins were free of tumor. No further adjuvant therapy was required. At follow-up 2-years later, the patient remains asymptomatic and showed no features of recurrence.



**Fig 1:** Firm Nodular Plaque on the thigh.



**Fig 2:** A. Low power view (H&E) B. Tumor-infiltrating deep into the dermis C. Irregular and Poorly defined deeper margins. D. High magnification showing malignant cells.

### Discussion

We present a rare case of DFSP in an adolescent female. DFSP is a relatively rare neoplasm representing <0.1% of all neoplasm with an annual incidence of 0.8-4.5 per million [8]. There are reports showing DFSP afflicting people of all ages and both sexes equally, however occurrence in an adolescent female is very rare [9].

DFSP is located primarily in the dermis and invades deeper layers in an irregular lace-like pattern but the origin is not well known. It is a slow-growing tumor and presentation is largely asymptomatic. At the initial stage, DFSP is often mistaken for similar cutaneous lesion like lipomas, epidermal cysts, keloids, and as it evolves in later stages, it needs to be differentiated from

a pyogenic granuloma or Kaposi sarcoma. Indolent growth pattern, together with rarity and variability in clinical appearance contributes to delay in diagnosis.

Fine-needle aspiration (FNA) might not allow an accurate diagnosis, so incisional or core-needle biopsy is the optimum tool to assess the histological features [10]. Histologically, the tumor demonstrates monomorphic, benign-appearing spindle cells, arranged in a storiform or matted pattern and intersecting at tight right angles around central vessels [4]. A thin tumor-free sub-epidermal zone (Grenz zone) is found in early lesions like in the present case. Bednar tumor or pigmented variant DFSP characterized by the presence of melanin-containing dendritic cells and myxoid DFSP are uncommon presentations [11]. In fact,

there can be significant heterogeneity within the single tumor revealing fibrosarcomatous, pigmented, juvenile, myxoid, atrophic, sclerosing, myoid or hybrid differentiation [7]. CD34 positivity is one of the most useful IHC markers to differentiate DFSP from dermatofibrosarcoma and other soft tissue tumors. The patterns are not absolute and may be seen in dermatofibroma and myxoid areas and smooth muscle actin positivity in myoid areas. Hyaluronate and CD44 staining are reported to be more useful in differentiating between the two [7]. The treatment goal of DFSP is complete surgical excision and options are well described and understood. One method is wide local excision with negative margins of 3 cm all around but this often leads to a bad scar and reported recurrence rate is 8.8%. A better option is Mohs micrographic surgery (MMS) with maximum conservation of tissues, which may require multiple stages but give better cosmesis especially in an anatomically challenging area and have a recurrence rate of 1.5%. Approximately 10-15% of cases of DFSP shows areas of fibrosarcoma (DFSP-FS) within them, which tend to exhibit aggressive course. Adjuvant radiotherapy or imatinib based targeted therapy is required in such cases [2].

Imatinib mesylate is used for the treatment of unresectable, recurrent, or metastatic DFSP. It can also be considered in unresectable cases, or if unacceptable functional or cosmetic results are anticipated with further resection [2]. In cases of positive surgical margins, postoperative radiation therapy can be considered if further resection is not possible [2]. Recurrent tumors should be resected whenever possible along with adjuvant radiation if feasible after surgery. In the case of metastatic disease, imatinib mesylate, chemotherapy, and radiation therapy should be considered. Single-agent or combined chemotherapy may be considered for DFSP. These include AIM (doxorubicin/ifosfamide/mesna), doxorubicin, ifosfamide, epirubicin, gemcitabine, dacarbazine, liposomal doxorubicin, temozolomide, vinorelbine, or pazopanib [2]. Local recurrence is common after incomplete excision and long term follow up is recommended to detect future recurrence.

**Conflict of interest:** None.

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