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Case Report

Dermatofibrosarcoma protuberans mimicking a benign lesion: case report on a rare diagnostic dilemma

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare, low- to intermediate-grade dermal sarcoma, representing less than 1% of head and neck malignancies. It is locally aggressive, with high recurrence rates but low metastatic potential. Parotid region involvement is exceptionally uncommon. A 35-year-old male presented with a recurrent swelling in the left pre-auricular region, one month after superficial parotidectomy. Histopathology of the earlier specimen had confirmed DFSP. Positron emission tomography (PET) revealed fluorodeoxyglucose (FDG)-avid nodular enhancement, and wide local excision (WLE) with reconstruction was performed. Margins were clear, and adjuvant radiotherapy was administered. DFSP in the parotid region is extremely rare and may mimic benign parotid tumors, leading to delayed or incorrect diagnosis. Complete surgical excision with negative margins remains the cornerstone of management, while radiotherapy can reduce recurrence risk in high-risk cases. Given its infiltrative nature and propensity for late recurrence, meticulous long-term follow-up is mandatory to ensure optimal outcomes.

Keywords: Dermatofibrosarcoma protuberans, Fine-needle aspiration cytology, Mohs micrographic surgery

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) was first described by Darier and Ferrand in 1924, but the definition of "DFSP" was established by Hoffman in 1925.[1] Sarcomas in general occur rarely in head and neck region accounting for less than one percent of head and neck malignancies. Dermatofibrosarcoma of head and neck is a rare neoplasm.[2] DFSP is a slow growing, low- to intermediate-grade dermal soft-tissue tumor. It has a high local recurrence rate but low metastatic potential. It is characterized by a uniform spindle cell arrangement, classically with a storiform pattern and CD34 immunoreactivity. The histomorphology and immunophenotype overlap with a broad range of other neoplasms. DFSP has characteristic t (17; 22) (q22; q13), resulting in a COL1A1-PDGFB fusion transcripts in more than 90% of DFSPs.[3]

DFSP clinically manifests as a firm, indurated plaque or nodule, most commonly presenting on the trunk or proximal extremities.[4] The typical age at diagnosis is between 20 and 50 years, but DSFP may present at any age, especially if congenital.[4] DFSP most commonly presents in males than females (57% vs. 43%) and occurs most frequently in black patients.[5]

DFSP is primarily managed through surgical intervention in most cases. Achieving complete resection during the initial surgery is strongly recommended, as the tumor carries a significant risk of local recurrence. Incomplete excision and repeated surgical procedures are associated with an increased likelihood of metastasis. Surgical modalities include wide local excision (WLE) and Mohs micrographic surgery (MMS), though the literature presents conflicting evidence regarding the optimal approach for DFSP management. Unresectable DFSPs are treated with radiation therapy and/or targeted therapy.[6]

We report this rare entity in a young male patient who was treated surgically. He had undergone a left superficial parotidectomy one month earlier, and the histopathological report of that specimen was suggestive of DFSP.

CASE REPORT

A 35 year old male patient presented to the ENT outpatient department with complaints of a swelling over the left cheek region of one-month duration. The patient had previously undergone a left superficial parotidectomy which was performed 1 month ago, based on fine-needle aspiration cytology (FNAC) findings suggestive of pleomorphic adenoma. Histopathological report of resected specimen suggestive of DFSP.

On local examination, inspection revealed a single, well-defined swelling in the left pre-auricular region, measuring approximately 3×1.5×0.5 cm. The overlying skin was tense but normal in color, without signs of ulceration or discharge. No regional lymphadenopathy was detected. Palpation confirmed a firm, non-tender lesion with restricted mobility over the deeper tissues.

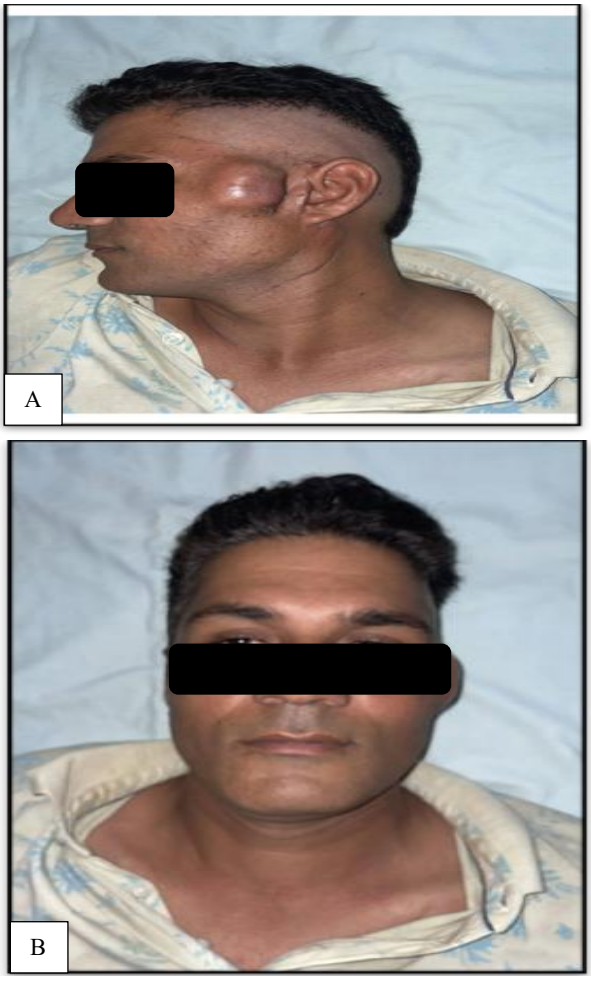

Figure 1: Pre operative clinical image (A) lateral view; (B) front view.

A positron emission tomography (PET) scan demonstrated fluorodeoxyglucose (FDG)-avid nodular enhancement in the left pre-auricular skin, with a maximum standardized uptake value (SUV_{max}) of 8.55. These findings suggested the possibility of a malignant lesion.

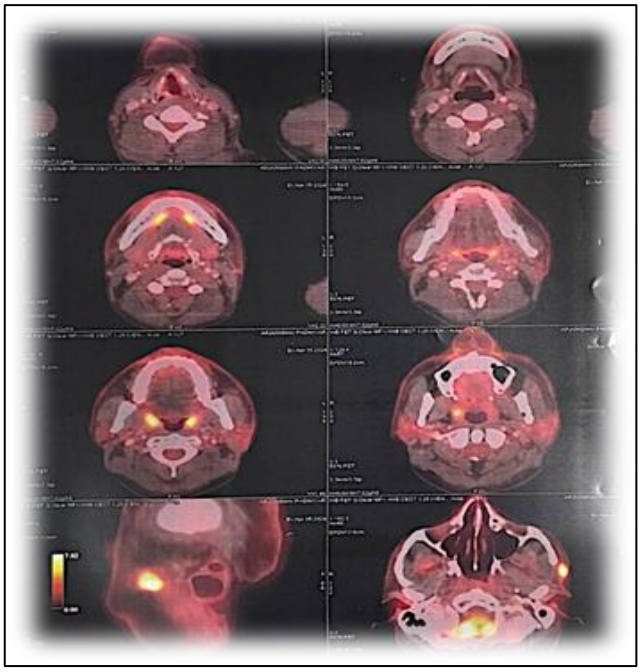

Figure 2: Pre operative PET scan.

The patient was taken up for surgery, and WLE of the swelling was performed with adequate margins, given the aggressive local infiltration commonly associated with such tumors. Intraoperatively, the lesion was found to be confined to the skin and subcutaneous tissue without deeper extension into parotid or bony structures. Care was taken to preserve facial nerve function. Reconstruction of defect of primary tumour excision part was done with anterolateral thigh flap and microanastomosis done with superficial temporal vessels.

The excised specimen was submitted for histopathological examination. Grossly, the lesion appeared as a well-circumscribed nodular mass with firm consistency. Microscopic evaluation revealed a dermal and subcutaneous tumor composed of spindle-shaped cells arranged in a storiform pattern. The tumor cells infiltrated the surrounding adipose tissue in a characteristic honeycomb pattern. Nuclear pleomorphism was minimal, and mitotic activity was noted. The surgical margins were free of tumor infiltration.

Given the infiltrative nature of the disease and the high risk of local recurrence, the patient was planned for adjuvant postoperative radiotherapy. He received external beam radiotherapy to the tumor bed with appropriate safety margins. The treatment was well tolerated, with no major acute complications.

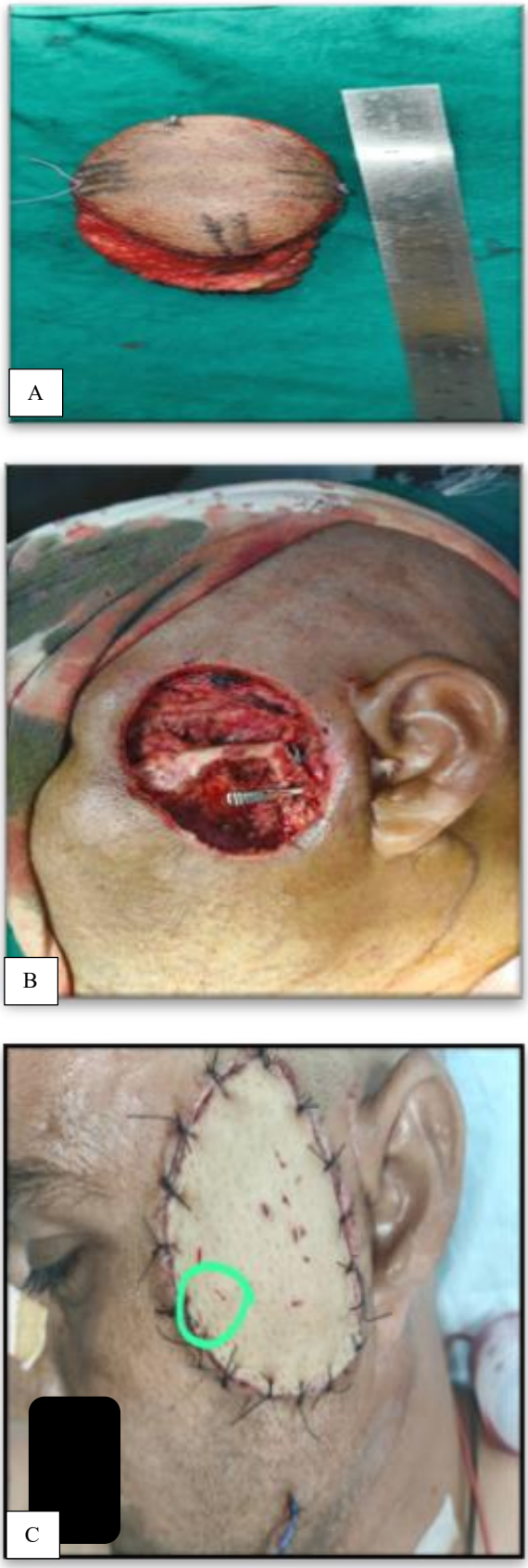

Figure 3: Intra operative image (A) intra operative specimen; (B) intra operative defect area; (C) intra operative area after reconstruction.

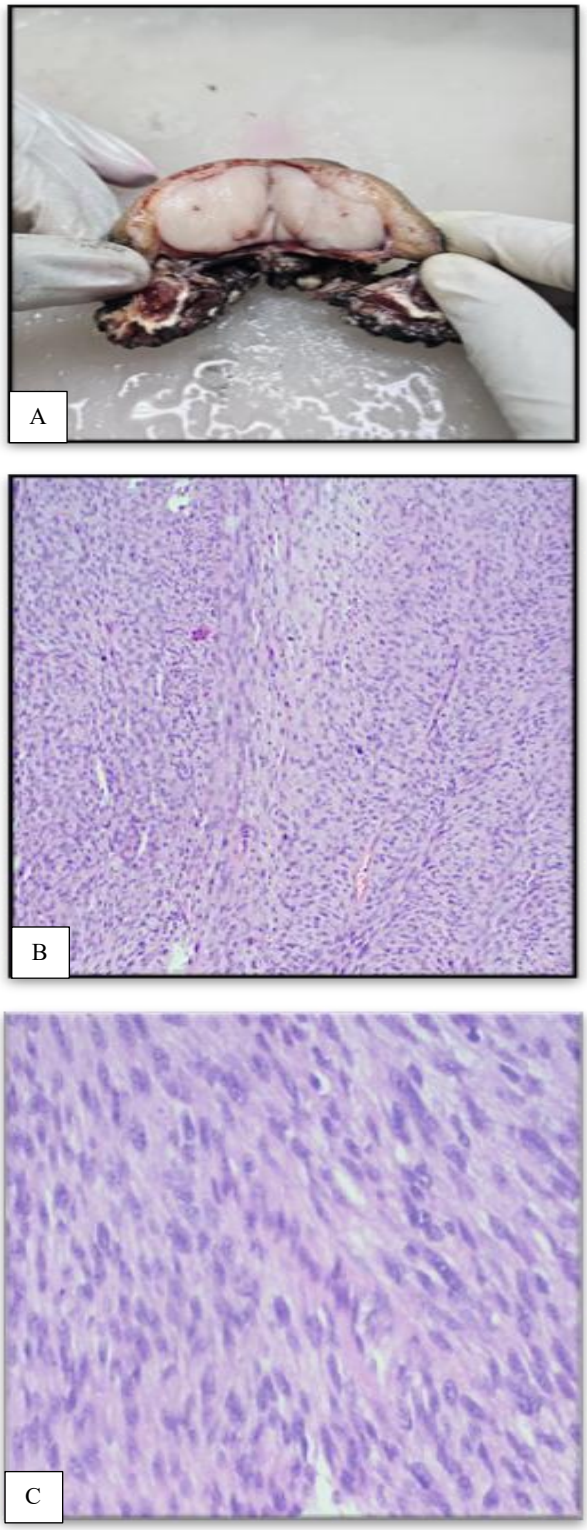

Figure 4: Specimen (A) gross specimen; (B) low power (10x) section; (C) high power (40x) section.

The patient's postoperative period was uneventful. He was discharged on regular follow-up and remains under close surveillance to monitor for any evidence of recurrence.

Figure 5 (A and B): Post operative 3 months and 9 months.

DISCUSSION

DFSP is a fibrohistiocytic tumor of low to intermediate malignant potential, characterized by infiltrative margins, a high rate of local recurrence (approximately 50%), and rare distant metastasis.[7] The most common location of DFSP is the trunk (42-72%) followed by proximal extremities (20-30%), and head and neck (10-16%). DFSP sites include surgical scars, old burns, trauma, radiation dermatitis, vaccination sites, central venous line puncture sites and even insect bites.[8]

The routine diagnostic workup for DFSP includes cross-sectional imaging, preferably magnetic resonance imaging (MRI), along with biopsy and histopathological confirmation. On computed tomography (CT), these lesions typically present as well-defined masses that are hypodense relative to muscle and exhibit homogeneous contrast enhancement. On MRI, DFSP usually appears homogeneous and iso- to hypointense compared to muscle, with strong post-contrast enhancement, particularly on T2-weighted sequences.[9]

The differential diagnosis of DFSP includes solitary fibrous tumor, benign fibrous histiocytoma, schwannoma, myoepithelioma, and other spindle cell sarcomas. Among these, solitary fibrous tumor poses the greatest diagnostic challenge due to its overlapping histological features and immunohistochemical profile. Solitary fibrous tumors often demonstrate a heterogeneous growth pattern with alternating hypercellular spindle cell areas, prominent hemangiopericytic vasculature, and regions of sclerosis or myxoid change. In contrast, DFSP is characterized by striking histological uniformity, absence of a hemangiopericytic pattern, and presence of a storiform architecture surrounding an inconspicuous vasculature. Given the potential overlap in immunohistochemical staining results, careful correlation with morphological features is essential for accurate differentiation between DFSP and solitary fibrous tumor.[10]

Surgery remains the cornerstone of treatment for DFSP, with the primary objective of achieving complete excision with negative margins. Treatment options include simple excision, WLE with reconstruction, and MMS. Among these, MMS is often preferred because of its tissue-sparing precision and superior local control rates. Adjuvant radiotherapy has an important role in cases with positive or close margins, where the risk of local recurrence is significantly higher. In such situations, radiotherapy can effectively reduce recurrence rates and contribute to improved long-term outcomes.[11-13]

MMS is considered the most effective approach for the management of DFSP, as the tumor often extends into surrounding tissue in finger-like projections. This technique allows for meticulous three-dimensional assessment of tumor margins using frozen or paraffin sections. Resection is performed with approximately a 2 mm safety margin, and horizontal sections from the deep surface of the specimen are sequentially examined until clear margins are achieved on frozen section analysis. This method not only minimizes the risk of local recurrence but also reduces surgical morbidity by preserving uninvolved tissue.[14]

Targeted therapy with tyrosine kinase inhibitors, particularly imatinib, has shown encouraging results in advanced or metastatic DFSP, especially in patients harboring the COL1A1-PDGFB fusion gene. While aggressive surgical resection remains the standard of care and is generally associated with low recurrence rates, adjuvant systemic therapy may be valuable in unresectable or progressive cases. Reported series indicate a 5-year local recurrence rate of approximately 2-3% following wide or aggressive excision, although the risk increases in the presence of positive margins or deep tissue invasion.[15-17]

The overall prognosis of DFSP is favorable, with 5-year survival rates reported between 93% and 100% following appropriate treatment. Metastatic spread, occurring via hematogenous or lymphatic routes, is rare and usually

associated with advanced disease or multiple local recurrences. Although DFSP is notorious for frequent local recurrence, the incidence of distant metastasis remains low, ranging from 1% to 4%.[18]

CONCLUSION

Aggressive surgical resection with negative margins remains the most effective treatment for DFSP in the head and neck region, offering low recurrence rates when complete excision is achieved. However, due to the tumor's infiltrative nature and potential for microscopic spread, long-term follow-up is crucial, as recurrences may develop many years after initial treatment. Adjuvant radiotherapy can be considered in high-risk cases, such as larger or deeper tumors or those with positive margins, although its benefit remains uncertain. DFSP of the parotid region is extremely rare, and awareness is essential for accurate diagnosis and timely management.

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