Dermatofibrosarcoma protuberans of the upper extremity: a case report

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Introduction

Dermatofibrosarcoma protuberans (DFSP) is a slow-growing, rare, sarcomatous skin tumor that constitutes around 1% of all soft tissue sarcomas[(1)]. The presentation of the tumor varies from a painless small plaque to a nodular, protuberant, or ulcerative lesion[(2)]. The tumor can be misdiagnosed as a simple scar or keloid. Though regional and distant metastasis is rare, a tendency to infiltrate adjacent and deeper structures like subcutaneous tissue, muscles, or bones is known[(3)]. Immunohistochemistry helps in confirmation of diagnosis as the tumor cells are positive for CD34, vimentin, apolipoprotein D and nestin. The reason behind development of this lesion is attributed to overproduction of platelet-derived growth factor (PDGF) as a result of chromosomal translocation, resulting in the fusion protein COL1A1-PDGFB that causes tumor growth. Inadequate resection can lead to recurrences.

Case Report

A 50-year-old male presented with a firm, mobile and non-tender swelling of size 5x4 cm over the right arm for six months, progressively increasing in size (Figure1). There was no associated right axillary lymphadenopathy. Fine Needle Aspiration Cytology (FNAC) of the swelling revealed features of DFSP.

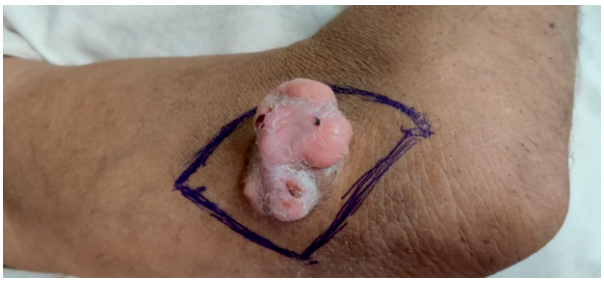

Figure 1 : Lesion over the right arm

X-ray chest and ultrasound of the abdomen showed no signs of distant metastasis. The patient underwent wide local excision with a 2.5 cm margin around the lesion with primary closure (Figures 2 and 3). Final histopathological report confirmed the lesion to be DFSP with spindle cell variation (Figure 4). Immunohistochemistry was positive for CD 34. The follow-up period of 8 months has been uneventful.

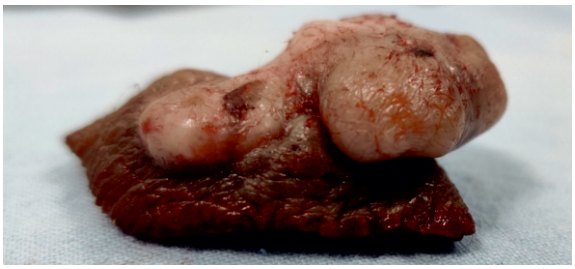

Figure 2: Excised specimen

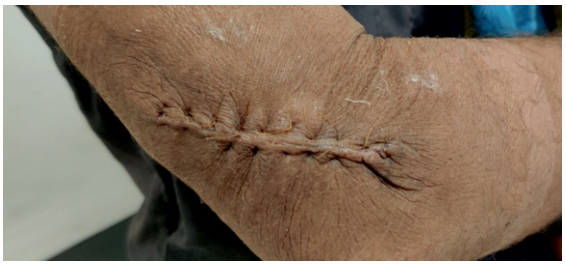

Figure 3: Primary closure of defect after surgical excision

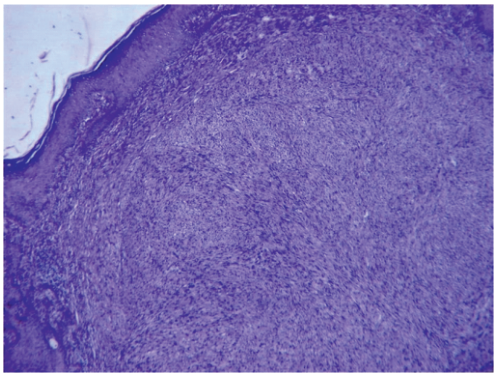

Figure 4: Well circumscribed tumor mass in the dermis (HP 10X) composed of spindle cells and histiocytic cells, arranged in fascicles and bundles

Discussion

Darier and Ferrand first described the entity DFSP in 1924, but the term DFSP was first coined by Hoffmann in 1925. Later in1962, Taylor and Helwig, in their series of 115 cases, described the characteristic features of a low-grade sarcoma with fibroblastic activity[(4)]. The tumor affects males and females equally, and the incidence is more common in the fourth and fifth decades of life[(5)].

The common sites for occurrence are the trunk and proximal aspects of the extremities. The presentation of the tumor

varies from a painless small plaque to a nodular, protuberant, or ulcerative lesion. Microscopically, the tumor comprises atypical spindle cells arranged in a cartwheel pattern in the matrix of fibrotic stroma in the dermis. Although the mitotic activity or nuclear pleomorphism is bare minimal, the local infiltration is seen in the form of small tentacles that may be the cause for local recurrence[6]. The diagnosis is confirmed upon histopathology with a few histopathological subtypes described in literature like pigmented (Bednar tumor), giant cell fibroblastoma-like, atrophic, sclerosing, granular cell variant, fibrosarcomatous and myxoid DFSP[7]. DFSP being a type of spindle cell tumor, the histopathology sometimes can have differential diagnoses of cellular dermatofibroma, fibrosarcoma, malignant fibrous histiocytoma, atypical fibroxanthoma, desmoplastic melanoma, Kaposi sarcoma, and solitary fibrous tumor. Therefore, in such cases, immunohistochemistry helps in differentiation as an expression of CD34 is seen in 80-100% of tumors with vimentin been considered as an alternative diagnostic marker for DFSP tumors[8]. In our patient, CD34 was used as a marker for immunohistochemistry, that came out to be positive and was the best marker available.

Surgical treatment is in the form of wide local excision, with margins of 2 to 5 cm from the tumor edge, including the skin, the subcutaneous tissue, and the underlying fascia[9]. Reconstructive surgery can be performed for larger tissue defects using a local skin flap, skin graft, or myocutaneous flap. Mohs micrographic surgery is now gaining popularity as an alternative to wide local excision for DFSP[10]. Unresectable, recurrent, and/or metastatic disease can be considered for adjuvant therapy using imatinib mesylate, a tyrosine kinase inhibitor. Radiotherapy alone or combined with surgery should be considered in cases of positive or inadequate margins, recurrence, or where cosmetic disfigurement is anticipated after wide excision[11]. The recurrence rate is high, especially if adequate margins are not taken care of. Recurrences may appear early within the first year of surgery or may be delayed even after five years. Thus, a long-term follow-up is necessary for these patients.

Conclusion

Although rare, a differential diagnosis of Dermatofibrosarcoma protuberans must be taken into account in any patient presenting with cutaneous soft tissue swelling, especially with absent features of loco-regional or distant metastasis.

Ethical consideration: Patient's written informed consent was taken.

Conflict of Interest: Nil

Source of Support: Nil

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