Dermatofibrosarcoma of Anterior Chest Wall: A Rare Entity

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ABSTRACT
We present a typical case of dermatofibrosarcoma protuberans (DFSP) of anterior chest wall managed by surgical approach. This is a rare tumor with frequent recurrence after surgery. So, there is definite need of observation after surgery. We also describe the histopathologic findings.

Keywords: Dermatofibrosarcoma protuberans, Skin tumor, Soft tissue tumor.

INTRODUCTION
Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing tumor of fibrohistiocytic nature considered of medium to low malignancy. It mainly affects young and middle-aged male adults. It grows as an asymptomatic bluish or otherwise brownish erythematous multinodular plate, and the areas most frequently involved are the trunk, chest wall, proximal extremities, head, and neck. It affects adult males between the second and fifth decades of life. Local recurrence after incomplete resection is common, although distant metastases are rare. Recognition of this tumor is very much important because of the excellent prognosis after adequate excision surgically, ensuring adequate margin of 3 to 5 cm. In histopathology, the tumor is located in the dermis and consists of uniform fusiform cells, densely grouped, with an elongated nucleus, showing strong positive reaction to CD34 and negative to S-100 and desmin. This DFSP case is reported for its rarity. The tumor accounts for less than 0.01% of all malignancies. Remote metastases are also rare, but when it does occur the common sites are the lungs and the regional lymph nodes.

CASE REPORT
The patient (40-year-old male) was admitted in outpatient department of Rohilkhand Medical College and Hospital, Bareilly, Uttar Pradesh, with complaints of swelling over the anterior chest wall (3 cm from left nipple and 4 cm from right nipple) from 5 years. According to the patient the swelling was gradually increasing in size. He had not taken any treatment before.

Physical examination reveals a mass of 6 × 4 cm with potato-like projections. The overlying skin is indurated. The tumor is nonmobile, i.e., fixed to the underlying tissues with fluctuations. Regional lymph nodes were not enlarged. Clinical examination did not reveal any other abnormality.

Chest X-ray shows no abnormality, so the fine-needle aspiration cytology was planned, which shows spindle-shaped nuclei with tapering ends and scant cytoplasm. Surrounding stoma is abundant myxoid, which shows the impression of spindle cell lesion. All other lab findings are in normal limits.

In view of the possibility of extension of the tumor tissue to the surrounding areas, a wide resection was planned so as to include more than a 3.5-cm margin. The excised tumor shows the uneven projections (potato like) (Fig. 1).
DISCUSSION

The term “DFSP” was first used in 1925 by Hoffman.\(^5\)\(^6\) The other terms used to describe this neoplasm are hypertrophic morphea, progressive and recurrent dermatofibroma, fibrosarcoma of skin, and sarcomatous tumor resembling keloid.\(^7\)\(^9\) The entity originates from the dermis and is a rare malignant dermal tumor. Although it can be seen at any age, it is found mostly between 30 and 50 years of age. Approximately 10% of patients are children. The incidence is 0.8 to 4.2 per million population. The tumor afflicts both sexes approximately equally but more in males. The cause of DFSP is not known. However, cytogenetic investigations have shown tumor cells with chromosomal anomalies together with mononuclear cellular origin. Asymptomatic multinodular bluish or brownish erythematous plate, developing over years, with its typical “protuberant” aspect, is the most frequently observed clinical aspect.\(^1\)\(^10\) Histopathology reveals relatively uniform densely grouped fusiform cells, with elongated nuclei without significant cytologic atypia or pleomorphism in characteristic storiform arrangement.\(^3\) The degree of nuclear atypia is higher in nodular lesions than in plates. Fibrosarcomatous focal alterations with a characteristic fish bone pattern are occasionally observed in DFSP.\(^11\) The standard treatment of the localized disease consists of wide local surgical resection with recommended surgical margins of 2 to 3 cm and three-dimensional resection including skin, subcutaneous tissue, and underlying fascia.\(^11\)\(^13\) There are current reports praising the use of Mohs micrographic surgery as a first-line therapeutic measure in cases of limited tumors for tissue preservation and reduction of recurrence rate.\(^14\)\(^17\) Radiation therapy is an adjuvant therapy in cases where adequate surgical margins are not easily reached or result in esthetic/functional defect, or in cases of positive margins, even after maximum resection. It is also indicated for patients with inoperable macroscopic lesions.\(^1\)\(^11\)

REFERENCES