Dermatofibrosarcoma Protruberans - A Case Report

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Abstract: Dermatofibrosarcoma is a relatively uncommon soft tissue neoplasm with an annual reported incidence rate of around 3 per million cases per year.it has intermediate to low grade malignancy. It is a locally aggressive tumour with high recurrence rate. We present a case of dermatofibrosarcomaprotuberans in a 65 year old man.

Keywords: dermatofibrosarcomaprotruberans; cutaneous soft tissue neoplasms; soft tissue tumours with intermediate to low grade malignancy; dermatofibroma

Introduction:
Dermatofibrosarcomaprotuberans (DFSP) is a relatively uncommon soft tissue neoplasm with intermediate to low-grade malignancy with an incidence of 3 per million per year. DFSP is a locally aggressive tumour with a high recurrence rate. Although the cellular origin is unclear, DFSP is considered to be a cutaneous soft tissue sarcoma. Common sites are torso but can be found at other areas also.

Darier and Ferrand first described it in 1924 as a distinct cutaneous disease entity called progressive and recurring dermatofibroma. Hoffman officially coined the term dermatofibrosarcomaprotuberans in 1925[1].

History:
A 65 year old male patient presented with a swelling on the upper part of back overlying the left scapula extending to the midline for aduration of 5 years. The swelling was progressively increasing in size, not associated with pain. There were no complaints suggestive of neurological involvement such as tingling, numbness or radiating pain. No history of trauma, loss of weight and appetite.

Examination:
The swelling was 10 X 7 cm in size, overlying the left scapula extending to the midline, variegated in consistency, non fluctuant, not fixed to underlying muscles or overlying skin. There was no axillary lymphadenopathy or loss of movements of the shoulder joint or spine.

FNAC of the swelling was inconclusive. Chest x ray was done which was normal. Decision was taken to do a wide local excision with 2cm margin for symptomatic relief. Intraoperative findings were a subcutaneous swelling with no involvement of underlying muscles.

The biopsy report was: section shows skin with flattened epidermis, dermis and subcutaneous tissue with tumour composed of bundles and fascicles of spindle shaped cells with plump, spindle shaped nuclei with mild hyperchromasia and anisonucleosis. Few mitotic figures were noted.

The patient was followed up for 6 months. There was no functional deficit. Post operative chest x ray was done to see for metastasis. As no metastases were found, no further treatment was given.

Discussion:
Dermatofibrosarcomaprotuberans (DFSP) is a slow growingcutaneous malignancy arising from the dermis and invades deeper subcutaneous tissue.

The cellular origin of DFSP is unclear. Evidence supports the cellular origin being fibroblastic, histiocytic, or neuroectodermal. Many authorities suggest pluripotential progenitor cells, such as undifferentiated mesenchymal cells, may be the origin of DFSP, because they have the capacity to differentiate into all 3 cell types [2].

Platelet derived growth factor activation has also been implicated. Cytogenic studies also reveal abnormalities like reciprocal translocations between chromosomes 17 and 22.

Epidemiology:
The annual incidence of DFSP is reported as 3 cases per million population from a population-based cancer registry from 1982-2002 in France [3].

DFSP is a locally aggressive tumour with a high recurrence rate. Metastasis in DFSP is rare (1-4%). However, almost all metastatic cases have been associated with local recurrence, anda poor prognosis [4].

DFSP is more often seen in the males between 20-50 years.

DFSP usually presents as a large, indurated plaque several centimetres in diameter. DFSP is composed of firm, irregular nodules varying in colour from flesh to reddish brown. Mostly the tumour is mobile; fixity to deeper structures such as fascia, muscle, and bone may occur in the later stages.

DFSP most commonly occurs on the trunk (42-72%), followed by the proximal extremities (16-30%). DFSP rarely occurs above the neck (10-16%) [1]

Other investigations useful in DFSP are Chest radiography: for pulmonary metastasis in high-risk cases, CT scan for bone metastasis; MRI for depth of invasion and atypical lesions and FDG-PET scan to monitor metastatic disease.

Histopathology:
A skin biopsy is essential for the definitive diagnosis of DFSP.
Histological types are plaque and nodular.
DFSP may show focal fibrosarcomatous changes with a characteristic herringbone pattern.
The pigmented variant of DFSP, also known as Bednartumour has the melanin-containing dendritic cells scattered between the neoplastic spindle-shaped cells [5].
Staging.
There are three stages. Stage 1 is localised primary tumor. Stages 2 and 3 have lymph node and distant metastases respectively [6].

Treatment:
Mohs micrographic surgery is the treatment of choice particularly for lesions located in the head and neck region, however some authors advocate Wide local excision [7,8].

A major cause of high recurrence is infiltration beyond clinical margins. Hence, a wide excision of 2-3 cm or more of the margins beyond identifiable tumour border, down to and including the fascia, is recommended for the treatment of DFSP.

Prognosis:
DFSP requires a close follow up because of high recurrence rates, mostly seen during the first three years following primary excision. Lymph node involvement represents poor prognosis; most patients die within 2 years. The lungs are the most common site of distant metastasis. Age above 50 is also a poor prognostic factor.

Photo 1:Showing swelling in the back

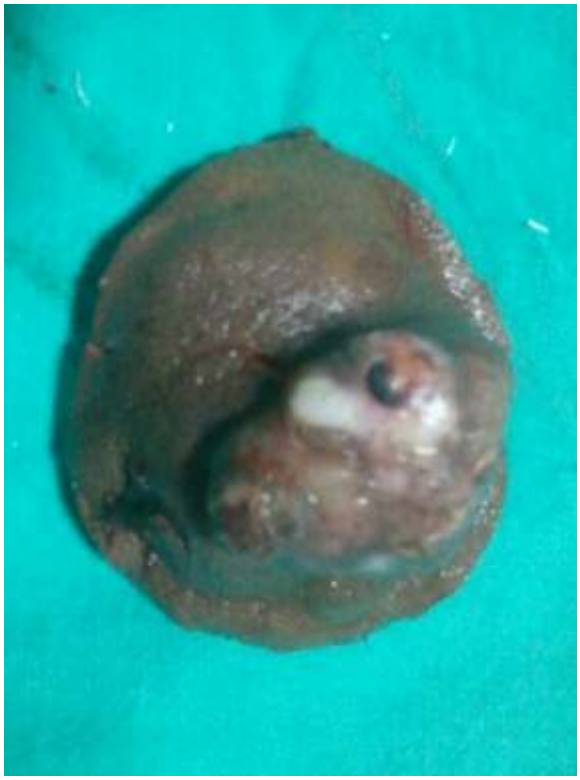

Photo 2: Resected specimen of dermatofibrosarcoma protruberans

Conclusion:

We are reporting this case of dermatofibrosarcoma-protruberans as it is a rare tumour with an incidence of 3 in one million.

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