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CASE REPORT

Dermatofibrosarcoma Protuberans: A Rare Cause of Neck Swelling

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Abstract

Dermatofibrosarcomaprotuberans (DFSP) is a low-grade soft tissue sarcoma (fibrosarcoma) originated from dermal and subdermal layer of the skin. The tumor infiltrates into the deeps in the form of villous or finger. For this reason, it is quite difficult to get clear surgical margins during tumor excision and the recurrence is a problem encountered frequently. We are reporting this rare tumour in a 40-year-old man presented with a 5-years history of slowly growing cutaneous lesion of the neck. Incisional biopsy confirmed the diagnosis of DFSP. Subsequently, the patient underwent wide local surgical resection, followed by reconstruction. Histopathology report revealed dermatofibrosarcomaprotuberans (DFSP). Although DFSP behave as non-aggressive malignancy, surgery with complete removal of the affected area is the treatment of choice. Moreover, adjuvant radiotherapy and follow-up of the patient is essential in order to prevent recurrence.

Keywords: *Dermatofibrosarcomaprotuberans, Recurrence, Radiotherapy.*

Introduction

Dermatofibrosarcomaprotuberans (DFSP) is a locally aggressive, slow growing, cutaneous, malignant tumor characterized by high rates of local recurrence and low risk of metastasis. It originates in the dermis, with a tendency to invade the surrounding tissue. This infrequent tumour typically presents as a solitary or a multilobulated and a painless cutaneous mass.[1]There is less clinical awareness because of its relatively rare occurrence and diagnosis is made on histology. The surface is characterized by irregular protruded multiple swelling, and a hard indurated plaque. It is commonly diagnosed in young and middle aged adults, but can involve children and elderly as well. Males are affected four times as often as females.[2] Surgical excision with wide margins is the main stay of treatment. The DFSP has high tendency of recurrence, due to its infiltrative asymmetric growth pattern. Although metastasis is rarely seen but usually preceded by multiple local recurrences. Surgery followed by post-operative radiotherapy may effectively reduce the local recurrence rate.[1]

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Case Report

A 40-year old male presented with right sided neck swelling for 5 years duration with progressive increase in size for the last six months. The swelling was localized over right sternocleidomastoid muscle.

The lesion was about 8.5x5.5cm, painless, multilobulated with erythematous overlying skin. There was no reported significant family history or history of addictions. No other abnormality was detected on examination of head and neck, systematic examination was unremarkable. Incisional biopsy shows a neoplasm, composed of spindle cells, arranged in storiform pattern, decision for wide local excision was taken. Intra-operatively right SCM was found to be free from the tumour, sparing the muscle. The tumour was excised with 3 cm margins along with overlying skin. Histopathologic studies showed uniform population of spindle cells arranged in storiform pattern and these spindle cells also showed infiltraltion into surrounding stroma.

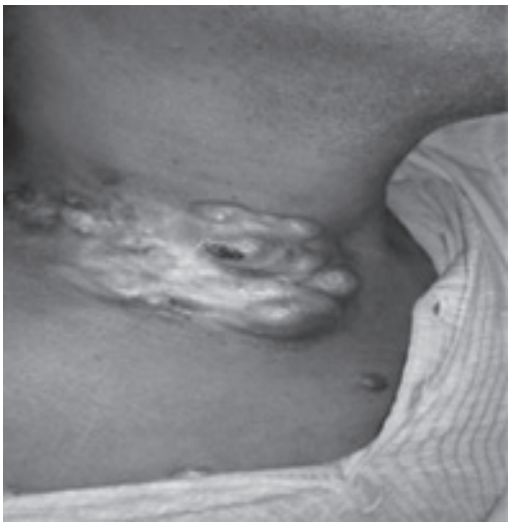

Fig.-1: *Photograph of neck showing multiple nodular swelling.*

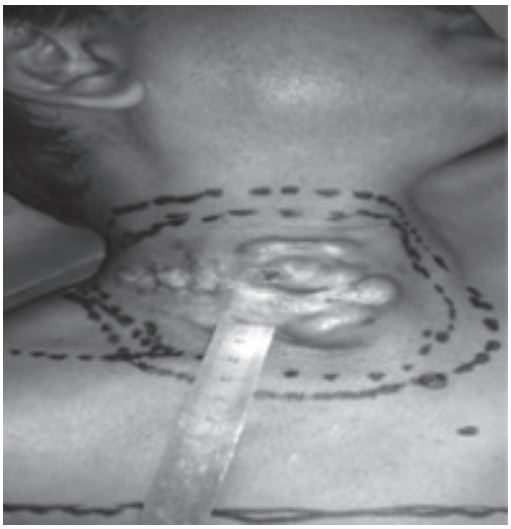

Fig.-2: *Pre-operative marking for 3 cm tumour margin.*

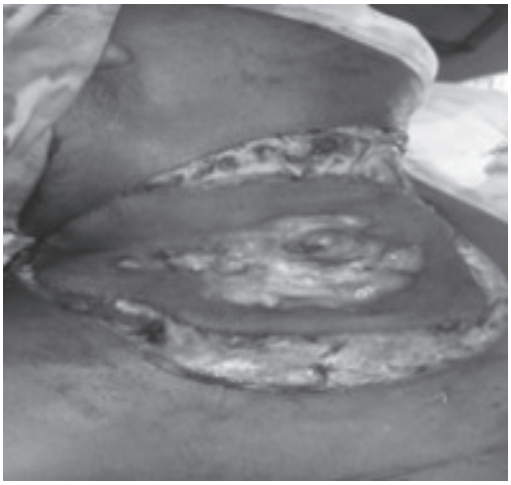

Fig.-3: *Excision of mass with 3 cm margin.*

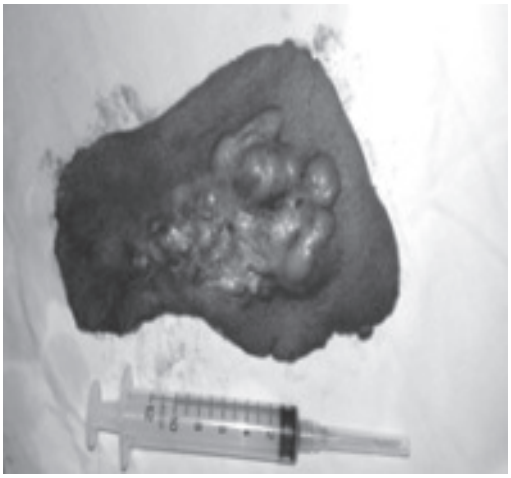

Fig.-4: *Excised mass*

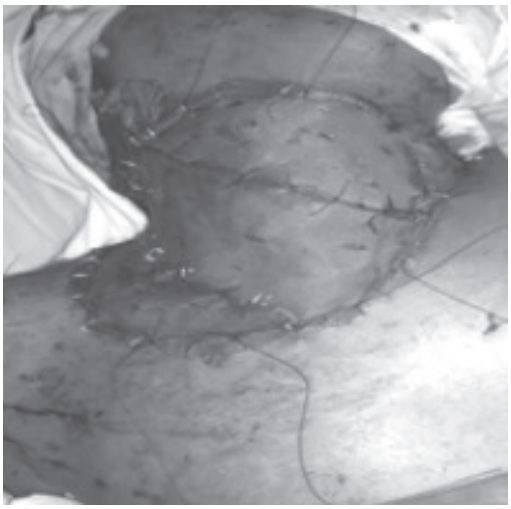

Fig.-5: *Reconstruction with skin grafting.*

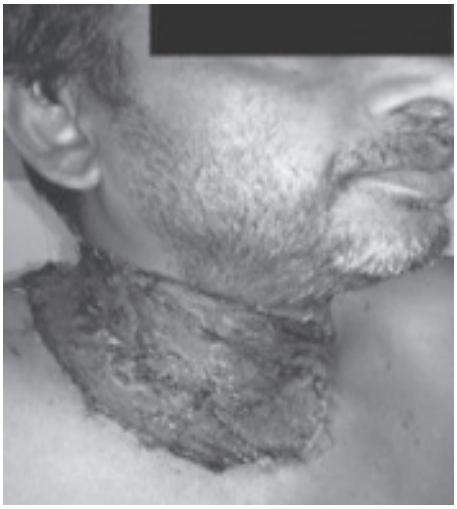

Fig.-6: *On 14th Post operative day.*

Discussion

DFSP comprises roughly 0.01% of all malignant tumours and approximately 2 to 6 percent of all soft tissue sarcomas.[3] It affects almost every age predominantly in adults (20-50 years). Genetic analysis has shown that virtually all cases of DFSP have a translocation that involves chromosomes 17 and 22, resulting in fusion of the collagen 1 alpha 1 gene and platelet derived growth factor B genes.[4]

Clinically the appearance of the tumour depends on the stage of the disease. Early it presents as a cutaneous pink to reddish, painless, trophic and or sclerotic plaque-like mass that develops into lumpy nodular and over time into ulcerative hemorrhagic protuberant tumor. It develops superficially, mobile upon palpation as it is adhered with its overlying skin, but not with its underlying tissues. Unfortunately, fixation to deeper structures such as fascia and muscle may present in the later stage of the tumour. Microscopically The DFSP has a characteristic histologic appearance of uniform spindle cells arranged in a storiform or "herring bone" pattern.

Since it is a slow growing tumor, delay in diagnosis and clinical misdiagnosis of the initial lesion is not uncommon. So, excisional or incisional biopsy is the diagnostic method of choice. MRI is used to evaluate local extension for the preoperative planning of large tumors and computed tomography is useful only when underlying bone erosion is suspected.

DFSPs show an extremely aggressive tendency to invade local surrounding tissue. Standard therapeutic approach used for the treatment of this tumor is wide and deep local excision, including the underlying fascia. It is generally agreed that 3-5 cm lateral and deep margins are adequate for the local control of the disease.[5] In order to achieve negative resection margins and simultaneously preserve the uninvolved tissue from resection.[5]Mohs Micrographic Surgery or staged wide excision (with formal histopathological sectioning and delayed reconstruction for complete circumferential peripheral and deep margin assessment) is becoming the standard surgical treatment for DFSP.[6] The use of radiotherapy in the treatment of DFSP has been investigated in

many studies.[7] It is particularly encouraged if resection is inadequate. The application of imatinib for DFSP has been limited and its precise role in DFSP is currently under investigation in many clinical trials.[8] In conclusion, DFSP is a rare tumour and clinicians must suspect it if there is a painless, cutaneous and multilobulated lesion. It can usually be well managed with wide local excision as a single modality or if indicated combined with radiotherapy.

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