

## Dermatofibrosarcoma Protuberans: an unusual case of neck swelling

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### Abstract

Dermatofibrosarcoma protuberans (DFSP) is a relatively intermediate to low grade malignant tumour with high proclivity for local recurrence if excised inadequately. It is a locally aggressive tumour and despite sharing some histological features with fibrohistiocytic tumours, it tends to grow in a more infiltrative manner. We are reporting this rare tumour in a 30-year-old woman where the diagnosis of DFSP was confirmed histologically and by positive immunomarkers at immunohistochemistry.

**Keywords:** Dermatofibrosarcoma protuberans, Recurrence, Radiotherapy.

### Introduction

Dermatofibrosarcoma protuberans (DFSP) is a slow growing soft tissue neoplasm with intermediate to low grade malignancy.<sup>1</sup> It originates in the dermis, with a tendency to invade the surrounding tissue. This infrequent tumour typically presents as a solitary or a multilobulated, slowly growing, and a painless cutaneous mass with

possible elusive features.<sup>2</sup> 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.<sup>3</sup> Trunk, proximal extremities, head and neck regions are most commonly involved.<sup>4</sup> 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.<sup>2</sup>

### Case Report

A 30-year old female presented with right sided neck swelling for 10 months duration with progressive increase in size for the last two months. The swelling was localized at the level of anterior surface of lower part of



Figure-1: Photograph of neck showing multiple nodular swellings at lower neck.

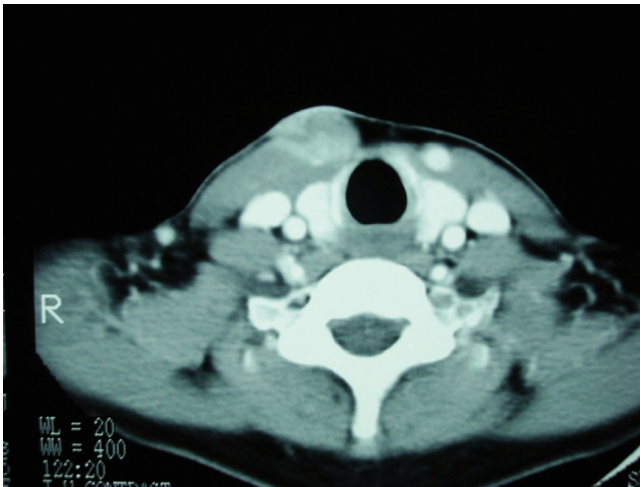


Figure-2: CT scan neck, axial cut, demonstrating heterogenous lesion anterior to sternocleidomastoid muscle involving skin.

sternocleidomastoid muscle (Figure-1). The lesion was about 4x3 cm painless, non-discharging, 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 un-remarkable.

CT scan revealed a heterogeneously enhancing lesion anterior to right sternocleidomastoid muscle (SCM), involving skin and subcutaneous tissue without significant lymphadenopathy (Figure-2).

Fine needle aspiration cytology (FNAC) showed spindle cell neoplastic lesion, decision for wide local excision was taken. Intra-operatively right SCM was found to be firmly adherent to the tumour, sparing the strap muscles. The tumour was excised with safe margins along with overlying skin and involved part of SCM. Frozen sections were sent, turned out to be negative. Histopathologic studies showed uniform population of spindle cells arranged in storiform pattern and exhibiting pleomorphism with increase mitotic activity. These spindle cells also showed infiltration into the muscle at the periphery and perivascular lymphocytic infiltrate. Immunohistochemical studies showed tumour cells positive for Vimentin and CD34.

The patient was then referred to radiation oncology services, and was treated with 68 Gy in 35 fractions. Post radiotherapy she developed symptoms of hypothyroidism for which oral thyroxine was started as a replacement. She is on regular follow up for past 2 years and has presented no evidence of local recurrence.

## Discussion

DFSP comprises roughly 0.01% of all malignant tumours and approximately 2 to 6 percent of all soft tissue sarcomas.<sup>4</sup> The DFSP usually has a long slow indolent course, with early tumours appearing as painless areas of cutaneous thickening. They may have pink, dark red or even bluish discoloration, particularly at its periphery. Over a period of time, they develop into a larger nodular mass, and ultimately can develop into a large fungating lesion. When they grow into the epidermal layer of the skin, they may eventually ulcerate. Unlike tumours of the subcutaneous tissue, DSPF is adherent or intimate with its overlying skin. Typically it is not adherent to underlying structures, with most tumours being superficial and less than 5cm in size at time of diagnosis.<sup>5</sup>

Typical presentation is in early or middle adult life with a slow growing soft nodular cutaneous mass on the trunk, although any part of the body may be involved. Given the indolent growth and long preclinical duration, it has been proposed likely that many of these tumours appearing in young adulthood, actually begin during childhood. 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.<sup>6</sup>

The DFSP has a characteristic histologic appearance of uniform spindle cells arranged in a storiform or "herringbone" pattern. Early lesions may demonstrate a "Grenz zone," which is a tumour-free region separating the tumour from the epidermis. Unusual variants of DFSP include the Bednar tumour that is denoted by melanin-containing cell, myxoid DFSP that contains areas of interstitial mucin, and the atrophic type. Immunohistochemical analysis can be utilized to aid in the diagnosis. Staining for Vimentin and CD34 is commonly employed, and sensitivity has been reported as being between 84 and 100 percent.<sup>7</sup>

Surgery is the mainstay of treatment and wide excision with a safety margin equal or more than 2 cm is recommended with an emphasis on clear margin for local recurrence.<sup>8</sup> Although it is a time consuming technique, Mohs micrographic surgery has been advocated by many professionals as a favourable resection option.<sup>9</sup>

The use of radiotherapy in the treatment of DFSP has been investigated in many studies.<sup>10</sup> It is particularly encouraged if resection is inadequate. Currently, there is limited objective data to support its routine use; however, successful application and recommendations have been reported in very few small series.<sup>11</sup>

Imatinib mesylate was designed to treat Philadelphia chromosome positive leukemia (chronic myelogenous leukemia). The application of imatinib for DFSP has been limited and its precise role in DFSP is currently under investigation in many clinical trials.<sup>12</sup>

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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