DERMATOFIBROSARCOMA PROTUBERANS – CASE REPORT

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ABSTRACT
Dermatofibrosarcoma protuberans (DFSP) is a rare type of skin cancer which is locally aggressive. Slow growing fibrohistiocytic neoplasm considered of low to medium malignancy. It starts in connective tissue cells in the middle layer of skin (dermis) and poorly circumscribed usually involves dermis and subcutis. DFSP might look like a pimple or feel like a rough patch of skin at first. It is a rare tumor with an incidence rate of 0.8 to 4.5 cases per million persons per year (2,3,4,5,6). Represents less than 5% of all soft tissue sarcomas. DFSP occurs most often in adults in 3rd to 5th decades of life but has been reported in all age groups, including congenital presentation. Tumour is usually positive for CD-34. Treatment consists of surgical resection with negative margins. In cases where surgical resection with negative margins is not possible, radiation and systemic therapy with tyrosine kinase inhibitors, such as imatinib mesylate, has been shown to be effective. Regular follow up of the patients is essential.
Keywords: Dermatofibrosarcoma protuberans, fibrohistiocytic neoplasm, surgical resection, negative margins, imatinib mesylate

Introduction

A uncommon, slow-growing fibrohistiocytic tumour categorised as low to medium malignancy is dermatofibrosarcoma protuberans (DFSP). Adults in their 20s and 30s are most at risk. The affected areas are typically the trunk, proximal extremities, head, and neck, and usually manifests as an asymptomatic bluish or brownish erythematous multinodular plate. Adults in their second to fifth decades of life are affected. Although distant metastases are uncommon, local recurrence following inadequate resection is frequent (1). (2).

Case presentation

A 45-year-old female was admitted in our department with chief complaints of swelling over left shoulder since 6-7 years (figure1). The patient reported an increase in size of the swelling in the last 1 year. Patient has history of colour changes of the skin over the swelling. No history of loss of weight, similar swellings. No significant family history.

On physical examination, a 4x3 cm swelling present over the left shoulder, 3cms medial to acromian process and 2cms below and lateral to the clavicle. Swelling is firm in consistency, non-tender with well-defined margins. The skin over the swelling is red compared to surrounding skin. There were no palpable cervical or axillary lymph nodes.

Soft tissue ultrasonography was done, which is suggestive of a fairly well defined heterogenously hypoechoic lesion of size 42.7x22.2x38.4 mm is noted inferior to the lateral one third of left clavicle showing few hyperechoic patchy areas within. Few densely echogenic foci, closely packed together are noted with the lesion. There is evidence of internal vascularity on colour doppler. Overlying skin shows no breach or discontinuity. No obvious deeper extension could be assessed. The above features suggestive of neoplastic etiology.

In core needle biopsy section studies shows fragmented tissue biopsy showing benign spindle cells arranged in fascicular pattern and interlacing bundles. These areas are hypercellular. No atypical features seen. The above findings suggestive of benign spindle cell lesion most likely schwannoma (neurilemmoma).

After obtaining anaesthetic fitness, patient posted for wide local excision with 2cms margin (figure 2 and 3). Excised specimen sent for histopathology. On gross examination, cut surface is greyish-white, smooth, firm mass noted measuring 5x3x2cms. Microscopically-showed spindle shaped cells arranged in storiform pattern. The calls have elongated nuclei with round ends with coarse chromatin. Occasional mitosis is noted. There is no evidence of necrosis. All surgical margins are free of tumor. CD-34 – positive.

PET-CT was done postoperatively suggestive of low grade metabolic active soft tissue stranding involving the subcutaneous and myofascial planes of left proximal arm, shoulder and supraclavicular region likely post operative changes. Advised for interval follow up. No evidence of metabolic active disease in rest of the body. Patient is regular followup once in 3 months.

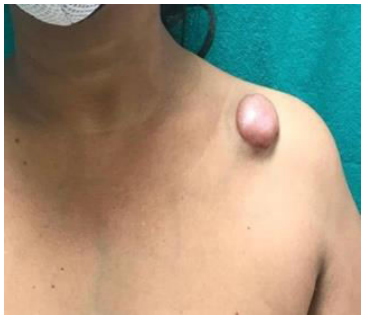

Figure 1

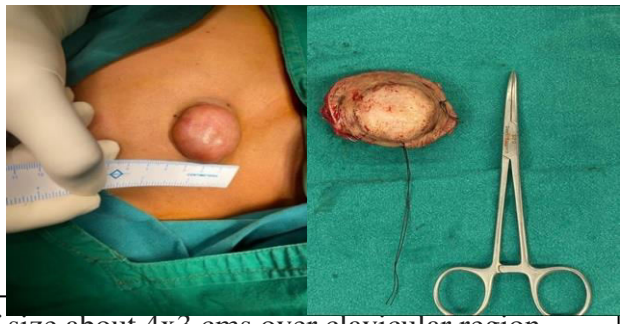

Figure 1: showing swelling of size about 4x3 cms over clavicular region.
Figure 2: showing intraoperative photo of the swelling.
Figure 3: showing wide local excision of the swelling with 2cms margins.

Discussion

An uncommon and locally aggressive form of skin cancer is called DFSP. When poorly circumscribed, it typically involves the dermis and subcutis and begins in connective tissue cells in the skin's middle layer (dermis). At first, DFSP may resemble a pimple or feel like a rough area of skin. With an incidence rate of 0.8 to 4.5 instances per million people per year, it is an uncommon malignancy (2-6). is a small percentage of all soft tissue sarcomas—less than 5%. DFSP has been recorded in all age categories, including congenital presentation, however it tends to affect adults in their third to fifth decades of life. DFSP can experience rapid development during pregnant. Black individuals experience DFSP, and particularly Bednar tumour, more commonly. There is no recognised cause for DFSP. The most frequent locations for DFSP are the trunk (40–50%), chest and shoulders (30–40%), proximal parts of limbs (10–15%), head and neck (10–15%), and toes, scalp, breast, and vulva (rare locations).
According to studies, a chromosomal translocation produces the fusion protein COL1A1-PDGFB, which stimulates the growth of tumours by producing too much PDGF. Over 90% of DFSP have the chromosomal translocation present. Rarely does DFSP develop into high grade sarcoma (1,4,5).
DFSP often begins as an indurated plaque with skin colour to red brown that is asymptomatic and later transforms into numerous elevated violaceous to red brown nodules. Their growth is sluggish. The tumour is frequently misdiagnosed, especially in its early stages when it can mimic a keloid or dermatofibroma. Some of them may develop painful ulcers as they get bigger. Rarely, between 1% and 4% of instances, distant metastasis develops, typically after numerous local recurrences. Lung is the most frequent location of metastasis due to hematogenous dissemination. Rarely are relational lymph nodes affected. A higher incidence of local recurrence (14%–52%) and distant metastasis (8%–29%) seen in the fibrosarcomatous type of DFSP.
Dermatofibroma, solitary fibrous tumour, spindle cell lipoma, keloid, angiosarcoma, angiomyoma, spindle cell melanoma, myxoid sarcoma, and peripheral nerve sheath tumour are examples of differentiations of DFSP.
The best course of treatment for DFSP is Mohs micrographic surgery (MMS), a surgical procedure that enables thorough margin assessment and tissue preservation due to the condition's propensity for a subclinical expansion. As an alternative, extensive local excision can be used to treat DFSP. Adults with metastatic, recurring, and untreatable DFSP are currently licenced for treatment with the chemotherapy drug imatinib mesylate by the FDA.

The tumour may be resectable, as in our instance, and there will be no recurrence or metastasis. Wide local excisions are performed with unrestricted margins. Patient was so instructed to have routine follow-up. Because recurrences are known to happen, all patients require ongoing imaging examinations. Patients who have entirely removed lesions have a fair prognosis. (7)

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