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Dermatofibrosarcoma Protuberans- A Case Report

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is well differentiated, primary fibrosarcoma of the skin. These tumours are slow growing, and locally aggressive and can recur, they rarely metastasize. It presents typically at mid-adult life with a slight male predominance. The trunk and proximal extremities are the most frequent locations of the disease. It accounts for <0.01% of all malignancies and <0.1% of all the cutaneous neoplasms, with reported incidence being 0.8 cases per million-persons. Radical wide local excision of the tumor along with surrounding area and subcutaneous tissue is the mainstay of treatment to avoid local recurrence. We report a case of DFSP over left popliteal fossa.

INTRODUCTION

Dermatofibrosarcoma protuberans-(DFSP) is a rare, slow growing, fibrohistiocytic neoplasm with intermediate to low grade malignancy, that commonly occurs in young to middle aged adults.[1] DFSP is locally invasive and rarely metastasizes. It is most commonly seen on the trunk followed by the proximal extremities and less commonly in head and neck.[2] DFSP has an incidence of only 0.8 cases per million and it rarely progresses to a high-grade fibrosarcomatous component. 1% to 4% of patients will develop distant metastasis, typically many years after the development of the initial lesion.[3] It was first recognized by Taylor in 1890 and described by Darrier in 1924, but the term Dermatofibrosarcoma was coined by Hoffman in 1925.[4]

According to the NCCN Clinical Practice Guidelines in Oncology, The gold standard treatment is complete surgical excision with appropriate reconstruction. Imatinib is approved for the treatment of advanced disease. [5]

CASE REPORT

28year male presented with complaints of a painless swelling in the back of left knee joint since 2 years, which was initially small (2X2cm) and gradually increased in size to 8x6cm.There was blackish discolouration over the swelling. Swelling became prominent on extending the knee.

There was no history of previous trauma, surgery or discharge from the swelling. The mass was adherent to the skin and was free from the underlying structures.

On examination swelling measured 8× 6 cm and was cystic in consistency. Fluctuation test was positive and transillumination test was negative. FNAC of the lesion was done and smears studied were cellular and showed benign spindle cells which were in groups and in singles, few having mild hyperchromatic nuclei associated with stromal matrix. Features were suggestive of spindle cell tumor. In view of increased cellularity and mild hyperchromasia, excisional biopsy was advised.

Excision biopsy of the swelling was done and the specimen was sent for histopathology. On gross examination, specimen consisted of skin covered tissue measuring 5.5x5x2 cm. Cut surface showed a well demarcated grey white to grey brown tumour mass with areas of haemorrhage. It was lobulated and homogenous. Microscopy showed structure of skin. Within the dermis there was an ill defined tumor made up of spindle cells(figure 1), arranged in storiform pattern, having scant bipolar cytoplasm and hyperchomatic nuclei. Mild pleomorphism with few mitotics were also seen. In between these cells were entrapped eccrine glands and subcutaneous adipocytes, giving it a honey comb appearance (figure 2&3). Mixed inflammatory cell infiltrate was also seen. Features were suggestive of dermatofibrosarcoma protuberans.

Immunohistochemistry showed immunoreactivity to vimentin (figure 4) , while non reactive to SMA and cytokeratin(figure 5&6).

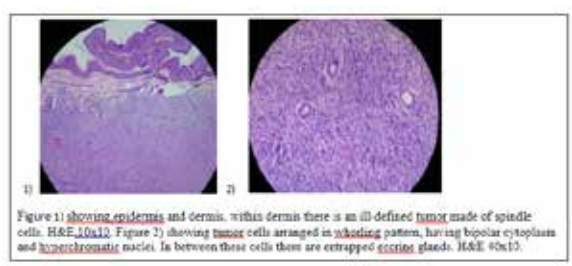

Figure 1) showing epidermis and dermis, within dermis there is an ill-defined tumor made of spindle cells. H&E,10x10. Figure 2) showing tumor cells arranged in whorling pattern, having bipolar cytoplasm and hyperchromatic nuclei. In between these cells there are entrapped eccrine glands. H&E 40x10.

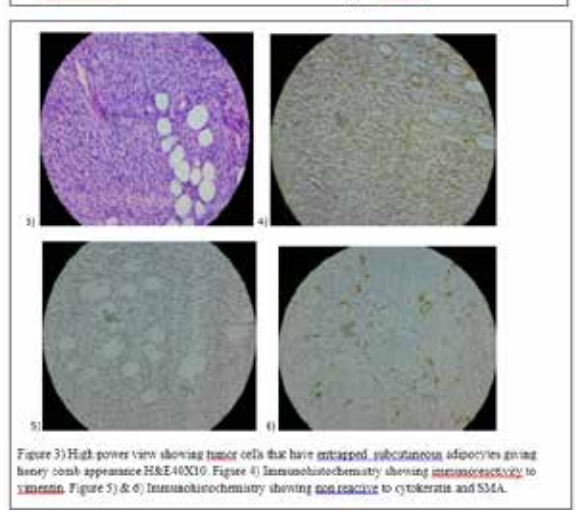

Figure 3) High power view showing tumor cells that have entrapped subcutaneous adipocytes giving honey comb appearance H&E40X10. Figure 4) Immunohistochemistry showing immunoreactivity to vimentin. Figure 5) & 6) Immunohistochemistry showing non reactive to cytokeratin and SMA.

DISCUSSION

Dermatofibrosarcoma protuberans is a rare tumour of the skin, which comprises 1% of soft tissue sarcomas and less than 0.1% of all malignancies. It is a malignant mesenchymal tumor originating from the dermis.[6] The exact cause is not well known. Predisposing factors include genetic mutation of the p53 gene, exposure to ionising radiations, post burn and other scars and exposure to certain carcinogens have been documented.[7]

A genetic link has been found in some patients with DFSP, which frequently (≥ 90%) exhibits translocation of chromosomes

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17 and 22, t(17;22).This rearrangement fuses the collagen, type I, alpha 1 (COL1A1) gene to the platelet-derived growth factor β chain (PDGFβ) gene. The resultant rearrangement causes unregulated expression of PDGF leading to constitutive activation of the platelet derived growth factor receptor (PDGFR).This step is believed to be a critical event in DFSP tumorigenesis.[1,8] No evidence of hereditary or familial predisposition exists.

Dermatofibrosarcoma is considered to be a pathologist's diagnosis more than that of surgeons. The relatively infrequent occurrence of DFSP lessens its clinical awareness and diagnosis is often made at histology.[2] Initially these cases present as a discrete asymptomatic plaque with reddish-brown or pink appearance with irregular borders, mimicking hemangioma and may be confused with localized scleroderma or solitary fibromatosis.[8,3,11] The usual pattern is an initial painless slow growing tumour. It may remain indolent for many years and sometimes grow to a very large size (more than 5cm in diameter over a few years). It may ulcerate but more commonly the patient intervenes one way or the other.[7,12] Four early clinical variants of DFSP can exist. These are: i) confluent nodular lesions forming a sclerotic plaque, ii) keloid like sclerotic plaque, iii) tumor-like, and iv) atrophic plaque form.[9]

The lesion is primarily located in the dermis with irregular infiltration of the subcutaneous fat in a lace-like pattern. The epidermis is usually spared, but can be hyperplastic. The lesion usually comprises fairly uniform spindle cells with elongated nuclei and scanty pale cytoplasm. Pleomorphism is minimal or absent. The cells are typically arranged in a storiform or a mat-like pattern.[1,6,9] Melanin pigment has been reported in some cases. This group is usually referred to as Bednar tumour and Bednar tumour with widespread metastasis has also been reported.[10] Immunohistochemical studies reveal positivity for CD34 that differentiates DFSP tumour cells from normal stromal cells and dermatofibroma. DFSP generally stains positive for CD34 and negative for S-100 and factor XIIIa16.[2,8]

The standard treatment of DFSP is radical wide local resection of tumor with surgical margin of 2-3 cm and three dimensional resections including skin, subcutaneous tissue and underlying fascia. Tyrosine kinase inhibitor imatinib has shown to induce regression in advanced DFSP where completely negative surgical margins are difficult to obtain.[2,3,7,8] Differential diagnosis of DFSP include dermatofibroma, atypical fibroxanthoma, malignant fibrous histiocytoma, myxoid liposarcoma and fibrous hamartoma of infancy.[2,12]

CONCLUSION

DFSP is a rare soft tissue tumor of cutaneous origin with an intermediate level of malignancy. Histopathology plays a pivotal role in the diagnosis which has prognostic significance. Wide local excision with pathologically negative margins plays a significant role in preventing local recurrence and distant metastasis. In young individuals, DFSP must be differentiated from fibrous hamartoma of infancy, a lesion that shares similar growth patterns and cytological features. Although metastasis rarely occurs but high rate of recurrence may occur so special attention has to be given to these patients even after surgery.

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