Case Report
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Myxoid Dermatofibrosarcoma Protuberance Presented as a Mass over Right Paraspinal Area of Back

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Abstract

Dermatofibrosarcoma protuberance is a histiocytic tumor of intermediate malignant potential. We are presenting a case of myxoid variant of dermatofibrosarcoma protuberance in a 37 year female patient having large 15 x 6 x 5 cm soft tissue mass on back region, over right paraspinal area of 8 years duration which suddenly increase in size in last 3 months. This case of dermatofibrosarcoma protuberance myxoid variant is presented for its rarity and clinico-histopathological findings.

Keywords: Soft Tissue Tumor; Fibrohistocytic Tumor; Histological Types.

Introduction

It is relatively uncommon, locally aggressive cutaneous tumor having high rate of local recurrence, but low risk of metastasis [1-3]. DFSP was first described in 1890 by Taylor [4], and Sherwell [5] independently. DarieranfFerrand termedit as a "progressive recurring dermatofibrosarcoma" which is a nodular cutaneous tumor with characteristic prominent storiform pattern [6]. Although DFSP has regarded as a low grade sarcoma, few cases have shown sarcomatous transformation.

Case Report

A 37 years female presented with history of insidious growing nodular cutaneous mass over back region at the right paraspinal area of 8 years duration. Initially small in size of 1 cm of long duration but within 3 months showed sudden increase in size and has reached to 15 x 10 cm in size.History of mild pain at the site, with increase in pigmentation over the skin was noted. Patient is unable to sleep on back. No history of trauma, local surgery or any systemic illness was there. No contributory family history was there. On radiological investigation show soft tissue mass on back, ?malignant.

Fine needle aspiration cytology of the mass was inconclusive and revealed only blood along with scattered spindle cells and myxoid material. Patient underwent wide local excision of the mass with 3 cm surgical margins. She was advised regular follow up.

Histopathology

Gross examination – We received specimen of excised growth from upper back, totally measuring 15 x 15 x 6 cm covered with skin. Skin showed multiple protrubences. Cut section showed multiple, variable sizes, grey white, firm subcutaneous nodular masses. The tumorwas grey white, nodular with area of translucency (Figure 1,2). Tumor was seen invading subcutaneous tissue and overlying skin. There was no evidence of haemorrhage, necrosis or cyst formation.

Microscopy

Multiple sections from the tumor mass showed a tumor composed of uniform population of slender fibroblasts arranged in distinct storiformpattern (Figure 3). In areas, tumor cells are in sheaths and in small clusters. Prominent area of Myxoid change is noted (Figure 4). The tumorcells showed mild

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pleomorphism, hyperchromasia with 3 mitosis per 10 high power fields (Figure 4). There was no area of haemorrhage and necrosis. On histopathology diagnosed as, DFSP – Myxoid variant. Immunohistochemistry (IHC) was CD 34 positive.

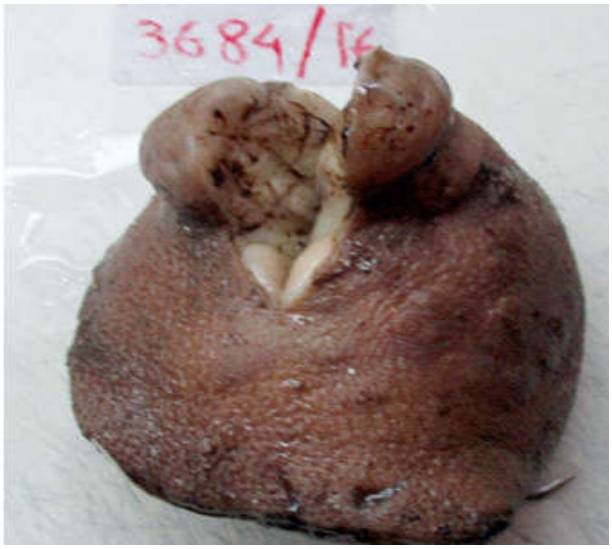

Fig. 1: Gross specimen of excised growth from upper back, totally measuring 15 x 15 x 6 cm covered with skin

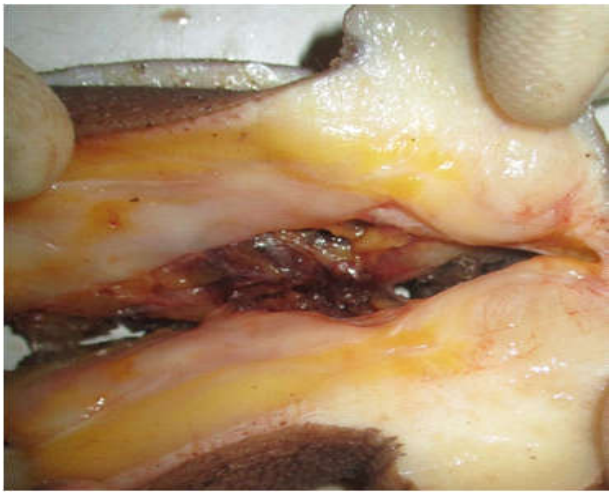

Fig. 2: Cut section showed multiple, variable sizes, grey white, firm subcutaneous nodular masses

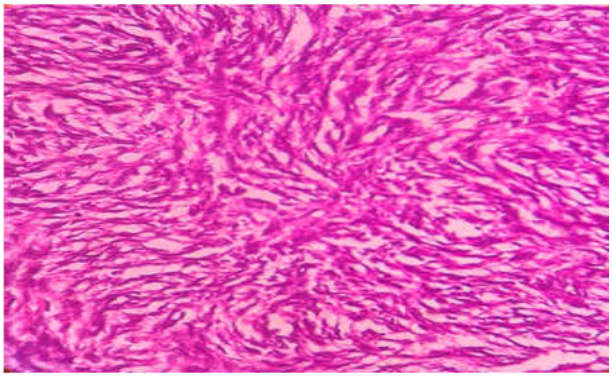

Fig. 3: Microphotograph of tumor composed of uniform population of slender fibroblasts arranged in distinct storiform pattern (H & E stain, 400 X)

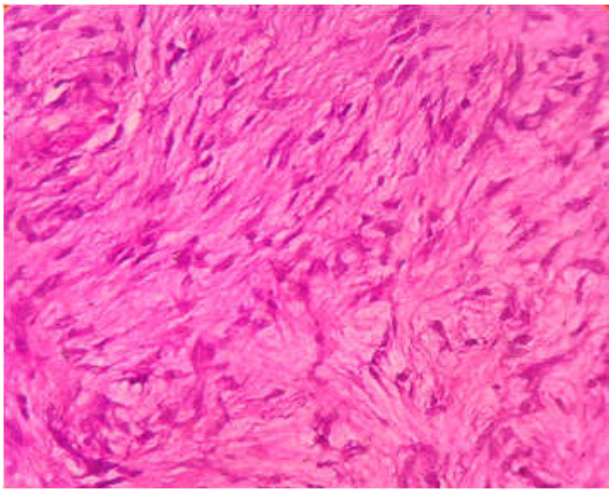

Fig. 4: Microphotograph of tumor cellsshowing mild pleomorphism, hyperchromasia and prominent myxoid change. (H & E stain, 100 X)

Discussion

In details histopathological characteristics of DFSP were described by Talyor HB and Helwig [7]. In his descriptionmentio the neoplasm was characterised by fibroblastic growth appearing as a low grade sarcoma having tumor cells arranged in fascicles with spiral or cart wheel pattern. Generally tumor cells show little or no pleomorphism, low mitotic rate. In WHO classification DFSP is referred as superficial, low grade, locally aggressive fibroblastic neoplasm. Clinically DFSP presents as nodular, slowly growing lesion but usually affects younger to middle aged adults . At the initial stage of disease history of trauma, burn or surgical scar is often found. Tumor in advanced stage may ulcerate, bleed or become painful .Tumor size may range from 2 -5 cm, but few cases showed upto 20 cm in diameter. Tumor is usually fixed to overlying skin but not to deeper structures. In our case, tumor was lager size which showed rapid increase in size in 3 months the underlying muscle and bone was free from tumor .

On histopathology DFSP were type into classical (Conventional) form, Pigmented, Myxoid, Plaque like, Atrophic, Sclerosing, Granular cell variant and DFSP with sarcomatous transformation.

In our case, it is of DFSP myxoidtype.It is considered when DSFP with more than 50% myxoid stroma [8,9]. For myxoid variant it should be differentiated form other conditions like myxoidliposarcoma [9], high grade myxofibrosarcoma [10], fibrous histiocytoma, signet cell fibroblastoma etc. In myxoidliposarcomatumor cells are bland, stellate or fusiform with prominent vasculature and presence of lipoblasts seen. For myxofibrosarcoma it occurs

commonly in subcutaneous tissue of the extremities of elderly patient. Histopathologically, myxofibrosarcoma is uniformly myxoid, lacks alternating fibrous areas and always has in greater degree of nuclear hyperchromasia with nuclear pleomorphism [10]. In case of giant cell fibrous histiocytotoma it is mainly seen in infants and children with painless nodules. Microscopy shows loosely arranged spindle cells with hyaline, myxoid area with giant cells.

On immunohistochemistry, DFSP is characterised by consistent and diffuse CD 34 immunostain positivity. The DSFP is a locally aggressive tumor that recurs upto one half of patients [11]. It is related to infiltrative nature of tumor. The risk of recurrence is 41% when excision margin is less than 2 cm. A gold standard practice is complete prompt wide local surgical excision. The Myxoid variant of DFSP have similar prognosis as classic DFSP.

Conclusion

Myxoid variant of DFSP is a rare fibrohistocytictumor of intermediate malignancy. We are presenting this case for its rarity, clinical behaviour and histological finding. It should differentiated from other soft tissue tumor with Myxoid change.

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