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Original Research Paper

Pathology

HIGH GRADE SARCOMA ARISING OUT OF DERMATOFIBROSARCOMA PROTUBERANS: A CASE REPORT OF A RARE OCCURRENCE.

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ABSTRACT Dermatofibrosarcoma protuberans (DFSP) is an intermediate grade soft tissue neoplasm. It has the potential of reccurence. Rarely it can transform into highly aggressive soft tissue neoplasm like Fibrosarcoma. We would like to report one such case of rare occurrence of high grade sarcoma arising in the Backdrop of DFSP. Histopathological examination forms the cornerstone in confirmatory tissue diagnosis of such an entity.

KEYWORDS : DFSP, sarcoma, histopathological examination

INTRODUCTION:

Dermatofibrosarcoma protuberans (DFSP) is a rare intermediate grade mesenchymal tumor[1]. It is a locally aggressive tumor with limited metastatic potential[2]. DFSP transformation into fibrosarcoma(DFSP-FS) is exceptional[3] ,while cases with fibrosarcomatous transformation have an approximately 15% metastatic rate[1]. Through clinical observation and this case report we can understand the epidemiologic, clinical, histological, and evolutionary characteristics of the disease.

Case-Report:

A 65 year male presented with a recurrent soft tissue swelling (8cm X 5cm X3 cm) over the back and had a history of similar swelling (6.5cm X 4 cm X 2.5cm) at same site which was surgically excised 2 years ago. A diagnosis of Dermatofibrosarcoma protuberans(DFSP) was made on histopathological examination (fig1) and IHC (Fig 2). Biopsy of the recurrent mass shows fungating growth on gross appearance with variegated areas on cut section .On microscopic examination, presence of both spindle cells arranged in storiform pattern as well as fascicles of pleomorphic spindle cells (Fig 4) at periphery with a transformation zone (Fig 3) are noted. Increased number of mitotic figures and necrosis are also seen. This shows transformation towards Fibrosarcoma. The high grade sarcomatous area turned out to be negative for CD34 on immunohistochemistry (Fig 5).

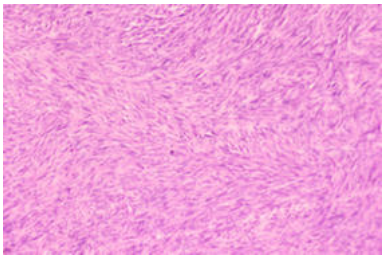

Fig 1 Spindle cells in storiform patterns in previous lesion. (H&E, 10X)

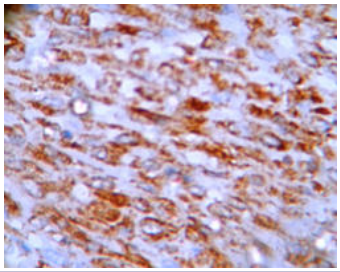

Fig 2: IHC For CD 34 Positive (40X)

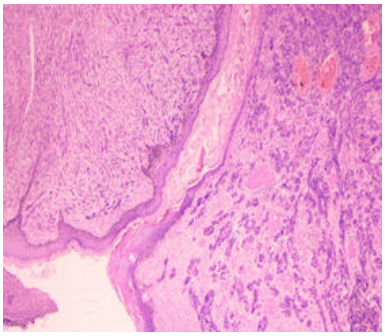

Fig-3: Transformation zone in present lesion. (H&E ,10x)

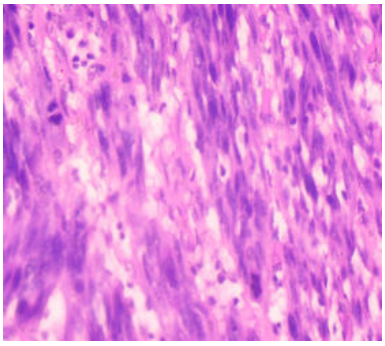

Fig- 4: Pleomorphic spindle cells in fascicles in present lesion. (H&E ,40X)

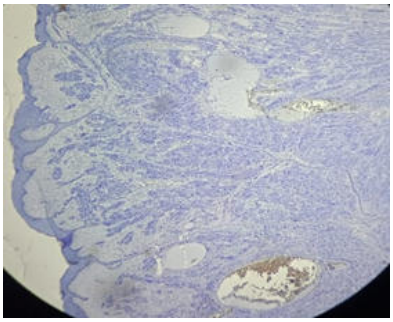

Fig 5: CD 34 negative in present lesion. (IHC ,10X)

DISCUSSION: Cutaneous sarcomas represent <1% of all cancers in humans. The DFSP is most commonly seen in the young adult between the age 20 and 40 years but there is a discrete male prevalence[4]. The transformation of a DFSP into

a Fibrosarcoma is exceptional; until now and since the first description of a metastatic DFSP by Penner in 1953, only 100 cases of FS-DFSP were reported[1]. The DFSP occurs in the trunk (47%), the upper limb (38%), or the cervico-cephalic area (15%)[3]. Genetically DFSP is a result of translocation of chromosome t(17;22)(q22;q13) in 90% of cases. It leads to fusion of the genes COL1A1 (collagen type 1 alpha 1) on chromosome 17q21-22 and PDGF (Platelet Derived Growth Factor Beta) on chromosome 22q12.The tumor cells of the DFSP express the CD34 in 90% of the cases[5] .The DFSP-FS is defined histologically by the presence in a cellular DFSP of ranges pointing towards an FS, the transition is abrupt or gradual. Cell areas are characterized by a fascicular architecture, pleomorphism, necrosis and high mitotic activity. Transformation into fibrosarcoma with CD34 negativity have been reported in only a few other studies in the past.

CONCLUSION:

The DFSP is a rare tumor, its diagnosis is mainly histological, and its treatment is surgical. Very rarely transformation of DFSP to a high grade sarcoma can occur Expression of CD34 becomes negative in the transformed sarcoma.

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