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Fibrosarcomatous Variant of Dermatofibrosarcoma Protuberans - A Case Report

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Abstract: ***Dermatofibrosarcoma protuberans is a fibroblastic tumour of intermediate malignant potential that can rarely metastasize. Fibrosarcomatous variant of DFSP is a rare but aggressive variant with higher rates of recurrence, more propensity to metastasize and increased mortality. We present a case of 54 year old male patient who presented with a swelling in the occipital region.***

1. Introduction

Dermatofibrosarcoma Protuberans is a fibroblastic tumour, which can be locally aggressive, hence considered to be of intermediate malignant potential. Microscopically, it has a storiform appearance. This tumour may acquire a metastatic potential with a morphological appearance of a fascicular or herringbone pattern which is termed as the Fibrosarcomatous variant of Dermatofibrosarcoma Protuberans (FS - DFSP).1

2. Case Report

A 54 years old male patient presented with a swelling in the nape of neck since 7 years with progressive increase in size since 5 months. The swelling was excised and sent for histopathological examination and a diagnosis of Dermatofibrosarcoma Protuberans was given. One year later, the patient presented with recurrence of the swelling at the same site. A wide local excision was done and specimen sent for histopathological examination.

On Gross examination, the soft tissue was skin covered and measured 7.5x4x1.5 cm. On serial sectioning, there was a well circumscribed, well capsulated lobular growth measuring 1x0.8x0.7 cm which was reaching up to the deep resection margin. Circumferential resection margins were grossly free.

Microscopic examination of the soft tissue mass revealed a well circumscribed lesion in the deep dermis infiltrating into the underlying subcutaneous tissue. The tumour predominantly comprised of uniform slender spindle cells arranged in storiform pattern. The spindle cells had elongated, hyperchromatic nuclei, inconspicuous nucleoli, and moderate amount of eosinophilic cytoplasm with indistinct cell borders. Occasional mitotic figures were seen (2/10HPF). These histological features were suggestive of a predominant component of Dermatofibrosarcoma protuberans. However, few areas showed an abrupt transition into areas revealing sweeping, intersecting fascicles of spindle cells with ovoid to elongated hyperchromatic nuclei, prominent nucleoli, and moderate amount of eosinophilic cytoplasm, occasional tumour giant cells and frequent atypical mitotic figures. (10/10 HPF). These features were suggestive of a sarcomatous component. The circumferential skin resection margins were free from tumour, however, the soft tissue resection margin and the deep resection margin showed tumour involvement.

On, Immunohistochemistry, the areas with DFSP showed CD 34 positivity whereas CD 34 was diminished in areas of the fibrosarcomatous variant. The fibrosarcomatous variant also showed increased immunoreactivity to p53 as compared to the conventional DFSP areas.

A diagnosis of Fibrosarcomatous variant of Dermatofibrosarcoma Protuberans was given.

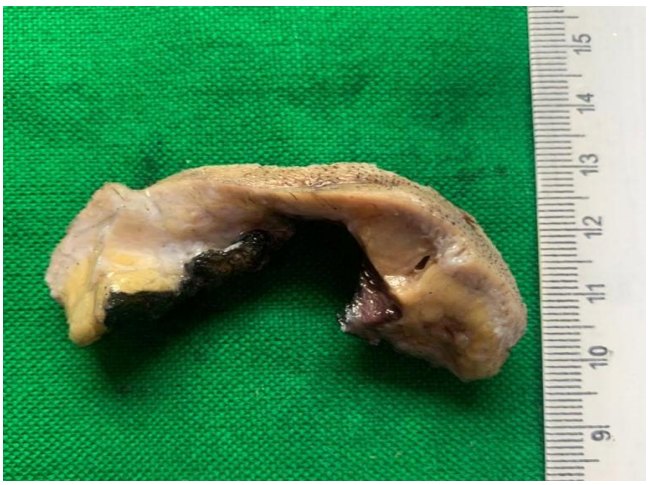

Image 1: Grossly showing a well circumscribed grey white tumor beneath the skin.

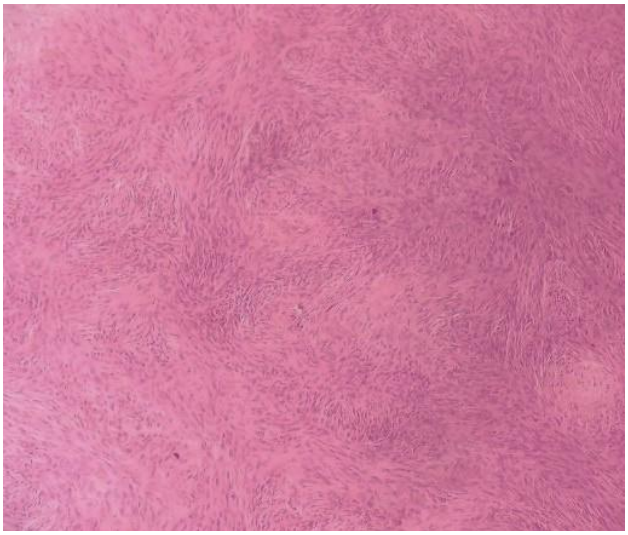

Image 2: The tumor predominantly comprised of uniform slender spindle cells arranged in storiform pattern. (Low Power)

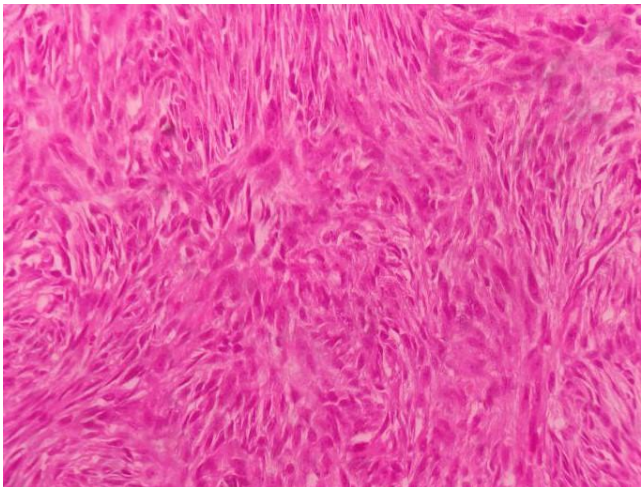

Image 3: The tumor cells are spindle cells having elongated, hyperchromatic nuclei, inconspicuous nucleoli, and moderate amount of eosinophilic cytoplasm. (High Power)

3. Discussion

Dermatofibrosarcoma Protuberans is a fibroblastic tumour with intermediate malignant potential. Microscopically, it is composed of uniform spindle cells arranged in a storiform pattern. Five percent of cases of DFSP show progression to FS - DFSP. The fibrosarcomatous variant can either occur de novo or in local recurring cases and the transformation may be abrupt or gradual. The fibromatous variant of DFSP is a rare but aggressive variant with a propensity to metastasize, most commonly to lungs. The following features are required to diagnose a sarcoma arising in DFSP. The sarcomatous component should constitute at least 5% to 10% of the tumour. The spindle cells have a higher nuclear grade and are arranged in a fascicular pattern. There is increased mitotic activity. As compared to DFSP, CD 34 is diminished. The FS - DFSP areas show a higher MIB - 1 labeling index and increased p53 staining.2

The most common fusion seen in DFSP is *COL1A1* - *PDGFB*. The genetic events which lead to the progression of FS - DFSP in DFSP are under study. Alteration in the Akt/mTOR signaling pathway in FS - DFSP has been reported by Park et al [3] and Hiraki - Hotokebuhi et al[4].

Various studies have been done on the prognostic significance of the sarcomatous variant of DFSP. In a study done by Mentzel et al[5] on 41 cases of FS - DFSP, 2 cases showed progression to pleomorphic sarcoma, local recurrence in 20 patients, and metastasis in 5 patients. Abbot J et al [6] studied the prognostic impact of FS changes in DFSP. Out of the 41 patient in his study, 8 patients had local recurrences, 4 patients (10%) had metastases, and 2 patients died of disease.

Liang C. A. et al compared the outcome of dermatofibrosarcoma protuberans with and without fibrosarcomatous changes and they concluded that risk of metastasis and death is elevated in DFSP - FS as compared to DFSP.[7] Rasheed A. A. et al analysed cases of locally advanced or metastatic DFSP. Fibrosarcomatous transformation was present in 10 out of 14 cases with a decreased progression free survival.

4. Conclusion

The Fibrosarcomatous variant of Dermatofibrosarcoma Protuberans is an aggressive variant signifying tumour progression with increased risk of metastasis and mortality.

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