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DERMATOFIBROSARCOMA SCALP – A MALIGNANT TURBAN OVER HEAD

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Article Info

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Abstract

Dermatofibrosarcoma protuberans is a rare fibrous tumour of skin which is slow growing and locally aggressive. DFSP is a malignant tumour which is characterized by high probability of local relapse with low metastatic potential. With less than 5% of the tumors involving scalp these tumors are frequently misdiagnosed and locally excised with insufficient margins. Wide local excision of tumour with 2-5cm of adequate margin is modality of treatment of dermatofibrosarcoma. We are reporting a case of 70yr male with bleeding scalp lesion. After thorough work up, wide excision with cover of defect with local transposition flap was done. There was no recurrence in Post-operative follow up of six months.

Key words: Dermatofibrosarcoma, Scalp, Transposition flap.

Introduction: -

Dermatofibrosarcoma protuberans is a rare tumour of skin which frequently involves the dermis with a tendency of local recurrence,[1] It is a fibrous tumour which is slow growing and locally aggressive. Metastasis to regional lymph nodes and distant sites is rarely seen. Most common involved site of DFSP is trunk, followed by extremities, with the head and neck affected less frequently, [2] Less than 5% of these tumors are located over the scalp. DFSP scalp are usually noticed because of its ugly effect on patient's appearance. Microscopic examination is mostly the diagnostic of tumour. Since excision of the tumour has excellent prognostic outcome, early recognition of this tumour is important ensuring adequate margin of 2 to 5cm. Here we are

reporting a case of dermatofibrosarcoma scalp in a 70-year-old male along with its modality of treatment.

Case Report: -

A 70-year-old male presented with a large swelling involving entire occipitoparietal region, which is gradually increasing in size. (Figure01 and 02) The swelling was examined and found to be soft to firm, non-tender, non-pulsatile and non-transilluminant. The skin overlying the tumour was stretched with vessels seen through it with some areas of skin erosion with bleeding on touch. Impulse on coughing was absent. There were no signs of any bony involvement in X-ray Skull. CT Scan showed a heterogeneously extra-calvarial lesion with mixed density overlying left parietal and occipital region extending on either side with peripheral enhancing and central necrotic component. There was also indentation but no erosion of underlying bone noted. Biopsy report came out to be suggestive of dermatofibrosarcoma protuberans. Microscopically, there was cellular spindle cell lesion with fascicular growth pattern, nuclear hyperchromatism with mild nuclear atypia and increased mitosis. Metastatic workup was negative.

Patient was admitted after required medical and surgical fitness. Patient was operated under general anaesthesia. Wide local surgical excision of tumour was done with 2cm margin from indurated area. Excision was achieved up to periosteum which was seen uninvolved by the tumour. Skin defect was covered with transposition flap from fronto-temporal area. Donor area was covered with split thickness skin graft. Post-operative period was uneventful. Patient was discharged after one week and was followed up every month. Biopsy report showed all margins free from tumour cells with specimen showing highly cellular tissue with spindle cells having blunt nuclei and eosinophilic cytoplasm showing mild to moderate atypia with interlacing fascicles (typical storiform pattern)

consistent with diagnosis of Dermatofibrosarcoma protuberans. [Figure 03] Tumour cells also stained positive for CD-34 and negative for S-100 and desmin, [3] After six months follow up, no signs of recurrence were noted. [Figure 04 and 05]

Legends of Photographs: -

Figure 01-Preoperative Photo [Anterior View]

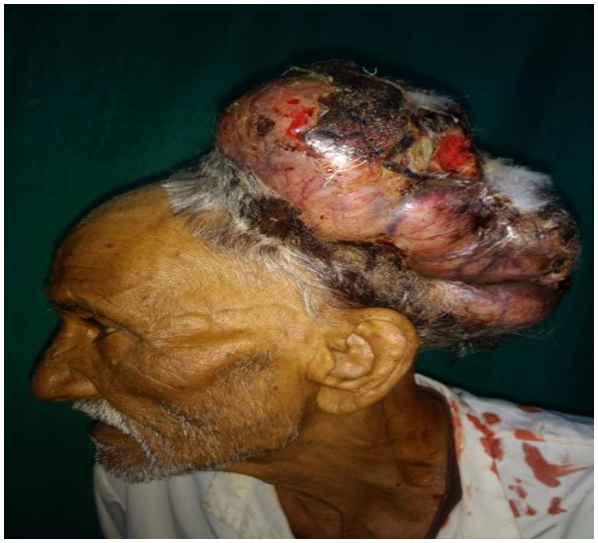

Figure 02- Preoperative Photo [Lateral View]

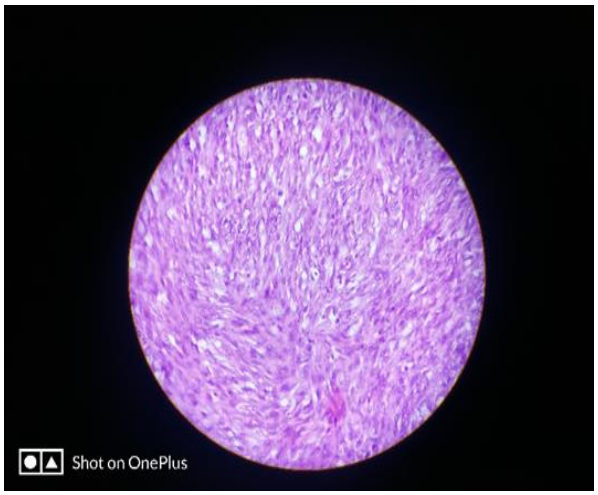

FIGURE 03 showing highly cellular tissue with spindle cells having blunt nuclei and eosinophilic cytoplasm showing mild to moderate atypia with interlacing fascicles (typical storiform pattern).

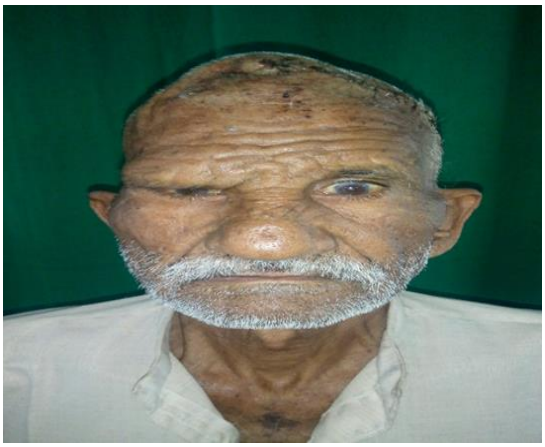

Figure 04 Postoperative Photo 1 Month Follow Up (Anterior View)

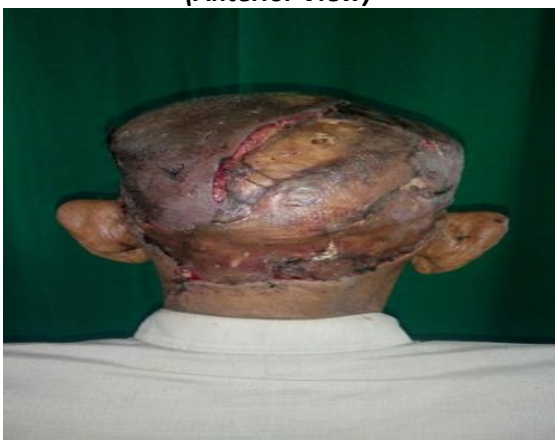

Figure 05 Postoperative Photo 1 Month Follow Up (Posterior View)

Discussion: -

DFSP is a malignant tumor which is locally aggressive tumour which is characterized by high probability of local relapse with low metastatic potential. Although metastasis is seldomly reported, before operation, investigations like chest X-ray and ultrasonography are done to exclude metastasis. On Histopathology it shows uniform grouped fusiform cells, with elongated nuclei in characteristic storiform arrangement. The factors associated with high recurrence rates are location over head and neck, histological subtype, cellularity, size, and high mitotic rate. The treatment of choice for DFSP is surgical resection with adequate margin followed by reconstruction, [4] Extent of initial surgical resection is considered as an important factor for relapse. Wide local excision of tumour leads to a lower rate of recurrence, but the term "wide local excision" is not well defined. Most surgeons consider margins of 2- 4 cm wide adequate for excision. Apart from wide local excision, Mohs micrographic surgery has also emerged as an attractive option with margin control approach,[5] DFSP is also radiosensitive and radiation therapy is only used in the setting of extremely large or recurrent tumours. In addition to radiotherapy, Imatinib chemotherapy is also effective in cases of DFSP recurrence and in cases that are unresectable.

Conclusion: -

A successful management and treatment of dermatofibrosarcoma depends on complete wide local excision of tumor with negative margins. Proper treatment of dermatofibrosarcoma scalp is very often delayed due to misdiagnosis and local excision,[6] Hence surgeons should be known about this very rare entity and should always perform a wide local excision of these tumors to decrease the risk of recurrence.

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