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CASE REPORT OF MERKELCELL CARCINOMA

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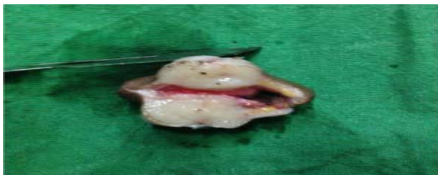
Abstract :
Merkel cell carcinoma (MCC) is an uncommon and aggressive primary neurocutaneous neoplasm that lacks distinguishing clinical features. More than half of Merkel cell carcinomas (MCCs) occur in the head and neck of elderly people in areas of actinically damaged skin. Here we present a case of merkel cell carcinoma of forehead in a 32 year old male patient for whom preoperative biopsy was reported as merkel cell carcinoma. We did a wide local excision biopsy, and the resected specimen showed tumour originating from merkel cell and IHC study also confirmed it.

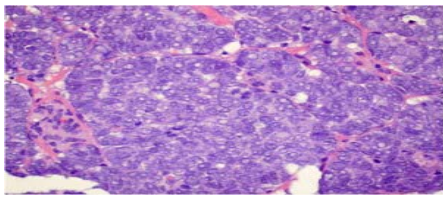
Keyword :
Merkel cell carcinoma, neuroendocrine neoplasm

A 32 year old male patient came with complaints of swelling in the right side of forehead for the duration of about 3 months. Initially it was small in size and progressively increased in size. History of dull aching pain was present. On examination a 3X2 cm sized, ovoid, violaceous nodule was present on right side of forehead 5cm above the right eyebrow, extending medially 3 cm from midline, and above upto hairline, with well defined margin, irregular surface with central umblication ,dilated veins and restricted mobility. Other systemic examinations were normal.

clinical picture
Skin biopsy showed features of merkel cell carcinoma.
Wide local excion of lesion with 2cm margin clearance was done.



specimen
Histopathological study showed aggregates of small, round tumor cells within the dermis. The configurations were broad sheets to trabeculae with focal spindling of cells in fascicles . The cytoplasm is scant, and the nuclei were oval with evenly distributed chromatin and inconspicuous nucleoli . All margins were negative, No lymphovascular invasionwas made out. IHC study showed NSE positive.



HPE
Wih these impression merkel cell carcinoma was confirmed. Since the lesion was >1.5cm patient was subjected to Radiation therapy - 45Gy over 5 weeks. Patient was on regular follow up and had no recurrence for the last 1year.

REVIEW OF LITERATURE
Merkel cell carcinoma (MCC), sometimes referred to as a neuroendocrine carcinoma of the skin, arises from the uncontrolled growth of Merkel cells in the skin. It mostly presents in sun-exposed skinperiorbital region of head&neck, arms and lower limb. Males are mostly affected, most comman age group being >65 yrs. The exact causes of MCC are idiopathic. Factors strongly associated with the development of MCC are age over 65 years, fair skin, chronic extensive sun exposure, chronic immune suppression . The most common genetic abnormality is

deletion of the short arm of chromosome 1 (1p36). Loss of heterozygosity has been observed in chromosome 3 , as well as in chromosomal arm 10q and chromosome 13. A new virus called Merkel cell polyomavirus (MCPyV) was found in 8 of 10 tumors tested, and it was associated with the DNA of the tumor cells Clinically it presents as painless, firm, red, blue or flesh-colored swelling, which grows rapidly breaking down the overlying skin. Lymphnode may be involved. ,Diagnosis made by **Light microscopy** is occasionally useful alone, because **66% of MCC** like many other undifferentiated small cell neoplasm, **will be misdiagnosed by using this alone** Histologic classification: 1.Intermediate cell type(50%),2.Trabecular 25% ,3.Small cell type 25% . Electron microscopy can be used to confirm the diagnosis of MCC, when needed. The most characteristic findings are perinuclear bundles of intermediate filaments and electron-dense neurosecretory granules . These features virtually confirm the diagnosis of MCC**.** Definitive diagnosis is based on the presence of antibodies to cytokeratin (CK),8,18,19, 20 (in the form of a perinuclear dotlike pattern typical of neuroendocrine tumors) and Neuron-specific enolase,[these 2 are present in almost 100%]and Neurofilament protein. CK19 experession is increasd in tumors positive for the Merkel cell polyomavirus and may help in diagnosis of CK20-negative tumors. CT may reveal regional nodal involvement, or systemic involvement with metastases to the lung, bone, liver, etc. , MRI accurately identifies metastases to soft tissue sites, brain, and bone marrow and should be performed if there is any suspicion for central nervous system involvement.

Treatment

Stage I &II: Wide local excision with 2-3cm margin +/- ELND +/- RT **ELND indicated in**: LN positivity[+ in 55% of pts.

If 1) >10mitosis/HPF]

2) presense of lymphatic invasion

3) tumours composed of small cell 4) midline lesion .

Radiotherapy: MCC is radiosensitive tumor, 45-50GY, if margin + 65GY over 5weeks. Radiotherapy is associated with a statistically significant improvement in local and nodal recurrence, but not in survival . Indicated for: 1) Primary tumor >1.5cm 2) Positive margin 3)Margin <2mm 4)Evidence of lymphovascular, perineural invasion 5)Stage III& IV Stage III : WLE with 2-3cm margin +modified radical neck dissection + RT to both primary site & nodal basins Stage IV: palliative RT +/- Chemotherapy CT agents: mainly **Cyclophosphomide, doxorubicin**, others 5FU, Bleomycin, vincristin, etoposide. But these do not improve survival .

CONCLUSION:

MCC is a rare skin cancer. It is about 40 times less common than melanoma. MCC has the potential to be lethal, and thus prompt aggressive treatment is warranted. At the time of diagnosis, half of the patients had disease localized to the skin, while the other half had MCC that was no longer confined to the skin. Most MCCs are diagnosed when a skin biopsy is performed to rule out another suninduced skin cancer or a cyst. In the vast majority of cases, both the doctor and the patient are surprised by the diagnosis of MCC. Microscopic examination and IHC studies play a major role in diagnosis. Surgery is the treatment of choice for operable lesions, chemotherapy and radiotherapy play a role for inoperable lesions but not improve survival.

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