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RESEARCH ARTICLE

MERKEL CELL CARCINOMA: A CASE REPORT

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Abstract

Merkel cell carcinoma is a rare but highly aggressive skin tumor. MCC clinically represents as cutaneous or subcutaneous nodule, and in 30% of the cases loco-regional metastasis are already there in primary diagnosis. Pathogenesis is essentially due to excessive exposure to UV light. The confirmation diagnosis is based on histological and immunological features on the lesion. The treatment methods require a complete surgical excision and radiotherapy.

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Introduction:-

Merkelcellcarcinomais a rare but highly aggressive skin tumor. MCC clinicallyrepresents as cutaneous or subcutaneousnodule, and in 30% of the cases loco-regionalmetastasis are alreadythere in primarydiagnosis (1). Pathogenesis isessentially due to excessive exposure to UV light (2). The confirmation diagnosisisbased on histological and immunologicalfeatures on the lesion (1) .Thetreatmentmethodsrequire a completesurgical excision and radiotherapy (3).

Case Report:

A 77 yearold male with a medicalhistoryoftype 2 diabetes and high blood pressure treated for 20years, burned inbothlegs due to a fire 10 yearsago.

The patient presents to the hospitalwith multipleindurated nodules.

Physicalexaminationshowsmanylesions: the main one wascauliflower-shaped,white, measuring 5 cm in diameters on the inside of the right leg. Itwasassociated to othersmallviolatedones. (Figure 1)

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Figure 1:-

The other lesion was in the right calf, ulcerated polypoid cutaneous nodule measuring 4.5 cm in diameters. (Figure 2)



Figure 2:-

The recurrent lesion was biopsied from the first white nodule to confirm the diagnosis.

Pathology examination revealed, on the standard hematoxylin-eosin stain, dermal infiltration with small round cells, with high nuclear to cytoplasmic ratio and focal necrosis.

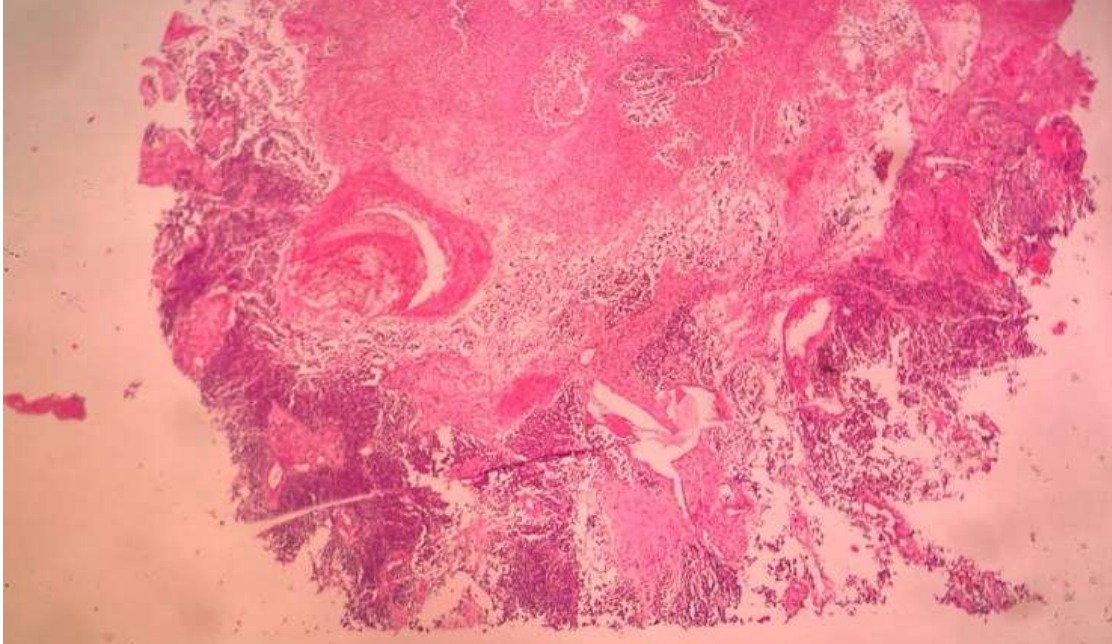


Figure 3: hematoxylin-eosin stain: Showing small round blue cells infiltrating the dermis, focal necrosis.

Expanded immunohistochemical panel revealed positivity with epithelial marker (CK20) and neuroendocrine markers (synaptophysin, chromogranin and CD56).

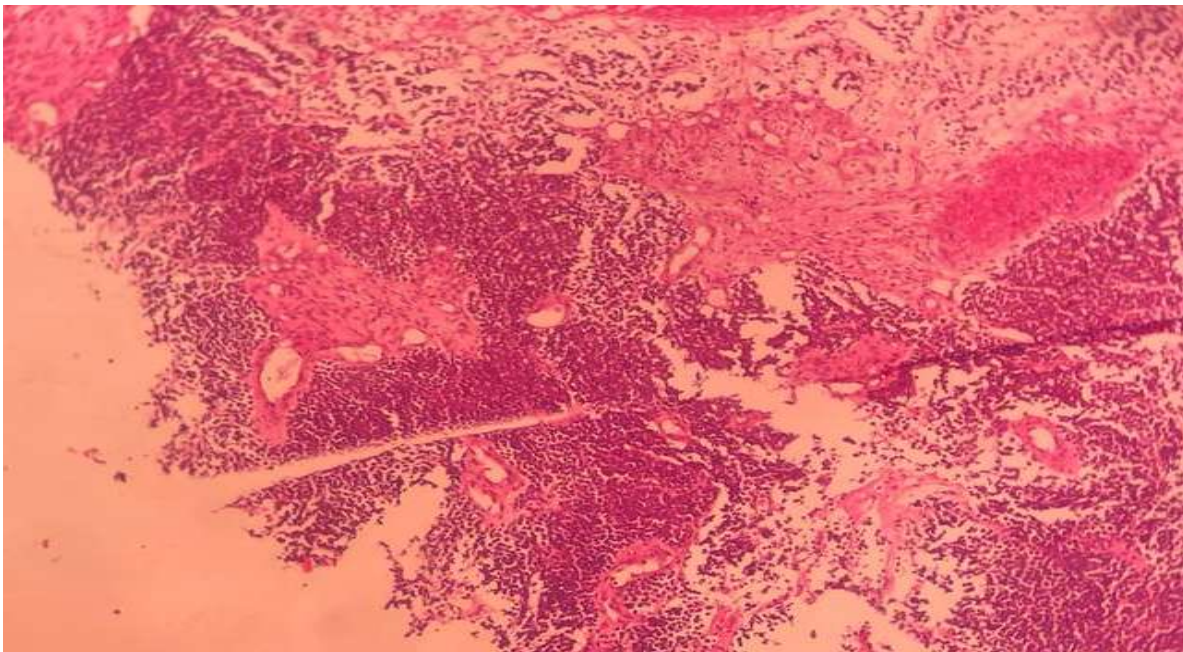


Figure 4:- Merkel cell carcinoma at medium magnification showing small blue cells have a high N/C ratio and hyperchromatic nuclei.

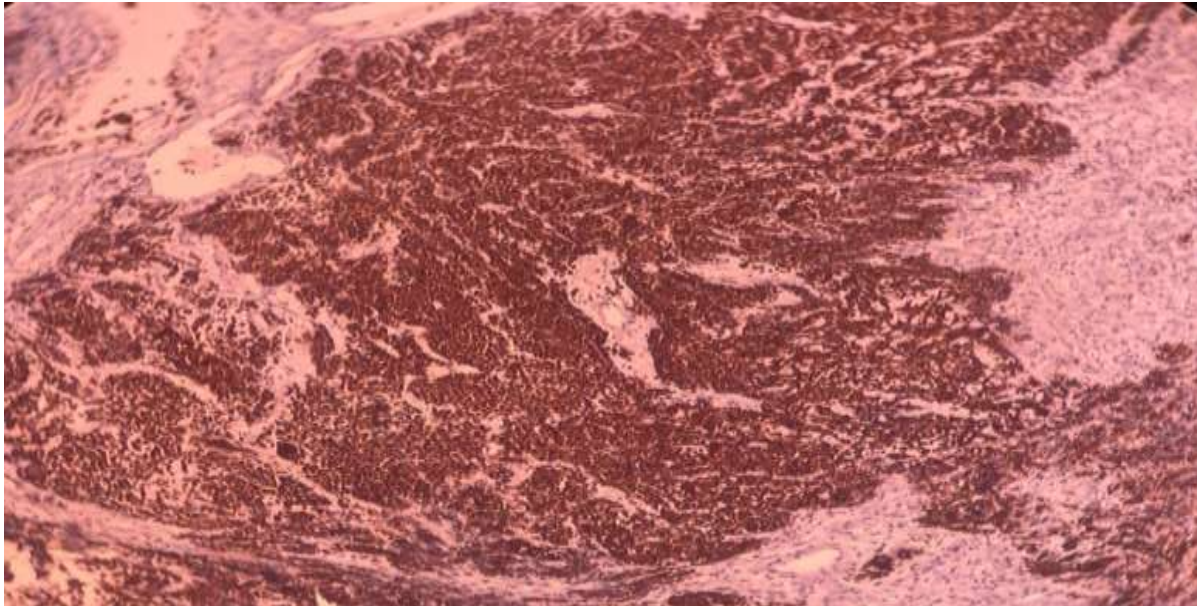


Figure 5: Diffuse cytoplasmic synaptophysin positivity.

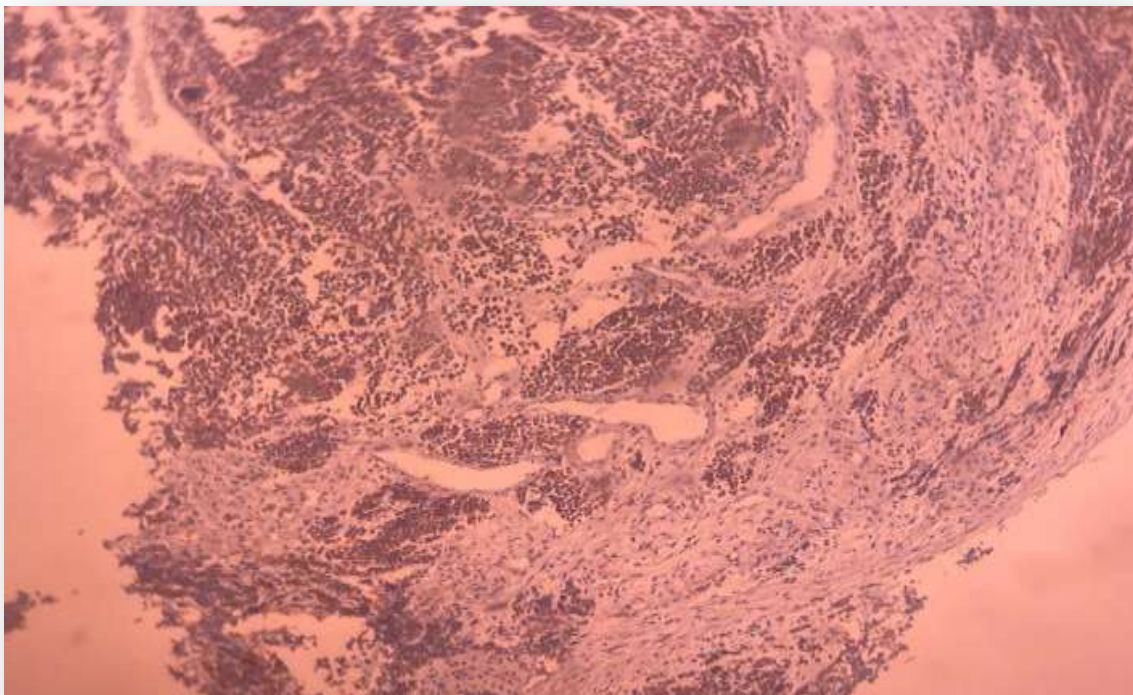


Figure 6:-Diffuse cytoplasmic chromogranin positivity in merkel cell carcinoma.

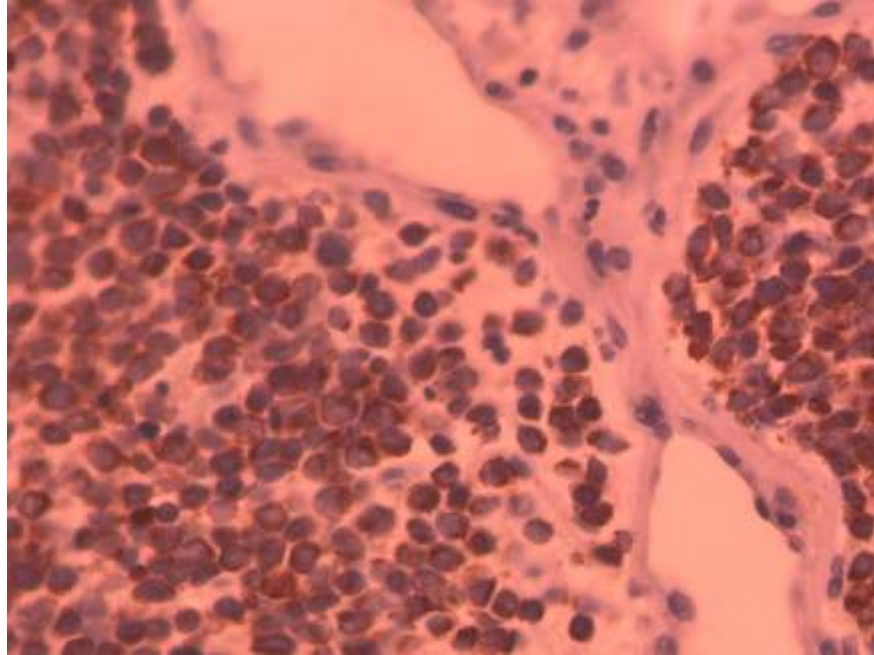


Figure 7:- Strong positivity for cytokeratin 20 (CK20) staining, with a dot-like perinuclear accentuation.

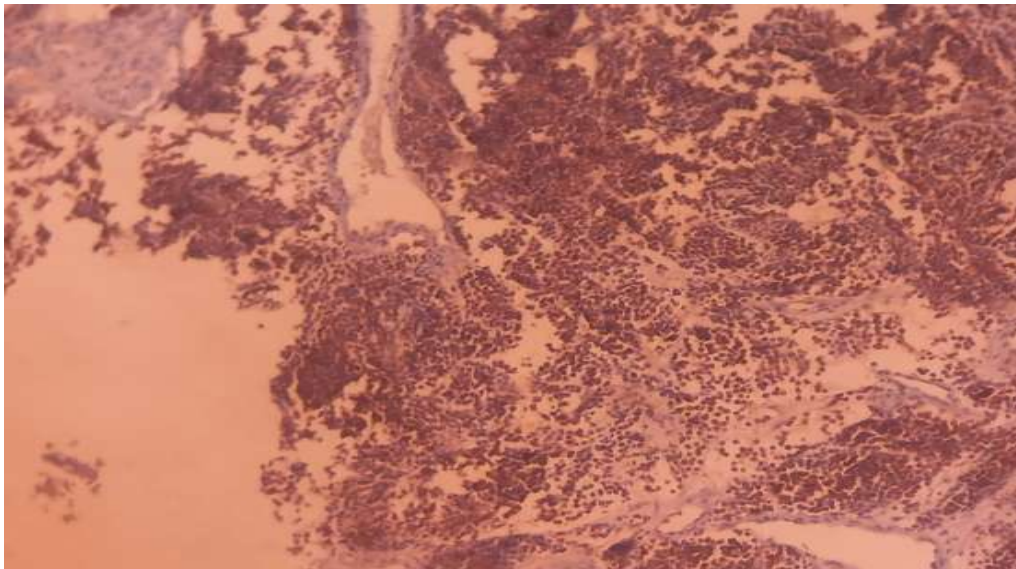


Figure 8:- CD 56 pattern showing strong positivity in neuro endocrine cells.

Discussion:-

Merkel cell carcinoma is a rare but highly aggressive tumor, it was named due to the resemblance between tumor cells and Merkel cells, which are present in the basal layer of the epidermis. (1)

MCC carcinogenesis suggests two main causes: the presence of clonally integrated Merkel cell polyomavirus, or chronic exposure to UV lights (1). In fact, UV exposure can also play a role in the viral cause by causing local immunosuppression (4).

Diagnostic indicators for the recognition of MCC can be described by the acronym AEIOU (A-asymptomatic; E-rapid expansion; I-immune suppression; O-over 50 years; U-UV-exposed skin). (2)

MCC presents as nodules on sun damaged skin in elderly, and it has a highly metastatic profile. (3) It is rapidly growing, cutaneous or subcutaneous nodules on sunexposed areas, (5,7) asymptomatic in most cases, that they can be confused with other benign lesions (inflammatory or cyst). (6)

However, clinical features are not enough to confirm the diagnosis, histological and immunochemistry analysis can confirm it. In standard hematoxylineosin stain, it presents as small blue round monomorphic cells with a high nuclear to cytoplasmic ratio and is composed of dermal and/or subcutaneous nodules or sheets (8)

There are three main types of MCC that has been described : small-cell, trabecular and intermediate but most cases present with overlapping features, and the classification of MCC according to these three variants does not have practical implications (1). Infiltrative (rather than circumscribed) growth pattern and the presence of lymphovascular invasion have been associated with increased risk of microscopic nodal metastases and a poor prognosis (1).

Immunohistochemical tumor cells mostly express cytokeratins, more specifically CK 20. And also neuroendocrine markers such as chromogranin and synaptophysin (1)

Differential diagnosis is basically with metastatic small-cell carcinoma, that also shows as small blue round cell morphology, but the positive staining for TTF1 and negative staining for CK20 helps to make the difference (1).

As for treatment, surgical **excision** with 1-2 cm **resection** margins is required, followed by adjuvant radiotherapy. (9) however larger excision for bigger lesions cause morbidity for the patients, such as loss of function and the need for skin graft, and this should be avoided, in that case some MCC treatment guidelines suggest only radiotherapy could be effective. (10)

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