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Merkel cell carcinoma of unknown primary site: A case presentation and review of the literature

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Merkel cell carcinoma (MCC) is a rare and highly aggressive neuroendocrine tumor of the skin. MCC was described first by Toker in 1972 as trabecular carcinoma of the dermis with high lymphatic metastatic risk. The incidence of this rare tumor is increasing rapidly; the American Cancer Society estimates 1500 new cases in the USA. Based on case reports, the risk factors include: ultraviolet exposure, Merkel Cell polyomavirus DNA infection, immunosuppression (HIV-patients, post transplant pts under immunosuppresive therapy).

MCC is characterized by a high incidence of early locoregional relapse and distant metastases. The clinical and pathologic diagnosis of MCC can be challenging, especially when it presents as nodal metastasis. As a "small round blue cell tumor," it can be difficult to differentiate from other small cell neoplasms of different primary origin. Diagnosis is based on typical histology representation on hematoxylin-eosin stained slides along with the results of immunohistochemistry. The tumour expresses both epithelial and neuroendocrine markers, so exhibits both epithelial and neuroendocrine differentiation.

Despite aggressive multimodality treatment, Merkel Cell Carcinoma outcome is primarily based on the stage of disease at presentation, with both increasing tumor size and lymph node positivity being associated with a worse prognosis. Moreover, the mortality rate of MCC is considerably higher than that of cutaneous melanoma.

Although it has been identified in various anatomical sites, LN metastatic MCC in the absence of a primary site is extremely rare and for this reason there is no standard approach to its management.

We report a case of a 39 year-old male, diagnosed with HIV infection 8 years ago, who presented to the hospital with an enlarged lymphnode in the left inguinal area, which revealed to be metastatic Merkel Cell Carcinoma in the absence of a primary skin lesion.

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