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Pathology

Merkel Cell Carcinoma of Skin - A Very Rare Case

Nanda Patil[1], Avinash Mane[2], Vaidehi Nagar[3*], Shivangi Mittal[3]

[1]Professor, Department of Pathology, KIMSDU, Karad, Maharashtra, India
[2]Assistant Professor, Department of Pathology, KIMSDU, Karad, Maharashtra, India
[3]Resident, Department of Pathology, KIMSDU, Karad, Maharashtra, India

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***Corresponding author:** Vaidehi Nagar
Resident, Department of Pathology, KIMSDU, Karad, Maharashtra, India

Abstract | **Case Report**

Merkel cell carcinoma is an uncommon primary neuroendocrine carcinoma of skin which affects elderly and immunosuppressed patients. The confirmatory diagnosis require histopathological examination as the clinical features are non - specific. Differentiation from metastatic neuroendocrine carcinoma requires clinicopathological correlation plays important role in differentiation. We present a rare case of merkel cell carcinoma. In a 68 year male patient who presented with exophytic growth arising from umbilicus with metastasis in inguinal lymph node.

Keywords: Merkel cell carcinoma, Neuroendocrine tumor, skin.

INTRODUCTION

Merkel cell carcinoma is a neuroendocrine carcinoma arising from the skin. It is highly aggressive neuroendocrine tumor and shows a tendency for local recurrence which varies in the elderly male patients [1]. The tumor shows distant metastasis and has very poor prognosis. We present a case of merkel cell carcinoma arising from umbilicus in a 68 year male patient to highlight the rarity and aggressive mature.

CASE REPORT

68 year male patient presented with exophytic growth on abdominal skin near umbilicus with history of bleeding since 10-12 days. The swelling was painless. On examination the mass was bluish in color measuring 4 x 3.5 x 2.4 cm. Patient has bilateral inguinal lymphadenopathy. There was no history of irradiation or immunosuppressive treatment. CT abdomen did not reveal other primary site. It revealed heterogeneously intervening soft tissue density lesion in the subcutaneous plane of umbilical region limited to linea alba and and no intraperitoneal extension. FNAC from Lymph nodes was positive for metastatic carcinoma. Incisional biopsy of swelling revealed features suggestive of Basal cell carcinoma. Complete excision of mass and bilateral inguinal lymph node dissection was done and specimen was sent to histopathological examination.

Histopathological Examination

The specimen of wide excision was received which revealed bluish color exophytic tumor measuring 4 x 3.5 x 2.4 cm, firm in consistency with all peripheral surgical margins were free from tumor. Cut section of tumor was grey white and homogenous.(Figure 1 and 2, Gross and cut section of exophytic growth in umbilical region.

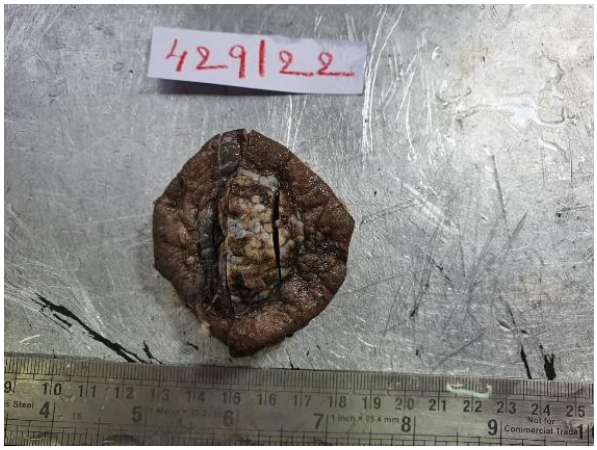

Figure 1

Figure 2

Microscopic examination of tumor reveals neoplastic cells arranged in the form of sheets, nests and trabeculae which were small with high N:C ratio, round to oval nuclei with finely dispersed salt and pepper chromatin. Tumor was seen located within the dermis and was reaching subcutaneous fat. Tumor reveal atypical mitotic figures. There was metastasis in right inguinal lymph node (Figure 3, 4 and 5). Immunohistochemistry reveals positivity for CK, Synaptophysin and negative for CK 20, TTF1, ISL1, PAX 8, PR. Considering these clinical and microscopic features the diagnosis was given as Merkel cell carcinoma of skin with metastasis in right inguinal lymph node.

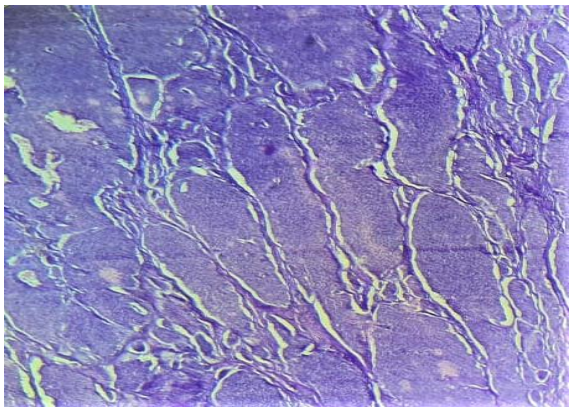

Figure 3: Tumor cells are arranged in sheets, trabeculae and nests, H and E stain, 100x

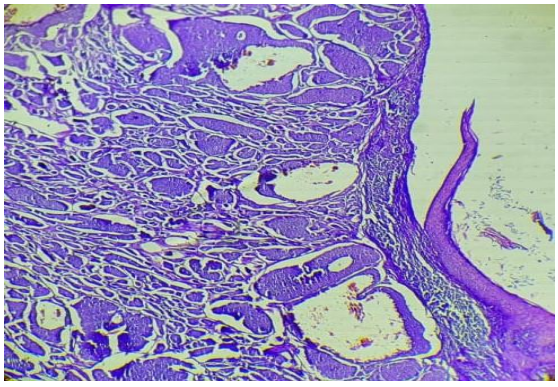

Figure 4: Tumor with superficial skin layer which is free from tumor, H and E stain, 100x

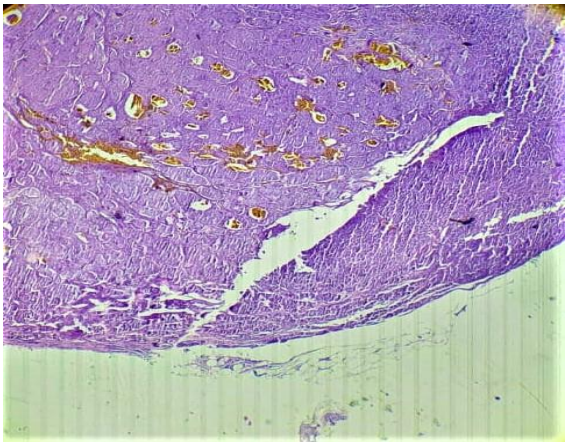

Figure 5: Metastasis in right inguinal lymph node, H and E stain, scanner view

DISCUSSION

Merkel cell carcinoma (MCC) is an uncommon, highly aggressive primary neuroendocrine carcinoma of skin. The tumor was first described by Toker in 1972 [2]. Tong and Toker revealed dense core granules in the cytoplasm of tumor cells by electron microscopy which led to its origin from Merkel cells [3]. De Wolff-Peeters proposed the rare Merkel cell carcinoma for this tumor [4]. Merkel cells are found in the basal layer of the skin forming mechanoreceptors and also function as neuroendocrine cells [5]. Risk of MCC increases in immunocompromised patients [6]. MCC presents as a rapidly growing painless nodule or plaque mostly from sun exposed areas of the body in elderly male patients. The nodule is solitary, firm with reddish blue color [1]. The tumor affects skin of head and neck in 50% cases, extremities in 40% and trunk and mucosa in 10% cases [7]. In our case the patient was elderly male and the tumor was located on abdominal skin near umbilicus. The clinical differential diagnosis are basal cell carcinoma, pyogenic granuloma, squamous cell carcinoma, amelanotic melanoma and adnexal tumor [1].

The tumor is seen in the dermis and infiltrates subcutaneous tissue. Neoplastic cells are small, round, monomorphic which show enlarged round nuclei with salt and pepper chromatin, prominent mitosis and arrangement in the form of trabeculae, solid nests or sheets, similar microscopic features were seen in our case. Sometimes MCC show differentiation. Basal cell carcinoma, squamous cell carcinoma or sweat gland tumor have been reported adjacent to MCC [1].

Immunohistochemistry of MCC shows both epithelial and neuroendocrine markers i.e. immunopositive for CK 20, synaptophysin, chromogranin and neuron specific enolase as it was seen in our case. Leucocyte common antigen differentiates MCC from lymphoma and S100 helps to distinguish MCC from melanoma [8]. Additional studies have proved that 80% of MCC contain merkel cell polyoma virus [9].

MCC are highly aggressive tumors with tendency for local recurrence. Lymph node involvement and lymph node metastasis. In our case, metastasis in inguinal lymph node was noted. Few of these tumors show spontaneous regression [10]. Treatment of MCC consist of surgery and radiotherapy.

CONCLUSION

MCC is a rare primary neuroendocrine carcinoma of the skin which is highly aggressive. Diagnosis reveals histopathological examination and clinicopathological correlation to rule out secondary deposits.

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