CASE REPORT

Merkel cell carcinoma is a primary small cell carcinoma of the skin that resembles oat cell carcinomas of the lung. This tumor has a predilection for head and neck region but may rarely occur as isolated vulvar tumor. It has aggressive malignant behavior and should not be missed while evaluating vulvar masses. We report a case of 45-year-old Mrs I, who presented with recurrent vulvar swelling and was diagnosed as stage III Merkel cell carcinoma of the vulva.

Synonyms: Primary neuroendocrine carcinoma, Trabecular carcinoma, Primary small cell carcinoma, Cutaneous apudoma.

Keywords: Vulvar carcinoma, Merkel cell carcinoma.

DISCUSSION

Vulvar cancer represents 3 to 5% of malignancies of the female genital tract. Approximately 90 to 92% of all invasive vulvar cancers are of the squamous cell type. Merkel cell carcinoma is a primary small cell carcinoma of the skin that resembles oat cell carcinoma of the lung. This tumor has a predilection for the head and neck region followed by the extremities and the buttocks, but may rarely occur as isolated vulvar tumor.

Merkel cell carcinoma is a relatively recently described entity, although the Merkel cell was described more than 100 years ago (in 1875, by Friedrich S Merkel). In 1980, the name Merkel cell carcinoma was first used. This tumor is a malignant proliferation of highly anaplastic cells which share structural and immunohistochemical features with various neuroectodermally derived cells, including cutaneous Merkel cells.

Clinical features are solitary, rapidly growing nodule, which are firm red, purple or skin colored. It is locally aggressive with local discontinuous and distant lymphatic spread. Histologically, small round blue cells are present in various growth patterns, out of which, a sheet-like growth is the most common. Characteristically, the edge of the tumor shows a trabecular infiltrating pattern. High mitotic rate, frequent single cell necrosis and often zonal necrosis are observed.

A “perinuclear dot”, pattern of cytokeratin-20 on immunohistochemical staining is essentially pathognomonic for Merkel cell carcinoma. S-100 is positive in most melanomas and negative in Merkel cell carcinoma.
According to American Joint Committee on Cancer, there are four clinical stages for MCC based on features, at time of presentation.\textsuperscript{1} Localized disease is either stage I (primary lesion \( \leq 2 \) cm) or stage II (primary lesion > 2 cm).\textsuperscript{1} Nodal spread of MCC is stage III, whereas patients presenting with metastatic disease beyond the local node bed have stage IV disease.\textsuperscript{1}

MCC shows aggressive malignant behavior.\textsuperscript{1} Treatment is challenging and controversial. Best outcome is obtained with multidisciplinary approach, including surgery, chemotherapy (cisplatin-based), immunotherapy and radiation therapy.\textsuperscript{1,3} In this case, recurrence can probably be explained by positive surgical margins and lack of chemo/radiotherapy.

Sentinel lymph node biopsy is also a sensitive test for detecting MCC spread to the lymph nodes.\textsuperscript{1}

Survival is highly dependent on the stage at presentation. Patients with truly local only disease have a greater than 90\% of chance of survival, which decreases to approximately 60\% with nodal involvement and below 10\% among those presenting with metastatic disease.

REFERENCES