MERKEL CELL CARCINOMA, A CASE REPORT AND LITERATURE REVIEW

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SUMMARY : Merkel cell carcinoma is a rare tumor that develops on or just beneath the skin and hair follicles. It occurs most often on the face, head and neck. Early detection and treatment are important because the disease can spread rapidly. Merkel cell carcinoma is difficult to cure once it spreads. Researchers believe that exposure to sunlight may increase risk of this disease. Herein, we report a 70 - year - old female with a painful, nodular skin lesion of right leg that enlarged rapidly. Total excision was done. Pathologic findings confirmed diagnosis of Merkel cell carcinoma.

Key words: Merkel cell carcinoma.

INTRODUCTION

Merkel cell carcinoma is a rare, malignant tumor of skin (1,2) characterized by significant incidence of local recurrence [30% to 45%], early involvement of the regional lymph nodes [40% to 70%] and distant metastases [30% to 50%] (3). The prognosis is worse than malignant melanoma (4). Due to its aggressiveness (3) and benign clinical appearance, the prognosis of this neoplasm is poor. Reported overall 5- year survival rates range from 30% to 64% (5,6).

Toker published the first complete description of this neoplasm in 1972, originally describing it as 'trabecular carcinoma' (7). Nearly 1000 cases are reported in the English literature since the initial description (8).

The tumor is also known as cutaneous small cell undifferentiated carcinoma. At present the tumor is classified as a neuroendocrine malignancy of the skin, generally occurring in elderly patients (9-11). It occurs, rarely, in children (12).

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The lesion is firm, nodular, and red-pink. They usually are non-ulcerated and range in size from 0.8 to 4 cm in diameter (1).

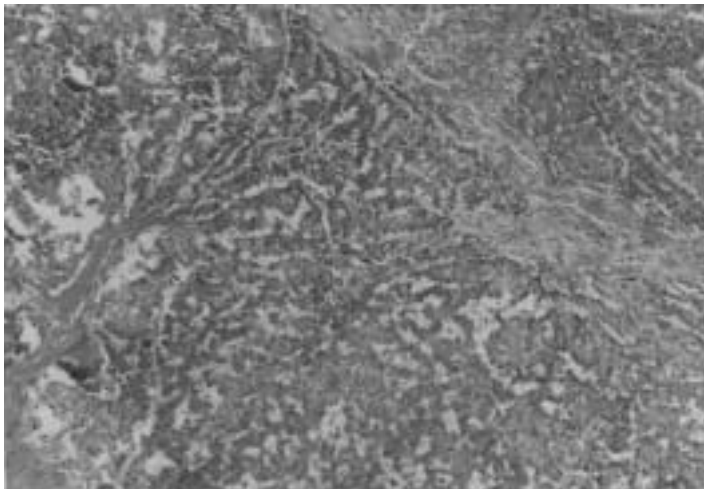
The commonest sites of presentation are the head and the neck and it is slightly more common in females (4,13,14).

Merkel cell carcinoma arises in the dermis and subcutaneous tissues from Merkel's cells located in the basal layer of the epidermis and expresses neuroendocrine markers such as neuron-specific enolase (NSE), chromogranin, synaptopysin and neurofilament proteins (1,4,15).

The most frequent sites of metastasis are distant lymph nodes, distant skin, CNS, and bone. The histologic diagnosis can be difficult, because with the conventional light microscopy Merkel cell carcinoma can be misdiagnosed as any other poorly differentiated small cell neoplasm (3). Immunohistochemical staining play an important role in the early diagnosis (2,16-19).

The most common staging system is that described by Yiengpruksawan *et al.*, stage I disease for isolated local lesion, stage II disease is characterized by metastatic spread to regional lymph nodes and stage III has evidence

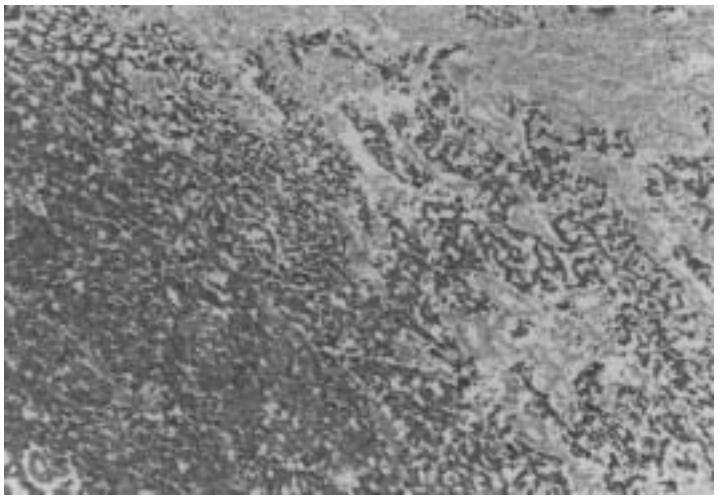
Figure 1: Merkel Cell Carcinoma (H and E * 40)



of distant metastatic disease at the time of initial presentation (20).

The management of Merkel cell carcinoma is still a challenging problem. Rare occurrence of this tumor and the lack of data concerning its true incidence and long - term responsiveness to therapies make it difficult to determine an 'ideal' management. Merkel cell carcinoma should be treated aggressively with wide excision of the primary lesion (2-3 cm margin) and prophylactic lymphadenectomy followed by irradiation to the primary site (21,22). Chemotherapy is preserved for systemic disease, though the success of this treatment is limited and no chemotherapy protocol has been shown to improve survival (22,23). Radiotherapy alone can be used as palliative treatment with good control of primary and lymph node metastases (24,25). The lymph nodal involvement is correlated with the 5- year survival [survival rates for nodal versus no nodal involvement were 48% and 88% respectively] (26). The local recurrence rate is frequently correlated to the progression of the disease (29). Tumor location on the trunk usually has the worse prognosis compared to those on the head and neck (28). Although systemic involvement indicates a poor prognosis and regression of Merkel cell carcinoma is exceedingly rare, almost 10 cases of spontaneous regression are presented in literature (30,31).

Figure 2: Merkel Cell Carcinoma (H and E * 40)



Case report

A 70-year-old female presented with a history of painful skin nodular lesion of right leg that enlarged rapidly, in recent 6 months. No lymphadenopathy was present. The nodule was pink, non-ulcerated and 4 cm in diameter. Total excision with wide margin was done. Histologically, the tumor composed of uniform round cells with scanty cytoplasm formed in diffuse pattern and focal glandular differentiation. The stroma was fibrotic and infiltrated with lymphoplasmacytic cells. Numerous mitoses, necrosis and apoptosis (Figures 1 and 2) were present. The immunohistochemistry staining for neuron-specific enolase (NSE) and chromogranin were positive but leukocyte common antigen (LCA) was negative.

Treatment consisted of surgical excision of the tumor with a wide margin as well as chemotherapy. The patient expired from extensive distant metastases 8 months after diagnosis.

DISCUSSION

The location of this case of Merkel cell carcinoma is unusual due to the occurrence of the tumor on leg; most Merkel cell tumors have been found on the sun-exposed origin of head and neck. It is difficult to diagnose this type of carcinoma because Merkel cells often resemble cells found in malignant lymphoma or small cell carcinoma. This carcinoma grows rapidly and often metastasizes, therefore early diagnosis and treatment of Merkel cell carcinoma are important factors in decreasing the chance of its spread. The value of an additional antineoplastic chemotherapy in the treatment of Merkel cell carcinoma is still controversial. In our case chemotherapy had no benefical influence on the prognosis.

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