CASE REPORT
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Neuroendocrine (Merkel cell) carcinoma of the auricle

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ABSTRACT

Merkel cell carcinoma is a rare locally aggressive malignancy. Most commonly occurs in the head and neck area and only six cases on the auricle have been reported. Immunocytochemistry may help with the differential diagnosis of small cell tumors that share cytomorphological features with merkel cell carcinoma. We present cytological findings as well as immunocytochemical ones in a case of this unusual entity of the auricle.

KEY WORDS: *Carcinoma, Merkel Cell; Ear Neoplasms; Ear, External; Immunocytochemistry; Cytology*

INTRODUCTION

Merkel cell carcinoma (MCC) is a rare primary neuroendocrine carcinoma of the skin. The Merkel corpuscle is located at the dermal- epidermal junction and functions as a slowly adapting type I mechanoreceptor that mediates sense of touch and hair movement (1). Merkel cell carcinogenesis seems to share pathogenic mechanisms with other neoplasms of neural derivation and most commonly occurs in the head and neck area or buttocks of elderly patients. Vortmeyer and colleagues (2) have found deletions involving chromosome 1p35-36, the same location that has been suspected for genetic changes in pheochromocytoma, neuroblastoma, and melanoma. Patients with MCC present with a rapidly enlarging, painless nodule that is locally aggressive and can metastasize. Sun exposure may be as important etiologic factor because most tumors were observed on sun-exposed areas of fair skinned individuals (3). Approximately 600 cases have been described in the literature (4) since 1972 when Toker reported the first case (5). Only six cases of this tumor on the auricle have been reported previously (6).

CASE REPORT

A 78-year old male presented to the department of otorhynolaryngology with two months history of rapidly enlarging painless nodule on the posterior surface of the auricle of his right ear and one palpable lymph node in the neck (Figure 1).

Imaging studies showed no tumor in the thorax and abdomen. The fine needle biopsy (FNA) obtained a poorly cohesive population of small uniform cells (Figure 2). The cells had rounded nuclei with powdery chromatin and scant cytoplasm. A few rosette-like aggregates were present and frequent nuclear molding was seen. Mitotic figures were also frequent. It was necessary to differentiate this tumor from lymphoma, small cell lung carcinoma, melanoma, and neuroblastoma. Immunocytology revealed that the tumor cells were totally negative for leukocyte common antigen (LCA), S-100 protein, and HMB45, whereas they were strongly positive for cytokeratin 20 and neuron specific enolase (NSE) (Figure 3).

These findings indicated the diagnosis of neuroendocrine (Merkel cell) carcinoma. Local wide excision of the tumor and the node was performed. Histological examination showed

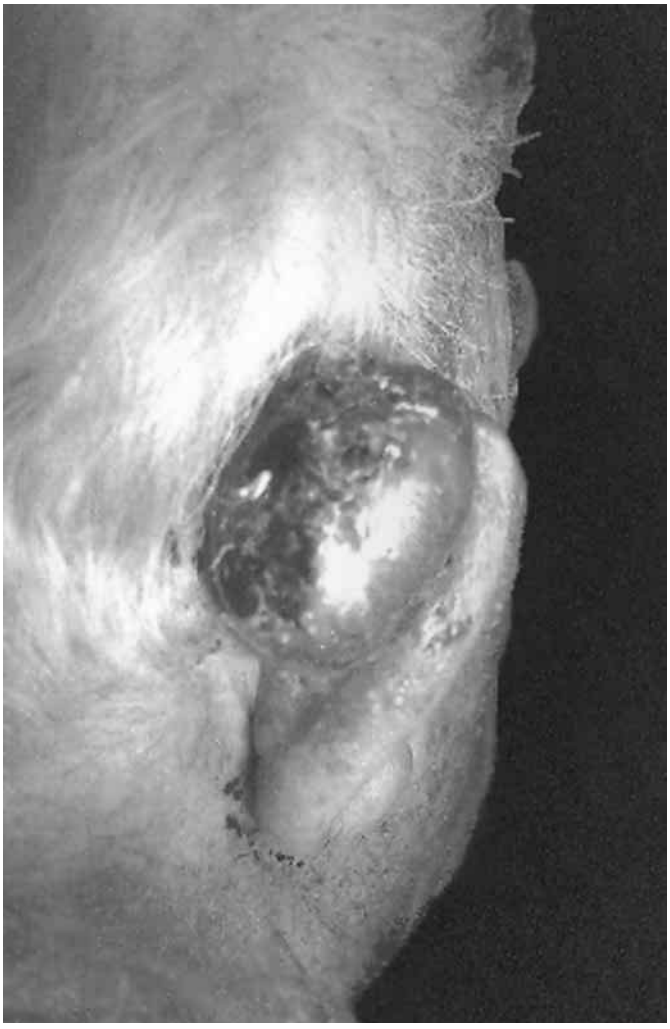

Figure 1. Painless nodule on the auride of the ear

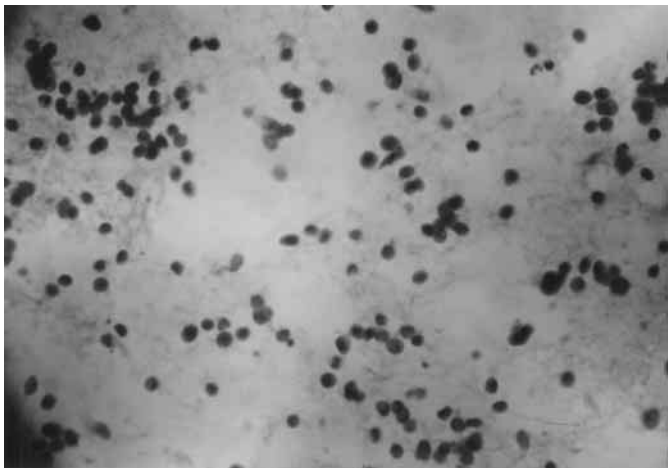

Figure 2. Merkel cell carcinoma, poorly cohesive population of small uniform cells (Pap stain x200)

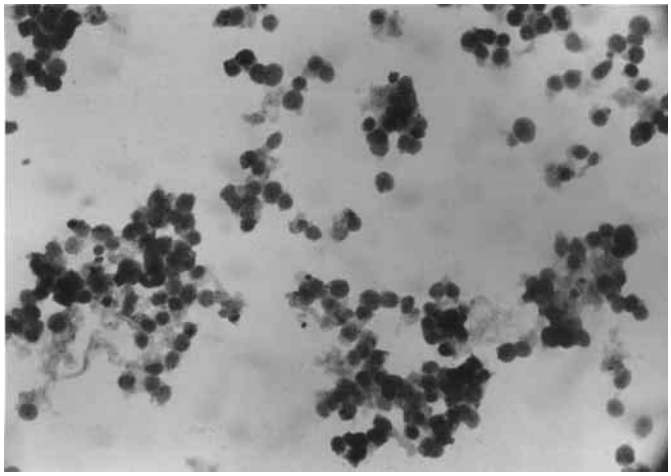

Figure 3. Merkel cell carcinoma. Positive staining of tumor cells with Neuron Specific Enolase x 200

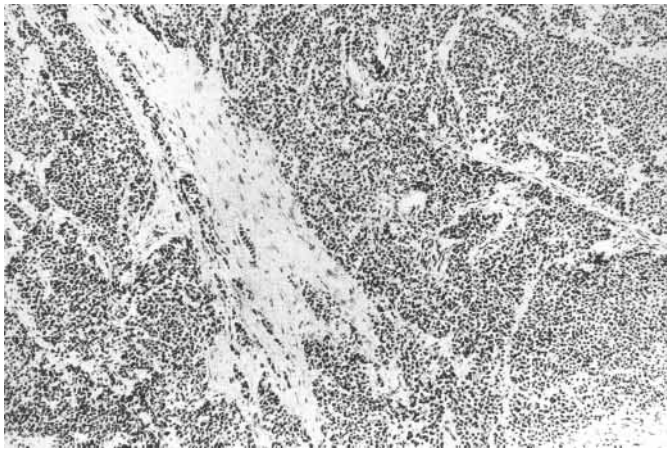

Figure 4. Merkel cell carcinoma. Histologic section of the tumor (Hematoxylin and eosin preparation x100)

Merkel cell carcinoma and tumor cell invasion in one lymph node (Figure 4).

Our patient underwent radiotherapy. Ten months after the operation no pathological lymph nodes were palpable in the neck and no evidence of local or distant recurrence was found.

DISCUSSION

The most common site of occurrence of the tumor is the head and neck (50%) followed by the extremities (35%) and other areas (15%) of elderly patients (7). MCC is a rare and locally aggressive cutaneous malignancy, which is potentially lethal. Distant metastases occur in 20% to 40% of the patients. Cases with MCC of unknown primary site are reported in the literature (8-10). Cytologically, it consists of a poorly cohesive monotonous population of small cell clusters and isolated cells with scanty, weakly basophilic cytoplasm. The nuclei are round or polyhedral with inconspicuous nucleoli, fine powdery chromatin, and well-defined nuclear membrane. Nuclear molding and mitotic figures are frequent.

Many primary and metastatic tumors may resemble Merkel cell carcinoma. The major differential diagnosis is small cell carcinoma of the lung. Distinguishing features of MCC include more uniform cells, absence of spindle-shaped cells, and a more dispersed cell pattern. It is also important to differentiate this tumor from malignant lymphoma, melanoma, adult neuroblastoma, basal cell carcinoma, metastatic carcinoid tumor or primary adnexal carcinoma. Immunocytochemical demonstration of neuron-specific enolase, intense staining for keratins (CK and CK20), and absence of S100 protein, leukocyte common antigen, and HB45 are enough to confirm the diagnosis. Cytokeratin 20 presents dotlike and crescent patterns. Occasional cases of MCC are negative for CK20 while there are some small cell carcinomas of the lung that are positive for the same marker. TTF-1 may help with the differential diagnosis, because no reactivity has been shown in MCC (11). Ultrastructural study revealed the presence of cytoplasmic extensions, complex intercellular junctions, collections of perinuclear intermediate filaments and dense-core, membrane-bound neurosecretory granules (12). Because of the documented aggressiveness of MCC, early and aggressive surgical excision has been advocated. In addition, the use of adjuvant radiotherapy is recently widely accepted (13) for advanced regional nodal disease.

In conclusion, otolaryngologists and cytopathologists should be aware of this rare carcinoma when faced with cutaneous tumors of the head and neck region.

REFERENCES

1. Halata Z. Sensory innervation of the hairy skin (light and electron microscopic study). J Invest Dermatol 1993;101(suppl):755-815.
2. Vortmeyer AO, Merino MJ, Boni R, Liotta LA, Cavazzana A, Zhuang Z. Genetic changes associated with primary Merkel cell carcinoma. Am J Clin Pathol 1998;109:565-70.
3. Marks ME, Kim RY, Salter MM. Radiotherapy as an adjunct in the management of Merkel cell carcinoma. Cancer 1990;65:60-4.
4. Voog E, Biron P, Martin JP, Blay JY. Chemotherapy for patients with locally advanced or metastatic Merkel cell carcinoma. Cancer 1999;85:2589-95.
5. Toker C. Trabecular carcinoma of the skin. Arch Dermatol 1972;105:107-10.
6. Virtaniemi J, Hirvikoski P, Pukilla M, Kumplainen E, Johanson R, Kosma VM. Merkel cell carcinoma of the auricle. Eur Arch Otorhinolaryngol 2000;257:558-60.
7. Trimas S, Stringer S, Pierson K. Merkel cell carcinoma or primary neuroendocrine carcinoma of the skin. Arch Otolaryngol 1992;9:998-1002.
8. Straka JA, Straka MB. A review of Merkel cell carcinoma with emphasis on lymph node disease in the absence of a primary site. Am J Otolaryngol 1997;18:55-65.
9. Kaplan GP, Bookbinder MJ, Hood DR, Bridgers SL. Merkel cell tumor of the skin. Hum Pathol 1988;19:615-6.
10. Pilotti S, Rilke F, Bartoli C, Grisotti A. Clinicopathologic correlations of cutaneous neuroendocrine Merkel cell carcinoma. J Clin Oncol 1988;6:1863-73.
11. Hanly AJ, Elgart GV, Jorda M, Smith J, Nadji M. Analysis of thyroid transcription factor-1 and cytokeratin 20 separates Merkel cell carcinoma from small cell carcinoma of the lung. J Cutan Pathol 2000;27:118-20.
12. Sibley RK, Dehner LP, Rosai J. Primary neuroendocrine (Merkel cell?) carcinoma of the skin. A clinicopathologic and ultrastractural study of 43 cases. Am J Surg Pathol 1985;9:95-108.
13. Brisseti AE, Olsen KD, Kasperbauer JL, Lewis JE, Goellner JR, Spott BE et al. Merkel cell carcinoma of the head and neck: a retrospective case series. Head and Neck 2000;982-8.