JAMER 2021;6(2):54-56

# Parotid Glandın Primer Merkel Hücreli Karsinomu

## Primary Merkel Cell Carcinoma Of The Parotid Gland

Fatma Şenel, Hatice Karaman

Kayseri City Hospital, Department of Pathology

### ÖZ

Merkel hücreli karsinom (MCC) cildin nadir ve agresif seyirli nöroendokrin tümörüdür. Bu tümör tükrük bezinde ise oldukça nadir görülür. MCC da lokal rekürrens, lenf nodu metastazı ve uzak metastaz sıktır. Biz burada parotid glandın primer Merkel hücreli karsinomunu sunduk.
**Anahtar kelimeler:** Merkel hücreli karsinom, parotid gland, tükrük bezi

### ABSTRACT

Merkel cell carcinoma (MCC) is a rare and aggressive neuroendocrine tumor of the skin. This tumor is very rare in the salivary gland. Local recurrence, lymph node metastasis, and distant metastasis are common in MCC. Here, we presented the primary Merkel cell carcinoma of the parotid gland.
**Keywords:** Merkel cell carcinoma, parotid gland, salivary gland

## INTRODUCTION

Merkel cell carcinoma is a rare neuroendocrine cancer of the skin (1). Initially, it was believed to originate only from the merkel corpuscles of the epidermis (2). However, it is considered that MCC may also originate from stem cells differentiated into different cells (3). The most common localization of MCC is the sun-exposed skin area, and it is observed in the head and neck region by 50% (4). MCC was also rarely reported in noncutaneous regions (5). MCC is considered an aggressive neoplasm with metastatic potential (6). Here, we presented the primary Merkel cell carcinoma of the parotid gland.

## CASE

A 71-year-old male patient applied to our center with the complaint of a mass that had emerged in front of the left ear 3 months ago and was growing gradually. In the physical examination of the patient, a solid, 12x10x5 cm mass extending to the mastoid apex posteriorly on the left, to the cricoid cartilage level inferiorly, and to the buccal regi on anteriorly was observed. In the superficial ultrasonography of the patient, a space-occupying formation with a heterogeneous internal structure of 57x35 mm at the level of the left parotid gland was observed. The lesion had a component of approximately 5 cm extending posteriorly to the posterior cervical region. Conglomerate lymph nodes, the largest of which was 32x15 mm in diameter, were partly observed inferiorly to it. Lymph nodes with a diameter of 22x13 mm, the largest of which was in zone 4, were observed in the left cervical chain. A 42x30 mm mass lesion with irregular borders and lobulated contours extending from the superficial lobe to the deep lobe and containing a cystic necrotic component in the left parotid gland was detected in the magnetic resonance examination. Many lymph nodes were observed in the left cervical chain and in the left posterior cervical region.

**Sorumlu Yazar/Corresponding Author:** Fatma Şenel
Kayseri Şehir hastanesi Patoloji Kliniği Kayseri.

**e.mail:** @gmail.com

**Tel:** 05370249110

**Geliş tarihi/Received:** 03.07.2021
**Kabul tarihi/Accepted:** 28.07.2021

## INTRODUCTION

Merkel cell carcinoma is a rare neuroendocrine cancer of the skin (1). Initially, it was believed to originate only from the merkel corpuscles of the epidermis (2). However, it is considered that MCC may also originate from stem cells differentiated into different cells (3). The most common localization of MCC is the sun-exposed skin area, and it is observed in the head and neck region by 50% (4). MCC was also rarely reported in noncutaneous regions (5). MCC is considered an aggressive neoplasm with metastatic potential (6). Here, we presented the primary Merkel cell carcinoma of the parotid gland.

## CASE

A 71-year-old male patient applied to our center with the complaint of a mass that had emerged in front of the left ear 3 months ago and was growing gradually. In the physical examination of the patient, a solid, 12x10x5 cm mass extending to the mastoid apex posteriorly on the left, to

the cricoid cartilage level inferiorly, and to the buccal regi on anteriorly was observed. In the superficial ultrasonography of the patient, a space-occupying formation with a heterogeneous internal structure of 57x35 mm at the level of the left parotid gland was observed. The lesion had a component of approximately 5 cm extending posteriorly to the posterior cervical region. Conglomerate lymph nodes, the largest of which was 32x15 mm in diameter, were partly observed inferiorly to it. Lymph nodes with a diameter of 22x13 mm, the largest of which was in zone 4, were observed in the left cervical chain. A 42x30 mm mass lesion with irregular borders and lobulated contours extending from the superficial lobe to the deep lobe and containing a cystic necrotic component in the left parotid gland was detected in the magnetic resonance examination. Many lymph nodes were observed in the left cervical chain and in the left posterior cervical region.

A tru-cut biopsy was performed from the patient's parotid gland. In the histopathological examination of the tissue, a monotonous-appearing tumor with a small round hyperchromatic nucleus and indistinct cytoplasm was observed. Acini structures of the salivary gland were observed in an area. Necrosis was observed in a small area. In the immunohistochemical examination (IHC), it was stained CD56, synaptophysin-positive, chromogranin, CD45, TTF1, CK7 negative. The Ki67 proliferation index was around 95%. The case was diagnosed as a "Neuroendocrine malignant tumor" with these results. Then, the patient underwent left total parotidectomy and left extended radical neck dissection. The cross-sectional face of the 12x10x6 cm excision material was solid-appearing, and the middle was necrotic. Cells with a rounded hyperchromatic nucleus were observed in the histopathological image of the tumor, and these cells formed a solid development pattern, sometimes trabecular arrays and rosette structures (Fig.1). Necrosis and frequent mitosis were observed. The tumor was infiltrated into the salivary gland, muscle, and adipose tissue. In the IHC examination, it was CD56, NSE positive (Fig.2), positive for PanCK, CK20 in a dot-like pattern (Fig.3), EMA positive, diffuse positive for P53, CD45, actin desmin, vimentin, S100 negative. The Ki67 proliferation index was around 95%. The case was diagnosed as "Merkel cell carcinoma" with these results.

Many metastatic lymph nodes were observed in the examination of the patient's lymph nodes. No primary skin lesion was observed in the dermatological evaluation. Chemoradiotherapy was then applied to the patient as a treatment.

## DISCUSSION

Merkel cell carcinoma constitutes 1% of all parotid gland carcinomas and 3.5% of all malignant minor salivary gland tumors (7). Merkel cell carcinoma occurs over the age of 50. Our case is 71 years old. It occurs in immunosuppressed patients, especially in organ transplant recipients, and sun exposure. Fong et al. detected the Merkel cell polyomavirus, a new polyomavirus in most MCCs (8). The relation of our case with polyoma virus could not be determined.

Of these tumors, 50% are present in the head and neck region (4). This tumor is a little more common in men (9). In the primary MCC of the salivary gland, neural crest cells are considered to differentiate into mature Merkel cells (10).

Atypical sites of primary MCC in the head and neck region were reported. These are the tongue, lips, alveolar mucosa, hard palate, nasal fossa, the floor of the mouth, and parotid gland (9).

Fornelli et al.(11) hypothesized that the MCC of the parotid gland develops from the neuroendocrine component of Warthin's tumor in the absence of a cutaneous MCC. Neuroendocrine salivary gland carcinomas are classified according to their immune phenotype. In CK20 positivity, it is called the merkel cell type. In the other type, the pulmonary type, CK20 is not expressed (12). In our case, it was also CK20 positive.Major tumors that can be confused with Merkel cell carcinoma are high-grade lympho-

mas and metastatic undifferentiated small cell carcinoma. Differential diagnosis can be made with an immunohistochemical panel.

Five-year overall survival is between 40-68% in head and neck MCCs, and the mortality rate is twice that of cutaneous melanomas (33% vs. 15%) (13). While the 5-year survival rate in cutaneous MCC is 74%, this rate is 47% in primary MCC of the parotid gland (14).

Metastasis is most common to the lymph nodes, mediastinum, lung, liver, and bone (15). In our case, there was only cervical lymph node metastasis. Merkel cell carcinoma has a poor prognosis and probably requires multimodal treatment. Most of the authors recommend surgery and chemoradiotherapy (16,17). Chemoradiotherapy was administered to our case due to extraparenchymal extension and lymph node metastasis.

## CONCLUSION

MCC of the salivary gland is very rare and has an aggressive course. This tumor should be considered in the differential diagnosis of salivary gland tumors. Histopathological appearance and IHC examination are very important for diagnosis. Multimodal treatment is recommended for this tumor.

**Informed Consent:** Written informed consent was obtained from patient who participated in this study.
**Conflict of Interest:** No conflict of interest was declared by the authors.
**Financial Disclosure:** The authors declared that this study has received no financial support.
**Author Contributions:** Conception/Design of Study- F.Ş.; Data Acquisition- F.Ş.; Drafting Manuscript- F.Ş., H.K.; Critical Revision of Manuscript- H.K.; Final Approval and Accountability- F:Ş.; Supervision- H:K.

## References

**1**. Toker C. Trabecular carcinoma of the skin. Arch Dermatol. 1972;105:107–10.
**2**. Tang CK, Toker C. Trabecular carcinoma of the skin: an ultrastructural study. Cancer. 1978;42:2311–21.
**3**. Saeb-Lima M, Montante-Montes de Oca D, Albores-Saavedra J. Merkel cell carcinoma with eccrine differentiation: a clinicopathologic study of 7 cases. Ann Diagn Pathol. 2008;12:410–4.
**4**. Ghaderi M, Coury J, Oxenberg J, Spector H. Primary Merkel cell carcinoma of the parotid gland. Ear Nose Throat J. 2010;89(7):24-7.
**5**. Knopf A, Bas, M, Hofauer B, Mansour N, Stark T. Clinicopathological characteristics of head and neck Merkel cell carcinomas. Head Neck.2017 ;39(1):92-97.
**6**. Agelli M, Clegg LX, Becker JC, et al. The etiology and epidemiology of Merkel cell carcinoma. Curr Probl Cancer. 2010;34:14–37.
**7**. Gnepp DR, Corio RL, Brannon RB. Small cell carcinoma of the major salivary glands. Cancer. 1986;58:705–14.
**8**. Feng H, Shuda M, Chang Y, Patrick S Moore. Clonal integration of a polyomavirus in human Merkel cell carcinoma. Science. 2008;319:1096–100.
**9**. Gioacchini FM, Postacchini V, Simonetti O, Offidani A, Magliulo G. Massimo Re: Merkel cell carcinoma : a systemic review of ENT presentation. Eur Arch Oto-Rhino-Laryngol. 2013;270:2191–9.
**10**. Szeder V, Grim M, Halata Z, Sieber-Blum M. Neural crest origin of mammalian Merkel cells. Dev Biol. 2003;253(2):258–63.
**11**. Fornelli A, Eusebi V, Pasquinelli G, Quattrone P, Rosai J. Merkel cell carcinoma of the parotid gland associated with Warthin tumour: report of two cases. Histopathology. 2001;39(4):342–6.
**12**. Chernock RD, Duncavage EJ, Gnepp DR. Absence of Merkel cell polyomavirus in primary parotid high-grade neuroendocrine carcinomas regardless of cytokeratin 20 immunophenotype. Am J Surg Pathol. 2011;35:1806–11.
**13**. American Joint Committee on Cancer. AJCC cancer staging manual. eighth ed. Springer; 2017. Chapter 46.
**14**. Agelli M, Clegg LX. Epidemiology of primary Merkel cell carcinoma in the United States. J Am Acad Dermatol. 2004:49 (5):832-41.
**15**. Basati M, Kassam K, Messiha A. Secondary Merkel cell carcinoma manifested in the parotid: case reports in dermatological medicine. 2013. p. 4.
**16**. Hui KK, Luna MA, Batsakis JG, Ordóñez NG, Weber R. Undifferentiated carcinomas of the major salivary glands. Oral Surg Oral Med Oral Pathol. 1990;69:76–83.