

Merkel Cell Carcinoma Clinically Mimicking a Traumatic Suture Scar in the Nasal Vestibule: Earlier Diagnosis for Better Prognosis

PETER JIANG¹ and JINPING LAI²

¹University of Florida College of Medicine, Gainesville, FL, U.S.A.;

²Department of Pathology and Laboratory Medicine,
Kaiser Permanente Sacramento Medical Center, Sacramento, CA, U.S.A.

Abstract. *Background:* Merkel cell carcinoma (MCC) is an aggressive form of skin cancer that is often found in sun-exposed regions of the skin in older or immunocompromised individuals. To date there have only been four cases reported of MCC found in the nasal vestibule. *Case Report:* We document an unusual case of MCC in the nasal vestibule of a 66-year-old female which was clinically believed to be an inflammatory reaction to a previous suture scar. The patient also had a history of dermatitis treated with UV light. The lesion was less than 5 mm in the greatest dimension and biopsied. There was small blue cell infiltrate in the dermis and the tumor cells were positive for synaptophysin, chromogranin and cytokeratin 20. They were negative for S100 proteins, melan A, human melanoma black 45 (HMB45) and lymphocyte common antigen CD45. For Ki-67, staining was positive in 90% of the tumor cells and tumor protein 53 immunoreactivity showed a wild type staining pattern. The pathological diagnosis was MCC and the patient underwent Mohs surgery with no tumor recurrence or metastasis identified at 2.5 years of follow-up. *Conclusion:* To the best of our knowledge, cases of MCC arising in the nasal vestibule with clinical features mimicking a suture scar have not been reported. The importance of early diagnosis and treatment of MCC to improve patient prognosis are discussed.

Merkel cell carcinoma (MCC) is a rare but aggressive form of skin neuroendocrine cancer and the second most common

Correspondence to: Jinping Lai, MD, Ph.D., Department of Pathology and Laboratory Medicine, Kaiser Permanente Sacramento Medical Center, Sacramento, CA 95825, U.S.A. Tel: +1 9169737260, Fax: +1 9169737283, e-mail: jinping.x.lai@kp.org

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cause of skin cancer-related death (1). MCC is often found on sun-exposed regions of the skin of the head and neck in older or immunocompromised individuals. MCC exhibits nonspecific physical characteristics and often presents as a red or pink lesion that is <2 cm. MCC also tends to present with intravascular invasion, a high degree of local recurrence, and distant metastasis that results in high mortality despite surgical removal and chemotherapy.

MCC involving the nasal mucosa is exceedingly rare. To date, as far as we are aware from a PubMed search, there have only been three other case reports of MCC in the nasal vestibule (2-4). We report an unusual case of MCC, which was diagnosed at less than 5 mm, clinically mimicking a suture scar status post trauma at the nasal vestibule. To the best of our knowledge, this is the first documented case of MCC clinically mimicking a suture scar. Early diagnosis and therapy is very important for a better prognosis and the etiology, diagnosis and treatment of this entity are also discussed.

Case Report

We report the case of a 66-year-old female with a past medical history of smoking, seborrheic keratoses, and dermatitis treated with UV. Her only relevant family history was basal cell carcinoma in her brother. She had no personal history of melanoma or other skin cancer. The lesion was first discovered by her dermatologist, who documented it as a 1 mm folliculocentric papule which he believed to be folliculitis and for which he prescribed topical Bactroban. Four months later, at a visit to a different physician, the lesion had grown to 5 mm. The lesion was thought to be an inflammatory reaction to a previous suture which had been placed in her nasal vestibule a few months earlier after a motor vehicle accident. The physician biopsied the lesion, fully anticipating the lesion to show a simple foreign body reaction.

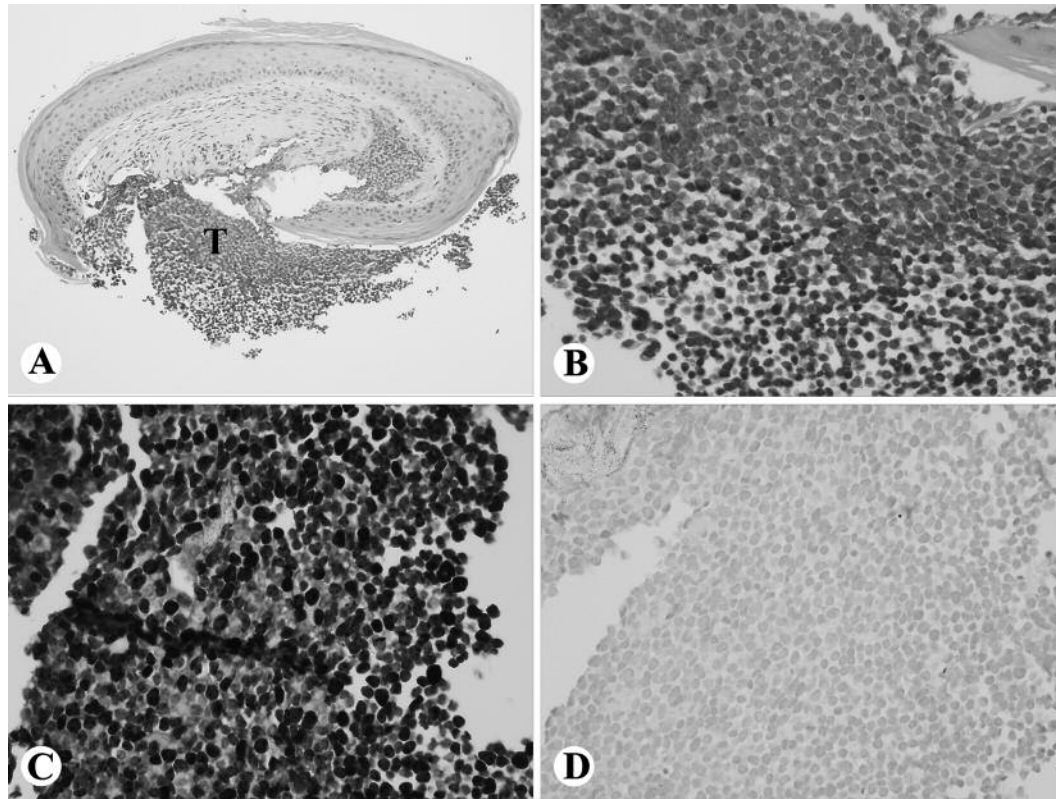


Figure 1. Low-power (A; $\times 100$) and high-power (B; $\times 400$) views of the histology of the specimen from biopsy of the nasal vestibule lesion, showing dermal small round blue cell infiltrate (T) (hematoxylin and eosin stain). Immunohistochemistry showed a high Ki-67 proliferative index (90%) (C; $\times 400$) and negativity for melan-A (D; $\times 400$).

To the physician's surprise, the biopsy results showed no foreign bodies nor any foreign body giant cell reaction. Instead it showed clusters of small blue cells with round nuclei and scant cytoplasm with plasmacytoid features showing invasion of the dermis (Figure 1A and B). Immunostaining showed high Ki-67 proliferative index (90%) (Figure 1C) and negativity for melan A (Figure 1D), S100, HMB45 and CD45. The tumor cells had a positive perinuclear staining pattern for cytokeratin 20 (Figure 2A) and positive for neuroendocrine markers synaptophysin and chromogranin (Figure 2 B and C). The pathological diagnosis was MCC. Due to the patient's history of UV exposure, tumor protein 53 (TP53) immunostain was also performed and showed a wild-type staining pattern (Figure 2D).

After the initial diagnosis, the patient had a thorough work-up including a baseline positron emission tomography-computed tomography (PET/CT) scan showing no evidence of metastasis. The patient elected to undergo Mohs surgery with reconstructive surgery of the *nare* performed with a postauricular full thickness skin graft. She declined lymph node dissection and adjuvant radiation due to her personal concerns about the side-effects of radiation. 2.5 years of

follow-up, the patient was doing well and there was no evidence of recurrence. PET/CT and neck ultrasound imaging examination showed no signs of metastasis.

Discussion

MCC in the nasal vestibule is exceedingly rare. There are only four cases reported in the English literature (2-4), with variable prognoses. It seems MCC of the nasal vestibule has a relatively better prognosis as compared to that seen in the other head and neck origins, but more case reports are needed to further characterize the disease.

This case is particularly interesting due to the unique nature of its presentation. Due to the previous history of minor nasal trauma and suture placement, the lesion had been initially overlooked as an inflammatory reaction. MCC had not been a consideration at all in this scenario. The histological features and immunoprofile including cytokeratin 20, synaptophysin and chromogranin expression are important in order to make the correct diagnosis.

Although MCC has a poor prognosis with high rate of metastasis, our case was incidentally diagnosed when the

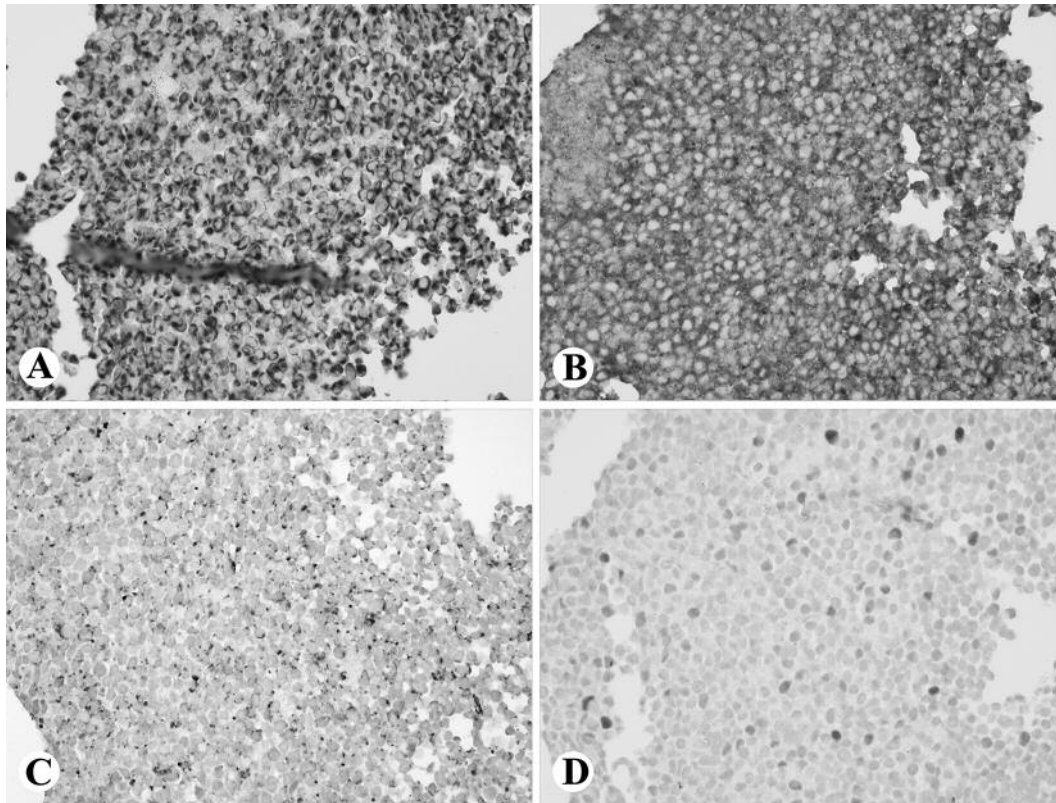


Figure 2. Immunohistochemistry for Merkel cell carcinoma. The tumor cells were positive for cytokeratin 20 (A), synaptophysin (B) and chromogranin (C), with a wild-type staining pattern for tumor protein 53 (D) ($\times 400$).

tumor was less than 5 mm, at the unusual location of the nasal vestibule. Subsequently, the patient underwent Mohs surgery and had a good prognosis. This case highlights the importance for clinicians to be aware that MCC can present in unusual ways. Due to the aggressive nature of MCC, it is often identified at much later stages of the disease and thus results in poor prognosis. However, we believe that if these lesions are identified and treated early, they can result in much more favorable outcomes.

Merkel cell polyomavirus has been highly associated with anywhere from 55% to 90% of reported cases of MCC (5). Those MCCs that are Merkel cell polyomavirus-negative often express DNA mutations secondary to UV exposure. From a molecular perspective, the main mechanisms of transformation promoting MCCs are still partially unknown. Mutations in *TP53* have been observed in 14-33% of patients with MCC and might be associated with UV exposure (6). Patients with these types of somatic mutations in MCC may have worse prognosis. A potentially important risk factor in this patient's history was UV radiation as treatment for her dermatitis. However, no *TP53* mutation immunostaining pattern was identified in this case.

In conclusion, we report a rare case of MCC arising in nasal vestibule with clinical features mimicking suture post trauma. We hope that this case of MCC of the nasal vestibule can help serve as an example to encourage clinicians to be aware of the unusual presentation of these tumors. Early diagnosis is vital to improved prognosis and outcomes of these patients.

Conflicts of Interest

The Authors declare that they have no conflicts of interest in regard to this case

Authors' Contributions

PJ wrote the article; JL made the diagnosis, collected and analyzed the data and finalized the article. Both Authors approved the final article.

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