Merkel Cell Carcinoma Of The Nasal Vestibule
Babak Sadoughi, MD, Elizabeth Guardiani, MD, Andrew Blitzer, MD, DDS
Department of Otolaryngology, St. Luke’s-Roosevelt Hospital Center, New York, NY

Abstract
Merkel cell carcinoma is a neuroendocrine malignancy of aggressive nature despite its occasionally indolent presentation. A patient presenting with a slowly growing nasal vestibular lesion, initially managed as folliculitis, underwent further outpatient workup studies leading to the diagnosis of Merkel cell carcinoma. No regional or distant metastasis was detected at the time of diagnosis, however the patient developed two submental lymph node metastases within 2 weeks, while obtaining multidisciplinary consultation. A wide local excision with cervical lymphadenectomy was performed followed by chemoradiation. Despite durable local control, the patient developed distant spine and perineal metastases 10 months after treatment completion.

Introduction
• Merkel cell carcinoma (MCC) is a rare skin malignancy, also described as primary neuroendocrine carcinoma, primary small cell carcinoma of the skin or cutaneous APUDoma, with a rapidly increasing incidence over the past few decades.
• Lesions typically arise in the sun-exposed areas of older individuals, with a predominance of head and neck locations.
• The hallmark of MCC is its aggressive nature, with a tendency to rapidly develop regional and distant metastases and a poor overall prognosis.
• Treatment is subject to controversy and commonly underestimated given the often indolent presentation.

Case Report
• A 75 year-old woman presented complaining of a painful “pimple” in the right nostril for several days.
• Physical examination: 1.5-mm papular lesion in the hair-bearing portion of the right alar vestibule, with local purulent discharge upon gentle palpation.
• The lesion was managed by incision, drainage and tissue biopsy under local anesthesia. Microbiology and pathology studies were suggestive of folliculitis, treated with directed antibiotics and local wound care.

Figure 1: Anterior rhinoscopy view of right nasal vestibular lesion. Fig. 2: Reconstruction with composite auricular (helical rim) graft.

Discussion
• This clinical vignette underscores the challenges associated with the management of MCC and its unusually hostile course; a tendency to develop regional metastases lesions during the early stages of the primary disease compromises the chances of curative treatment.
• The atypical biology of MCC may be linked to its peculiar histology and pathogenesis: UV radiation exposure and immune suppression have been proposed as significant risk factors, but an infectious etiology has also become plausible with the recent discovery of the Merkel cell polyomavirus (MCPyV) and its strong clinical association with MCC. MCPyV antibody titers might also be promising as forthcoming prognostic factors.
• Various MCC staging systems have been proposed, lacking uniformity for collaborative research purposes. A recent international consensus panel described a definitive prognostic and staging system for MCC. Dedicated International Classification of Diseases diagnostic codes were also created. These measures should encourage coordinated efforts to better understand MCC and suggest reliable prognostic parameters, which will subsequently help elaborate standardized treatment protocols.

Conclusion
This report underlines the highly aggressive nature of MCC, which should not be underestimated even in the context of a seemingly benign presentation. Recent developments have generated new paradigms awaiting translation into clinical practice, and helped design more effective epidemiologic tools. Further research is needed to identify prognostic parameters and design validated treatment algorithms in the management of MCC.

References