A 91-year-old white man with a history of atrial fibrillation, benign prostatic hyperplasia, dysphagia, gastroesophageal reflux disease, hypertension, hypothyroidism, osteoarthritis, and laryngeal cancer presented with an 8-mm firm, painless, pink lesion with telangiectasia on the left wrist. The lesion had been present for an unknown period of time and was asymptomatic at presentation.

WHAT’S THE DIAGNOSIS?

a. acneform lesion  
b. basal cell carcinoma  
c. cyst  
d. dermatofibrosarcoma protuberans  
e. Merkel cell carcinoma
A partial biopsy was performed during the dermatology examination. Histopathology demonstrated a dense dermal infiltrate of small, dark blue, pleomorphic cells (Figure 1). On high power, the individual cells were noted to have vesicular nuclei with finely granular and dusty chromatin (Figure 2). Numerous mitotic figures were present. Immunohistochemical stains were performed and revealed positive staining for cytokeratin 20 (with a perinuclear dot pattern), synaptophysin, and chromogranin.

Merkel cell carcinoma (MCC) is an uncommon carcinoma of the epidermal neuroendocrine cells with approximately 1500 cases a year in the United States.1 Merkel cell carcinoma has a poor prognosis with approximately one-third of cases resulting in death within 5 years and with a survival rate strongly dependent on the stage of disease at presentation.2 A complete surgical excision with histologically verified clear margins is the main form of treatment of the primary cancer.3 Although the effectiveness of adjuvant therapy for MCC has been debated,4 retrospective analysis has shown that the high local recurrence rate of the primary tumor can be reduced by combining surgical excision with a form of radiation therapy.5

A systematic cohort study of 195 patients diagnosed with MCC summarized its most clinical factors with the acronym AEIOU: asymptomatic, expanding rapidly, immunosuppression, older than 50 years of age, and UV-exposed site on a fair-skinned individual.6 The role of immune function in MCC was highlighted by a 16-fold overrepresentation of immunosuppressed patients in the studied cohort as compared to the general US population. The immunosuppressed patients included individuals with human immunodeficiency virus, chronic lymphocytic leukemia, and iatrogenic suppression secondary to solid organ transplantation.6

In 2008, Merkel cell polyomavirus (MCPyV) was found in 80% (8/10) of MCC tumors tested.7 Since then, many different studies have suggested that MCPyV is an etiologic agent of MCC.8-10 A natural component of skin flora, MCPyV only becomes tumorigenic after integration into the host DNA and with mutations to the viral genome.11 Although there currently is no difference in treatment of MCPyV-positive and MCPyV-negative MCC,12 research is being done to determine how the discovery of the MCPyV could impact the treatment of MCC.

REFERENCES

FIGURE 1. A biopsy of the lesion showed a small blue cell tumor located primarily in the dermis (H&E, original magnification ×4).

FIGURE 2. On high power, the cells were noted to have vesicular nuclei containing small nucleoli as well as granular and dusty chromatin (H&E, original magnification ×20).


