Merkel cell carcinoma (MCC) is an uncommon aggressive neuroendocrine skin carcinoma. It usually affects sun-exposed skin of white elderly people. MCC is characterized by a high incidence of early locoregional relapse and distant metastases. Because of its rarity and the resulting lack of prospective randomized trials, data regarding the optimal treatment of MCC are limited. Despite aggressive multimodality treatment, the prognosis of patients bearing MCC is often poor. We report three cases of lymph node metastases of MCC with unknown primary sites. Two patients died 17 and 28 months after diagnosis due to brain and pancreatic metastases, respectively, without evidence of cutaneous disease. The third patient is alive and free of tumor at 16 months from the diagnosis. After an accurate diagnosis of lymph node metastases from MCC, the absence of a primary tumor at complete initial evaluation and during adequate follow-up can confirm this particular clinical scenario. The prognosis seems to be analogous to that of cases with similar disease stage (lymph node involvement) but a known primary site.