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A Review of Merkel Cell Carcinoma for Dermatology Nurses

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ABSTRACT: Merkel cell carcinoma (MCC) is an uncommon but frequently deadly form of skin cancer. It can present in a variety of ways, which appear very similar to many other benign and malignant skin neoplasms. New diagnostic and treatment methods are being implemented to try to increase survival and improve quality of life. Still, because MCC is so rare and because there is no one standard of care, management of this cutaneous malignancy can be a challenge. This article provides an overview of MCC and its clinical features, diagnosis, treatment, and management.

Key words: Anaplastic Carcinoma of the Skin, Chemotherapy, Merkel Cell Carcinoma, Neuroendocrine Tumor, Non-melanoma Skin Cancer, Primary Small Cell Skin Cancer, Trabecular Cell Carcinoma, Radiation Therapy

Merkel cell carcinoma (MCC) is a rare, aggressive, potentially lethal malignant solid tumor. MCC is known by many terms, including *trabecular cell carcinoma*, *neuroendocrine or primary small cell carcinoma of the skin*, and *anaplastic cancer of the skin*. Classic clinical presentation of MCC is identical to that of numerous other benign and malignant neoplasms (Heath et al., 2008). Malignant cells are found on or just beneath the skin and in hair follicles. Establishing a high index of suspicion is difficult because MCC is uncommon and lacks unique clinical features. Its aggressive nature makes prompt diagnosis and adequate treatment essential (Table 1).

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EPIDEMIOLOGY

Merkel cell carcinoma accounts for far less than 1% of all cutaneous malignancies in the United States (Rigel et al., 2005). Each year in the United States, approximately 1,200 new cases of MCC are diagnosed, compared with 60,000 new melanoma cases, and >1 million new nonmelanoma skin cancers (Heathet et al., 2008; Rigel et al., 2005). Incidence of MCC has tripled since 1986 (Petrou, 2006).

The average age at diagnosis is 69 years; 5% of cases are diagnosed in persons younger than 50 years (Nghiem & Jaimes, 2008; Rigel et al., 2005). Most MCCs occur in Caucasians; cases have been reported in Japanese people; very few Blacks have been diagnosed with MCC. Extensive sun exposure is a risk factor; older White men (65 years or older) are at higher risk. MCC is more common in immunosuppressed patients. Prognosis is poor; overall, 2- and 5-year survival rates are 50%–75% and 30%–64%, respectively (Rigel et al., 2005).

Etiology, Disease Course, and Prognostic Features

The histogenesis of MCC is controversial. Merkel cells (MCs) appear during the eighth week of gestation (Shea & Prieto, 2007). It is thought that they might be derived from embryonic epidermal stem cells. They occur most densely on the lips, hard palate, palms, finger pads, proximal nail folds, and dorsal feet (Shea & Prieto, 2007). Their function is not fully understood. It is thought that MCs stimulate nerve bundles in the skin, induce keratinocyte proliferation, and release bioactive chemicals into the dermis (Shea & Prieto, 2007).

It was initially thought that MCC arises from epidermal MCs. However, most tumors arise intradermally, rarely involving the epidermis only. There are reports of MCC consisting of intraepidermal spread with dermal involvement; and recently, intraepidermal MCC without dermal

TABLE 1. Key Aspects of Merkel Cell Carcinoma

Incidence has tripled since 1986 but still considered rare.
It appears as a solitary, firm, painless, smooth, shiny, erythematous, nonulcerated lesion.
Mortality rates exceed those of malignant melanoma.
Risk factors for development of MCC include male gender, age >65 years, Caucasian, and high ultraviolet radiation exposure.
Metastasis to regional lymph nodes and to other organs is common.
Combination therapy is usually recommended, consisting of wide local excision with radiation therapy and sometimes chemotherapy.
Favorable prognostic indicators include Stage I disease, tumor on the extremity, being female, age <65 years, and absence of comorbidities.
Median overall survival is 31 months.

involvement has been described (Krasagakis & Tosca, 2003). There are several possible cells of origin, including epidermal MCs, dermal MCs, a neural-crest-derived cell, and a residual epidermal stem cell. MCs have been found free in the dermis and in association with terminal axons, where they probably function as slowly adapting mechanoreceptors.

No predisposing conditions have been consistently identified among patients with MCC. The etiologic role of ultraviolet light or ultraviolet radiation has been proposed. There is a higher incidence of MCC among Whites, and MCC has a predilection for sun-exposed areas (head, neck, and extremities). MCC has been described in patients treated with psoralen, a light-sensitizing medication, combined with ultraviolet A phototherapy and ultraviolet B phototherapy (Rigel et al., 2005). However, MCC can present in non-sun-exposed areas (penis and vulva), indicating that there are other etiological factors involved.

Other risk factors include immunosuppression, erythema ab igne, irradiation, congenital ectodermal dysplasia, and Cowden's disease (Rigel et al., 2005). There are numerous reports of spontaneous remission, presumably immune mediated, illustrating the importance of the immune system in the development, prognosis, and treatment of MCC.

Feng, Shuda, Chang, and Moore (2008) noted that MCC occurs most frequently in elderly and immunosuppressed patients. This suggested the possibility of an infectious etiology. Recently, the identification of gene sequences of a polyomavirus, termed *Merkel cell polyomavirus* (MCV), has been detected in MCC tumors (Feng et al., 2008; Sullivan, 2008). This suggests that MCV may be a contributing factor in the development of MCC (Feng et al., 2008; Sullivan, 2008).

Merkel cell carcinoma tumors can be solitary or multiple and have high rates of both local recurrence (25%) and regional lymph node metastases (25%–50%; Wolff, Johnson, & Suurmond, 2005). These tumors frequently disseminate to the viscera and central nervous system; distant metastasis occurs in 33% of cases. MCC mortality rates exceed those of melanoma; mortality rates of MCC and melanoma are 33% and 15%, respectively (Heath et al., 2008; Nghiem & Jaimes, 2008). Thirty-five percent of those diagnosed with MCC die of it (Gagnon, 2004).

In recent years, 40% of patients with MCC experience local recurrence; 55%, regional metastases; and 36%, distant metastases (Krasagakis & Tosca, 2003). Current rates of local recurrence have been reduced to nearly 25% possibly due to successful treatment with wide local excision of the primary tumor (Krasagakis & Tosca, 2003). Unfortunately, rates of regional and distant metastasis have not followed this trend and have remained stagnant.

Clinical Features

The most common symptom of any skin cancer is a change in the skin, especially a change in an existing mole or the growth of a new lesion; this is true of MCC. MCC typically presents as a solitary, firm, painless, smooth, shiny, telangiectatic, nonulcerated, skin colored to erythematous, bluish red to violaceous purple, fixed, intracutaneous nodule that may resemble a cyst. When MCC develops in the deeper dermis and involves the subcutaneous tissue, it can look like a skin-colored plaque. These deeper tumors appear uncharacteristic of a malignant skin cancer and may more closely resemble metastatic tumors or lipomas (Krasagakis & Tosca, 2003). MCC is usually found on sun-damaged skin of the head, neck, or extremities of White people older than 50 years (Nghiem & Jaimes, 2008).

Merkel cell carcinoma can resemble basal cell carcinoma. Generally, overlying skin is intact, but there may be superficial ulceration in larger lesions. The median size of a primary MCC is 2 cm, but tumor size ranges from below 2 cm to 15 cm (Gagnon, 2004). Tumor size varies by location; MCCs occurring on the face tend to be smaller than those found on other parts of the body (Krasagakis & Tosca, 2003). Satellite lesions may occur, but multifocal or disseminated lesions are rare.

Merkel cell carcinoma begins as a slow-growing tumor, which undergoes a period of rapid growth, prompting the patient to seek medical attention. Although MCC has a predilection for the periorbital area, it has been reported on non-sun-exposed areas (trunk, nasal, and oral mucosa).

At initial diagnosis, most patients have localized disease; 76%–89% of patients have one primary skin lesion, 10%–18% have nodal disease, and 1%–2% have distant disease. Fifty to 70% of patients develop regional lymph node metastases; 30%–50% develop distant metastases (Rigel et al., 2005). The most common sites

TABLE 2. Frequency of the Location of Primary Merkel Cell Carcinoma (Gagnon, 2004)

Location	Frequency (%)
Head and neck	50
Extremities	40
Trunk	10

of distant metastases are distant lymph nodes, liver, bone, brain, lung, and skin; the site of distant metastasis does not correlate with primary tumor location. Approximately 2% of patients who present with nodal or distant disease have no apparent primary skin lesion (Gagnon, 2004; Rigel et al., 2005; Table 2).

PATIENT EVALUATION, DIAGNOSIS, AND DIFFERENTIAL DIAGNOSIS

The nonspecific characteristics of MCC create a lengthy differential diagnosis, including squamous cell carcinoma, keratoacanthoma, amelanotic melanoma, epidermal cyst, pyogenic granuloma, adnexal tumor, and lymphoma (Gagnon, 2004). Diagnosis is rarely made before histopathologic evaluation. Histological diagnosis can be difficult because MCC resembles many other widely recognized small blue-cell tumors. The most challenging differentiation is between primary MCC and metastatic small cell carcinoma of the lung (SCLC).

If histology confirms a diagnosis of MCC, chest x-ray must be performed to rule out SCLC; additional studies are performed as clinically indicated, based on patients' symptoms.

Natural development of MCC proceeds in a stepwise fashion, beginning with local disease, then with regional metastasis to the lymph nodes, and finally with distant metastases. MCC staging is important for determining prognosis and treatment options.

MERKEL CELL CARCINOMA STAGING

The most consistently reported adverse prognostic feature is tumor stage followed by tumor size. Several staging systems exist, including those created by the Memorial Sloan-Kettering Cancer Center (Table 3), the Seattle Cancer Care Alliance, and Yiengpruksawan et al. (1991).

TABLE 3. Merkel Cell Carcinoma Staging With Description (Krasagakis & Tosca, 2003)

Stage	Description
Stage I	Primary tumor only
Stage II	Regional or regional nodal metastases
Stage III	Distant metastases, most commonly to lymph nodes, skin, liver lung, bones, and brain

Staging workup should include palpation of lymph nodes, liver, and spleen; liver function blood tests; chest radiograph; and magnetic resonance imaging or computed tomography (CT) of the chest, abdomen, and pelvis to assess for dissemination to lymph nodes and viscera. Routine head CT is controversial in asymptomatic patients. Chest imaging is important because SCLC can metastasize to the skin (see Table 3).

Fine-needle aspiration is used to assess metastatic spread. Octreotide scans help evaluate visceral metastases. Immunohistochemical analysis of sentinel lymph nodes (SLN) increases sensitivity of detecting clinically occult lymph node metastases, suggesting that SLN mapping and biopsy may be useful in staging and management of MCC. It is now recommended that all patients with MCC have routine sentinel lymph node biopsy (SLNB; Petrou, 2006). SLNB is much more sensitive than CT scan for detecting nodal disease. CT scans are helpful and should be used in patients with positive SLNB to help rule out distant metastasis (Petrou, 2006).

PATHOLOGY

Merkel cell carcinoma usually arises from the dermis, extends into the subcutis, and rarely involves the epidermis. Diagnosis by light microscopy is difficult because of the similarity between MCC and many other poorly differentiated small cell neoplasms, including SCLC, cutaneous large cell lymphoma, neuroblastoma, metastatic carcinoid, amelanotic melanoma, sweat gland carcinoma, Langerhans cell histiocytosis, and Ewing's sarcoma. Sixty percent of MCCs are misdiagnosed using light microscopy alone; ancillary techniques, such as electron microscopy and immunohistochemistry, are usually needed to make a definitive diagnosis. MCC tumors are classified into three cellular patterns: intermediate, small-cell type, and trabecular (see Table 4).

TABLE 4. Differential Diagnosis Based on Cellular Pattern (Petrou, 2006)

Cellular Pattern	Description and Differential Diagnosis
Intermediate	Most common cellular pattern
	Small blue cell tumors
	Melanoma
	Lymphoma
Small cell	Second most common cellular pattern
	Small cell carcinoma of the lung
Trabecular	Least common cellular pattern
	Metastatic carcinoid

Merkel cell carcinoma exhibits immunocytochemical properties of both epithelial and neuroendocrine cells. Immunoreactivity for intermediate filaments, including cytokeratins (CKs), distinguishes MCC from other undifferentiated tumors. Immunohistochemical detection of intermediate filaments, thyroid transcription factor-1, and neuroendocrine markers differentiates MCC from metastatic small cell cancer. Definitive diagnosis requires negative reactivity for S100, leukocyte common antigen, HMB-45 and NKI/C3, and low molecular weight CK 18 and 20 to rule out malignant melanoma and cutaneous lymphoma (Krasagakis & Tosca, 2003). Perinuclear dot-like staining pattern for CK 20 is specific for MCC and distinguishes it from oat cell carcinoma, a type of lung cancer. New evidence suggests great value in staining tumors with both thyroid transcription factor-1 and CK 20 to distinguish MCC from SCLC (Krasagakis & Tosca, 2003).

Electron microscopy helps confirm the diagnosis and may be essential if tumor identity is uncertain. Characteristic features, not found in any other primary cutaneous neoplasms, seen on electron microscopy include membrane-bound dense-core granules 75–200 nm wide and perinuclear whorls of intermediate filaments 7–10 nm wide. These findings confirm the diagnosis of MCC.

Many chromosomal abnormalities (gains, losses, and rearrangements) have been detected in MCC. The relationship of these genetic changes to pathogenesis is not clear. Similar chromosome gains and losses are seen in SCLC.

TREATMENT

No standard protocol exists; however, although optimal therapy is controversial, it is commonly agreed that multidisciplinary management yields the best patient outcomes. Therapy is primarily based on the presence or absence of metastases. Surgery is the primary treatment modality for MCC; there is no consensus for postsurgical adjuvant treatment with radiation therapy (RT) or chemotherapy.

Most treatment guidelines recommend wide excision of the primary tumor with or without adjuvant RT. A 1- to 3-cm margin of surrounding normal-appearing skin should be taken and confirmed by frozen section. Mohs micrographic surgery, a specialized surgery that provides precise removal of cancerous tissue while sparing healthy tissue, yields more favorable results in terms of local recurrence and tissue sparing compared with wide excision (Rigel et al., 2005).

Sentinel lymph node biopsy is important in the staging and treatment of MCC. Wide local excision followed by locoregional RT or elective lymph node dissection (ELND) is associated with longer time to recurrence, decreased local recurrence, and possibly improved survival (Rigel et al., 2005). Gagnon (2004) reported that SLNB was able to project which patients would need adjuvant chemotherapy and therapeutic lymph node dissection.

REGIONAL LYMPHADENECTOMY

Patients with clinically or radiographically diagnosed nodal disease should undergo therapeutic regional lymphadenectomy (Rigel et al., 2005; Tai, Yu, Tonita, & Gilchrist, 2000). ELND and prophylactic RT are controversial (Tai et al., 2000). Approximately 50% of patients eventually develop locoregional recurrence after resection of tumor alone (Tai et al., 2000). ELND is often used for patients with Stage I disease, without nodal involvement; however, no data exist to determine if ELND prolongs survival (Tai et al., 2000). Some researchers suggest that ELND be used only for patients with head and neck primary tumor sites, tumors larger that 1.5 cm in diameter, or when there is histological evidence of lymphatic or vascular invasion (Tai et al., 2000). There are no reliable factors to determine relative risk for regional recurrence. All MCC patients should be considered high risk (Rigel et al., 2005; Tai et al., 2000).

SENTINEL LYMPH NODE MAPPING AND SELECTIVE LYMPHADENECTOMY

Intraoperative SLN mapping and selective lymphadenectomy are commonly used for melanoma patients (Rigel et al., 2005; Tai et al., 2000). As Merkel cell carcinoma is a very rare tumor, treatment for MCC is based on what we have learned to be effective treatments for melanoma. This procedure has less inherent morbidity than that of total lymphadenectomy. The technique involves preoperative injection of radioactive sulfur colloid and radiolymphoscintigraphic localization and/or intraoperative SLN localization with vital blue dye injection (Rigel et al., 2005). If the SLN is histologically negative, the likelihood of other residual nodal disease is low (Tai et al., 2000). If the SLN is histologically positive, a formal lymphadenectomy is required (Tai et al., 2000). Positive SLN is associated with higher rates of recurrence.

RADIATION THERAPY

New data support RT as a standard treatment in most cases of MCC. Petrou (2006) asserted that RT decreases the rate of local recurrence and therefore reduces morbidity. According to the National Comprehensive Cancer Network, if SLNB is not performed following local excision of the primary tumor, postoperative RT to the primary site, in-transit lymphatics, and draining nodal basins are recommended. If SLNB is negative, postoperative RT to only the primary site is indicated. If SLNB is positive, postoperative RT to primary tumor site, in-transit lymphatics, and draining nodal basins, with or without therapeutic lymph node dissection, are advised. In cases of positive SLN by immunohistochemical methods only, RT may be considered. When lymph nodes

are clinically positive, in the absence of distant metastatic disease, treatment mirrors that of patients with positive SLNB (Rigel et al, 2005; Tai et al., 2000). In patients with inoperable disease, RT may be used as monotherapy (Petrou, 2006).

Merkel cell carcinoma is a radiosensitive tumor (Krasagakis & Tosca, 2003; Rigel et al., 2005; Tai et al., 2000). RT to the primary tumor site after local excision is used as adjunctive therapy (Tai et al., 2000). RT has been advocated to decrease locoregional recurrence rates, prolong time to disease progression, and improve survival. RT is recommended in cases of local recurrence and regional lymph node involvement. Post-excision RT is associated with lower rates of local and regional recurrence. Adjuvant RT is associated with significantly higher 2-year disease-free intervals and fewer local recurrences at 18 months. Response to RT is reported in up to 96% of cases. Unfortunately, adjuvant RT has not had a significant impact on overall survival and has not been shown to improve survival (Krasagakis & Tosca, 2003; Tai et al., 2000).

The National Comprehensive Cancer Network guidelines recommend using adjuvant nodal RT when SLN biopsy is not performed or when regional lymph nodes are clinically positive (Tai et al., 2000). It is recommended that the surgical bed and the draining regional lymphatics be irradiated, if technically possible (Rigel et al., 2005; Tai et al., 2000). All regional lymphatics must be irradiated to avoid geographic misses (Rigel et al., 2005; Tai et al., 2000). Studies show that a median wait of 24 days for RT commencement was associated with a high risk of disease progression (Tai et al., 2000).

Radiation therapy is recommended for patients with high-risk features including tumor larger than 2 cm in diameter, positive resection margins or tumors closely approximating the margins, angiolymphatic invasion, and positive regional lymph nodes or when regional lymph nodes were not pathologically staged and for immunocompromised patients (Tai et al., 2000).

Postoperative (Adjuvant) Chemotherapy

Merkel cell carcinoma is a chemotherapy-sensitive tumor; yet, adjuvant chemotherapy for MCC has not been extensively studied. In the literature, adjuvant chemotherapy is associated with significantly worse outcomes because patients who received it had very high risk or recurrent tumors (Tai et al., 2000). It is controversial whether adjuvant chemotherapy is beneficial for more advanced locoregional disease (Tai et al., 2000).

Chemotherapy for MCC is based on success in treating neuroendocrine carcinomas in other sites. There is no specific MCC chemotherapy regimen; a number of chemotherapy regimens are used. Nghiem and Jaimes (2008) recommended using carboplatin and etoposide. Chemotherapy would be indicated for recurrent and metastatic MCC, but these most often occur in older patients who tolerate aggressive treatment poorly (Rigel et al., 2005).

Most clinicians and institutions use chemotherapy, with or without surgery, only in cases of distant metastatic disease because the benefits of adjuvant chemotherapy have not been demonstrated in clinical trials. Generally, patients are treated according to small cell chemotherapy regimens, most commonly cyclophosphamide/doxorubicin/vincristine or carboplatin/etoposide/vincristine; both regimens produce overall response rate of 76% and complete response rate of 35% (Tai et al., 2000). The 2- and 5-year survival rates after treatment with both chemotherapy regimens are 36% and 17%, respectively (Tai et al., 2000). Use of etoposide with carboplatin or cisplatin may be preferable for patients with cardiac disease (Tai et al., 2000).

Although no prospective studies currently exist to show that adjuvant chemotherapy prolongs survival in patients with Stage II or regional nodal disease, some authors recommend it as a "last measure" to prevent distant metastases (Garneski & Nghiem, 2007; Krasagakis & Tosca, 2003, p. 673). In patients with multifocal metastatic spread, in cases where RT did not result in remission, or in conjunction with RT, Krasagakis and Tosca (2003) suggested that chemotherapy may be beneficial (2003).

For patients with recurrent or locally advanced MCC, local excision with combined chemotherapy and RT is the treatment of choice (Tai et al., 2000). Despite the conclusion of several studies that there is no association between adjuvant chemotherapy and survival for patients with Stage II disease, adjuvant chemotherapy should be considered for treatment of high-risk disease. Chemotherapy with RT may provide better palliation than chemotherapy alone for patients with advanced disease (Tai et al., 2000). Krasagakis and Tosca (2003) recommended chemotherapy for palliation in patients with Stage III disease. In these patients, response rates are around 50% and are usually short-lived; second-line chemotherapy is almost always required.

Adjuvant chemotherapy is not recommended for patients with node-negative disease. It is important to report treatment results to help identify optimal chemotherapy regimens (Rigel et al., 2005; Tai et al., 2000).

In comparison, some researchers do not support the use of adjuvant (Petrou, 2006). Some data suggest that, in addition to unpleasant chemotherapy-related morbidity, including fever, neutropenia, and sepsis, adjuvant chemotherapy is associated with lower rates of survival in Stage II disease and a decreased quality of life (Petrou, 2006). In addition, the immune system plays a complex and not fully understood role in the battle against MCC. Until more research is done about the way the immune system affects MCC, Petrou (2006) suggested that chemotherapy should not be used because it suppresses the immune system.

COURSE AND PROGNOSIS

Merkel cell carcinoma can have a variable course. Some patients with localized primary tumors have long-term control with local excision only (Tai et al., 2000). Most MCC tumors behave aggressively, like thick or ulcerated melanomas, in their propensity for locoregional recurrence and early lymph node metastases. Survival rates for MCC with nodal or systemic disease are similar to those of malignant melanoma (Tai et al., 2000).

Significantly favorable prognostic factors for overall survival include initial localized disease with negative lymph nodes, tumor occurring not on the head or neck, being female, age younger than 60–65 years at time of diagnosis, and absence of comorbidities. Histological features associated with poor survival rates include large tumor size, high mitotic rate, and small cell size (Krasagakis & Tosca, 2003). Researchers have found that gender and the presence of nodal disease are significant predictors of survival and distant metastasis. Median survival with and without regional nodal involvement is 13 versus 40 months, respectively (Tai et al., 2000).

Prognosis is impacted by primary tumor site (Tai et al., 2000). Truncal lesions, especially those in the vulvar or perianal areas, have the worst prognosis, possibly related to late detection (Rigel et al., 2005; Tai et al., 2000). MCC occurring on the legs has a high recurrence rate due to poor blood supply and poor tolerance of high-dose RT (Rigel et al., 2005; Tai et al., 2000). Systemic disease is associated with particularly poor prognosis (Rigel et al., 2005; Tai et al., 2000).

Following initial treatment, local recurrence occurs in 29%–43% of patients, at a median of 4 months, and usually within 1 year of initial therapy. Following margin-negative excision, local recurrence rate is reduced to 8% (Tai et al., 2000). Nodal and distant metastasis occurs in 33% of cases, each. Patients with an initial nodal recurrence are at higher risk of developing other distant metastases than are patients without nodal recurrence (Tai et al., 2000).

After primary tumor resection, the median time to develop clinically detectable nodal recurrence is 7 to 8 months. Once local or nodal recurrence occurs, combination therapy provides the best survival potential (Rigel et al., 2005; Tai et al., 2000). Eleven to 66% of patients who present with Stage II disease or have local recurrence die of MCC within 5 years (Tai et al., 2000). When recurrence occurs and is treated, median overall survival is 27 months (Tai et al., 2000). Median overall survival is 31 months.

Most recurrences and deaths from MCC occur within 3 years of diagnosis. Mean time to local or regional recurrence is approximately 8 months; mean time to distant or systemic metastasis is approximately 18 months. Within 2 years of diagnosis, nearly 50% of patients develop systemic recurrence; 65%–75% die of MCC (Tai et al., 2000).

FOLLOW-UP

It is generally agreed that patients should have regular skin and lymph node examinations every 3–6 months for the first 3 years and annually thereafter. Annual chest radiograph is indicated. CT scans of the chest, abdomen, or head may be needed for symptomatic patients (Tai et al., 2000). If recurrence is detected, a full staging workup should be performed (Tai et al., 2000).

Second Primary Cancers

Merkel cell carcinoma is associated with high incidences of other skin tumors and hematologic malignancies. Up to 25% of patients with MCC have had a second neoplasm, half of which are squamous cell carcinoma. Patients who develop second neoplasms usually have higher MCC-specific mortality rates.

ROLE OF THE DERMATOLOGY NURSE AND NURSE PRACTITIONER

Because MCC is rare, it is unlikely to be encountered on a regular basis. However, because of its aggressive nature and quick progression to advanced disease, it is necessary that all dermatology nurses and nurse practitioners be knowledgeable about MCC so that rapid diagnosis may be made and appropriate treatment initiated.

In addition, most patients are not familiar with MCC. Therefore, patient education must be comprehensive. Patients will undergo multiple procedures, surgeries, and treatments and may be apprehensive or confused about their care. Having a thorough explanation about their diagnosis, prognosis, and treatment options will be comforting for the patient and his or her family. Having a unified, integrated approach to treatment, with health-care providers from dermatology, surgery, oncology, and radiation oncology working together, is encouraged to facilitate treatment and management of these patients. Dermatology nurses may be required to treat radiation dermatitis, dress surgical wounds, or manage stomatitis. Being aware of the most common clinical features of MCC and being able to anticipate treatment complications will simplify the management of and be comforting to patients with MCC.

CONCLUSION

Despite advances in diagnosis, overall prognosis for patients with MCC is poor. Recent discoveries in pathology of disease and advances in disease treatment and management paint a brighter picture for patients with MCC. With the discovery of MCV, more research is needed to determine possible methods of prevention. Meanwhile, optimal treatment modalities still remain poorly defined due to the lack of randomized control studies and the rarity of MCC (Rigel et al., 2005). Currently, wide local excision of the primary lesion remains the standard treatment of MCC (Rigel et al., 2005). It remains unclear whether SLN

mapping versus ELND will improve overall patient survival (Rigel et al., 2005). More research is needed so that providers can rapidly diagnose, treat, and manage these unique and aggressive tumors. ■

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