

Merkel Cell Carcinoma: Questions and Answers

Key Points

- Merkel cell carcinoma (MCC) is a rare, aggressive type of skin cancer that forms on or just under the skin (see Question 1).
- MCC has been linked to sun exposure, with most cases occurring on sun-exposed areas of the body (see Questions 3 and 5). MCC has also been linked to immunosuppression, exposure to other sources of ultraviolet light, and certain diseases (see Question 3).
- MCC differs from most other skin cancers in that it grows rapidly over a few weeks or months (see Question 4).
- Surgery and radiation are the usual treatments for MCC (see Question 7).
- People with cancer are encouraged to enroll in clinical trials (research studies with people) that explore new treatments (see Question 8).

1. What is Merkel cell carcinoma?

Merkel cell carcinoma (MCC) is a rare, aggressive type of skin cancer that forms on or just under the skin. It is also called primary small cell carcinoma of the skin, trabecular carcinoma, APUDoma, neuroendocrine carcinoma, endocrine carcinoma, or primary undifferentiated tumor of the skin (1). MCC is believed to start in neuroendocrine cells called Merkel cells. These cells release hormones into the blood when stimulated by the nervous system. They migrate from part of the nervous system called the neural crest to the skin (2). Merkel cells are believed to play a role in making the skin sensitive to touch (3).

2. How often does Merkel cell carcinoma occur?

Approximately 1,200 new cases of MCC are diagnosed in the United States each year (4), compared with almost 60,000 new cases of melanoma and more than 1 million new cases of nonmelanoma skin cancer. The incidence of MCC has been rising, with a 3-fold



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increase between 1986 and 2001 (4). Most patients diagnosed with MCC are over age 50 at diagnosis (the average age is 69), with only 5 percent of cases diagnosed in those under age 50 (5). MCC is more common in white people than in other racial/ethnic groups. Some cases have been reported in Japanese people, but very few have been seen in black people (6).

3. What are the possible causes of Merkel cell carcinoma?

The exact cause of MCC is unknown, but it appears to be linked to sun exposure and immunosuppression (suppression of the body's immune system and its ability to fight infections or disease) (2). Sun exposure as a risk factor for MCC is supported by data that show a rise in incidence corresponding with the solar UVB index (scale indicating the intensity of solar ultraviolet-B (UVB) radiation at noon for a particular location) (6). MCC has been linked to conditions such as HIV infection, chronic lymphocytic leukemia, Hodgkin lymphoma (cancer of the lymph system), ectodermal dysplasia (a disease involving abnormal tissue development), and Cowden disease (a disease in which masses of abnormal but benign tissues grow in multiple sites in the body). Other possible causes include exposure to arsenic and treatment for psoriasis that uses psoralens (a medication that causes the skin to become sensitive to light) and ultraviolet-A light (PUVA) (2).

4. What are the symptoms of Merkel cell carcinoma?

The most common symptom of any skin cancer, including MCC, is a change in the skin, especially a change in an existing mole or a new growth. MCC appears as a firm, painless lump within the skin that may resemble a cyst but is fixed; i.e., cannot be moved. The lump is usually less than 2 cm (about $\frac{3}{4}$ inch) in size and can be red, pink, or blue-violet. MCC is different from other skin cancers in that it grows rapidly over a few weeks or months (5).

5. Where does Merkel cell carcinoma develop?

MCC is usually found on sun-exposed areas of the body. Fifty percent of cases occur on the head and neck, especially around the eye and on the eyelid (1). Forty percent of cases occur on the arms and legs (2). MCC has also been found on the trunk and other areas of the skin that are not usually exposed to the sun (1).

6. How is Merkel cell carcinoma diagnosed and staged?

The doctor may use the following procedures and tests to diagnose MCC. Some of these tests are also used to help determine the stage of the disease. Stage is a description of the extent of cancer.

- A **biopsy** is the removal of cells or tissue from a tumor for examination by a pathologist. The pathologist may study tissue samples under a microscope or perform other tests on the cells or tissue. Biopsies are used for both diagnosis and staging.

The surgeon may also remove lymph nodes (small, round organs that trap cancer cells, bacteria, or other harmful substances) to help determine the stage of the disease.

- **Sentinel lymph node (SLN) biopsy** is a procedure in which the sentinel lymph node is removed and examined under a microscope to determine whether cancer cells are present. The sentinel lymph node is the first lymph node to which cancer is likely to spread from the primary tumor. SLN biopsy is used to help determine the stage of the disease. SLN biopsy may cause fewer side effects than standard lymph node removal because fewer lymph nodes are taken out.
- **Immunohistochemistry** (staining of cells with agents that react with antibodies on the surface of cancer cells) is a laboratory technique used to tell the difference between MCC and other types of cancer (2).
- **Computed tomography (CT)**, a procedure that uses special x-ray equipment to obtain cross-sectional pictures of the body, can distinguish MCC from small cell lung cancer and show whether the disease has metastasized (spread) to other parts of the body (5).
- In an **octreotide scan** (sometimes called Somatostatin Receptor Scintigraphy or SRS), the doctor injects a small amount of a radioactive drug into a vein. The drug travels through the bloodstream and attaches to tumor cells. A machine called a scanner detects the radioactive material and creates scans (pictures) showing where the tumor cells are located in the body. For MCC, this test can be used for both diagnosis and staging (5).
- A **PET scan** uses radioactive sugar, which is absorbed by cancer cells and appears as dark areas on the scan. It can be used for both diagnosis and staging of MCC.

7. **How is Merkel cell carcinoma treated?**

Surgery is the most common treatment for MCC. Surgery with wide margins (a large border of healthy tissue removed with the tumor) is the recommended treatment for MCC. Mohs micrographic surgery, a technique in which individual layers of tissue are removed and examined under a microscope until all cancerous tissue has been removed, may be used instead of traditional surgery with wide margins. Mohs micrographic surgery may be a good alternative for MCC tumors on highly visible areas such as the face, and in areas where the surgeon would not be able to obtain wide margins (5).

The surgeon may remove lymph nodes to help stage the disease or to prevent recurrence (cancer coming back). The patient may also receive adjuvant radiation therapy (treatment given after the primary therapy) to decrease the chance of recurrence. Chemotherapy is the usual treatment if the disease has spread beyond the lymph nodes to areas that are not treatable by radiation therapy.

Supportive care is treatment given to improve the quality of life of patients who have a serious or life-threatening disease, such as cancer. It prevents or treats as early as possible the symptoms of the disease, side effects caused by treatment of the disease, and psychological, social, and spiritual problems related to the disease or its treatment. For example, anticancer drugs such as carboplatin and etoposide may be given to relieve symptoms in some patients with MCC. Radiation may be used to relieve pain from MCC that has metastasized to the brain or bones, and to reduce discomfort from skin problems associated with MCC (2). Additionally, meeting with a social worker, counselor, or member of the clergy can be helpful to those who want to talk about their feelings or discuss their concerns. A social worker can often suggest resources for help with recovery, emotional support, financial aid, transportation, or home care.

8. Are clinical trials (research studies) available? Where can people get more information about clinical trials?

Yes. The National Cancer Institute (NCI), a component of the National Institutes of Health, is sponsoring clinical trials that are designed to find new treatments and better ways to use current treatments. Before any new treatment can be recommended for general use, doctors conduct clinical trials to find out whether the treatment is safe for patients and effective against the disease. Participation in clinical trials may be a treatment option for patients with MCC.

People interested in taking part in a clinical trial should talk with their doctor. Information about clinical trials is available from the NCI's Cancer Information Service (CIS) (see below) at 1-800-4-CANCER and in the NCI booklet *Taking Part in Cancer Treatment Research Studies*, which can be found at <http://www.cancer.gov/publications> on the Internet. This booklet describes how research studies are carried out and explains their possible benefits and risks. Further information about clinical trials is available at <http://www.cancer.gov/clinicaltrials> on the NCI's Web site. The Web site offers detailed information about specific ongoing studies by linking to PDQ®, the NCI's comprehensive cancer information database. The CIS also provides information from PDQ.

9. What is the prognosis for patients with Merkel cell carcinoma?

Prognosis describes the likely course and outcome of a disease—that is, the chance that a patient will recover or have a recurrence. The prognosis for MCC patients depends greatly on the stage of the disease at the time of diagnosis. If the tumor is small (less than 2 cm or about $\frac{3}{4}$ inch) and cancer cells have not spread to the lymph nodes, the 5-year survival rate is more than 90 percent. Patients with MCC that has spread to the lymph nodes have a 5-year survival rate of about 50 percent. Overall 5-year survival for patients diagnosed with MCC is 64 percent (7), but half of patients with advanced MCC will live only 9 months (2). The disease recurs in about 50 percent of patients (7). It is important to keep in mind, however, that these statistics are averages based on large numbers of patients. Statistics cannot be used to predict what will happen to a particular patient because each person's situation is unique.

Selected References

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3. Camisa C, Weissmann A. Friedrich Sigmund Merkel. Part II. The cell. *American Journal of Dermatopathology* 1982; 4(6):527–535.
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7. Allen PJ, Bowne WB, Jaques DP, et al. Merkel cell carcinoma: Prognosis and treatment of patients from a single institution. *Journal of Clinical Oncology* 2005; 23(10): 2300–2309.

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Related NCI materials and Web pages:

- National Cancer Institute Fact Sheet 5.2, *Computed Tomography (CT): Questions and Answers* (<http://www.cancer.gov/cancertopics/factsheet/Detection/CT>)
- National Cancer Institute Fact Sheet 5.32, *Staging: Questions and Answers* (<http://www.cancer.gov/cancertopics/factsheet/Detection/staging>)
- National Cancer Institute Fact Sheet 6.7, *Cancer: Questions and Answers* (<http://www.cancer.gov/cancertopics/factsheet/Sites-Types/general>)
- National Cancer Institute Fact Sheet 6.20, *Metastatic Cancer: Questions and Answers* (<http://www.cancer.gov/cancertopics/factsheet/Sites-Types/metastatic>)
- National Cancer Institute Fact Sheet 7.44, *Sentinel Lymph Node Biopsy: Questions and Answers* (<http://www.cancer.gov/cancertopics/factsheet/therapy/sentinel-node-biopsy>)
- *Taking Part in Cancer Treatment Research Studies* (<http://www.cancer.gov/clinicaltrials/Taking-Part-in-Cancer-Treatment-Research-Studies>)
- *What You Need To Know AboutTM Skin Cancer* (<http://www.cancer.gov/cancertopics/wyntk/skin>)

For more help, contact:

NCI's Cancer Information Service

Telephone (toll-free): 1-800-4-CANCER (1-800-422-6237)

TTY (toll-free): 1-800-332-8615

LiveHelp[®] online chat: <https://cissecure.nci.nih.gov/livehelp/welcome.asp>

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