

TREATMENT UPDATE:

Merkel Cell Carcinoma

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Treatment Update: Merkel Cell Carcinoma

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Merkel cell carcinoma (MCC) is a rare, aggressive type of skin cancer. Most cases of MCC are found in older people or those with weakened immune systems.

Merkel cells are found in the epidermis, the outermost layer of the skin. MCC starts when Merkel cells start to grow out of control. It usually appears as a painless bump, often on sun-exposed areas such as the face, head or neck.

Diagnosis

The diagnostic process for MCC often begins with a person pointing out a rapidly-growing bump (on or under the skin) to their doctor. The bump (tumor) may be flesh-colored or appear in shades of red, blue or purple.

A skin biopsy will be performed if MCC is suspected. In this procedure, a doctor removes a small sample of skin from the tumor, which is then examined by a pathologist to determine if it contains abnormal cells.



Other tests may be conducted to determine if the MCC has spread beyond the skin, including:

- **Sentinel node biopsy.** A sentinel node biopsy is a procedure to determine if the MCC has spread to the lymph nodes, small organs located throughout the body that remove waste and fluids and help fight infection. Dye is injected near the tumor and flows through the lymphatic system to the lymph nodes. The first nodes to receive the dye (the sentinel nodes) are removed and examined under a microscope for the presence of cancer cells.
- **Imaging tests.** In the majority of cases, a positron emission tomography (PET) or computerized tomography (CT) scan is recommended at the time of diagnosis. Other types of imaging tests that may be used include chest X-rays and octreotide scans (a test that uses an injection of a radioactive tracer to look for the spread of abnormal cells).

Treatment Options

Treatment options for MCC often depend on whether the cancer has spread beyond the skin. You and your doctor will discuss the approach that is right for you, based on your individual circumstances. Often, treatment will consist of a sequence of therapies rather than just one approach.

Surgery

The tumor, along with a surrounding border of normal skin, is removed with a scalpel by a surgical oncologist. If diagnostic tests showed evidence that the MCC has spread to lymph nodes, those lymph nodes are also removed. This is called lymph node dissection.

Radiation

Radiation therapy is often administered after surgery to destroy small traces of cancer that could not be seen and removed during surgery. Radiation can also be used to treat the main tumor if surgery is not an option, and is a possible treatment approach if the MCC has spread to the lymph nodes or beyond.

If MCC recurs (returns) after initial treatment, it can be treated with external beam radiation therapy (EBRT), either alone or in combination with surgery or other treatment types. EBRT uses a machine that directs a beam (or multiple beams) of radiation to the tumor. The use of imaging tests allows radiation oncologists to accurately target the MCC, helping to spare healthy tissues.

Immunotherapy

Our immune system is constantly working to keep us healthy. It recognizes and fights against danger, such as infections, viruses and growing cancer cells. In general terms, immunotherapy uses our own immune system as a treatment against cancer.

In 2017, the U.S. Food and Drug Administration (FDA) approved avelumab (Bavencio) for the treatment of MCC. Avelumab is an “immune checkpoint inhibitor” that works by targeting PD-L1, a protein that can prevent the body’s immune system from attacking tumors. It is given as an infusion into a vein.

Other immune checkpoint inhibitors, including nivolumab (Opdivo) and ipilimumab (Yervoy) are being studied in clinical trials for the treatment of MCC. Nivolumab works by blocking the PD-1 cellular pathway (a pathway that restricts the body’s immune system from attacking cancer cells). Ipilimumab blocks the action of a protein called CTLA-4. By doing so, it is thought to help the immune system destroy cancer cells.

Chemotherapy

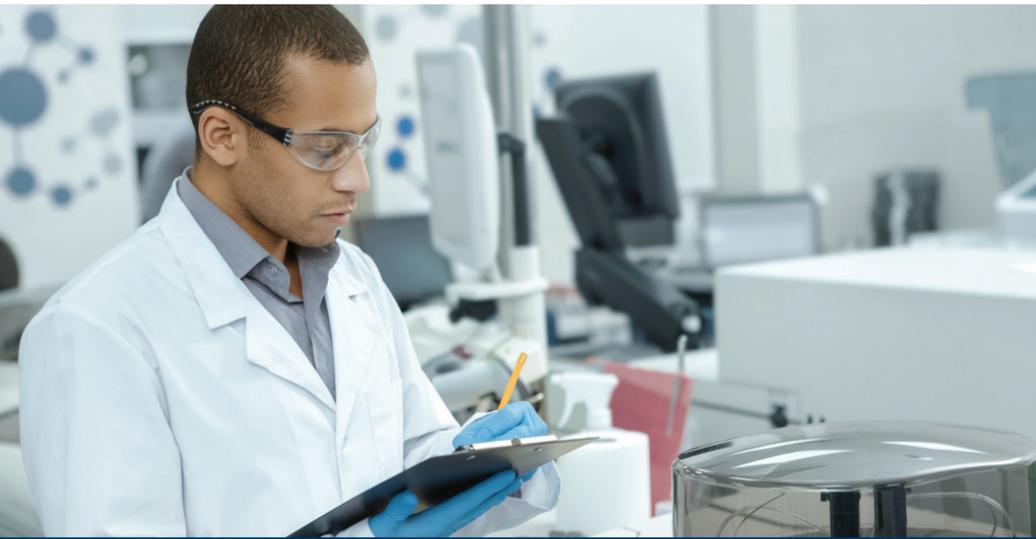
Chemotherapy is the use of medication to treat cancer by stopping the ability of cancer cells to grow and divide. It is usually given intravenously (into a vein through a needle) or via a pill or capsule.

In the treatment of MCC, chemotherapy is most likely to be beneficial when the cancer has spread beyond the skin to other organs. However, it's important to note that immunotherapy (not chemotherapy) is typically used as a firstline (initial) treatment for MCC that has spread beyond the skin.

The types of chemotherapy most often used to treat MCC include:

- Cisplatin (Platinol, Platinol AQ)
- Carboplatin (Paraplatin)
- Etoposide (Vepesid)
- Topotecan (Hycamtin)

A combination of chemotherapies may also be given, consisting of cyclophosphamide, doxorubicin and vincristine.



Treatment Side Effects

All cancer treatments can cause side effects. It's important that you report any side effects you experience to your health care team so they can help you manage them. Report them right away—don't wait for your next appointment. Doing so will improve your quality of life and allow you to stick with your treatment plan. It's important to remember that not all people experience all side effects, and people may experience side effects not listed here.

Side Effects of Radiation Therapy

Changes to the skin are the most common side effects of radiation therapy. The changes can include dryness, swelling, peeling, redness and blistering. If a reaction occurs, contact your health care team so the appropriate treatment can be prescribed. It's especially important to contact your health care team if there is any open skin or painful area, as this could indicate an infection. Infections can be treated with an oral antibiotic or topical antibiotic cream.

Side Effects of Immunotherapy

Immunotherapy travels through the bloodstream, helping to prompt an immune response. Sometimes the immune system attacks healthy cells as well as cancer cells, and certain side effects may be experienced, including fatigue, decreased appetite and digestive tract symptoms. The management of these potential side effects is discussed later in this publication.

Some people being treated with avelumab may experience side effects that can include fever, chills, flushing of the face, rash and trouble breathing. There are medicines that can be given prior to each infusion to lower the risk of this reaction occurring.

Side Effects of Chemotherapy

The side effects of chemotherapy depend on the type and dose of drugs given and the length of time they are used, and can include:

- Achiness
- Hair loss
- Increased risk of infection (from having too few white blood cells)
- Easy bruising or bleeding
- Changes in memory or thinking
- Peripheral neuropathy (numbness or tingling in hands and feet)



The Importance of Clinical Trials

Clinical trials are the standard by which we measure the worth of new treatments and the quality of life of patients as they receive those treatments. For this reason, doctors and researchers urge people with cancer to take part in clinical trials.

Your doctor can guide you in making a decision about whether a clinical trial is right for you. Here are a few things that you should know:

- Often, people who take part in clinical trials gain access to and benefit from new treatments.
- Before you participate in a clinical trial, you will be fully informed as to the risks and benefits of the trial, including any possible side effects.
- Most clinical trials are designed to test a new treatment against a standard treatment to find out whether the new treatment has any added benefit.
- You can stop taking part in a clinical trial at any time for any reason.



General Side Effects

Some side effects may occur across treatment approaches. This section provides tips and guidance on how to manage these side effects should they occur.

Managing Digestive Tract Symptoms

Nausea and vomiting

- Avoid food with strong odors, as well as overly sweet, greasy, fried or highly seasoned food.
- Nibble on dry crackers or toast. These bland foods are easy on the stomach.
- Having something in your stomach when you take medication may help ease nausea.

Diarrhea

- Drink plenty of water. Ask your doctor about using drinks such as Gatorade, which provide electrolytes. Electrolytes are body salts that must stay in balance for cells to work properly.
- Over-the-counter medicines such as loperamide (Imodium A-D and others) and prescription drugs are available for diarrhea but should be used only if necessary. If the diarrhea is bad enough that you need medicine, contact a member of your health care team.
- Choose foods that contain soluble fiber, like beans, oat cereals and flaxseed, and high-pectin foods such as peaches, apples, oranges, bananas and apricots.
- Avoid food high in refined sugar and those sweetened with sugar alcohols such as sorbitol and mannitol.

Loss of appetite

- Eating small meals throughout the day is an easy way to take in more protein and calories, which will help maintain your weight. Try to include protein in every meal.
- To keep from feeling full early, avoid liquids with meals or take only small sips (unless you need liquids to help swallow). Drink most of your liquids between meals.
- Keep high-calorie, high-protein snacks on hand such as hard-boiled eggs, peanut butter, cheese, ice cream, granola bars, liquid nutritional supplements, puddings, nuts, canned tuna or trail mix.
- If you are struggling to maintain your appetite, talk to your health care team about whether appetite-building medication could be right for you.



Managing Pain

There are a number of options for pain relief, including prescription and over-the-counter medications. It's important to talk to a member of your health care team before taking any over-the-counter medication to determine if it is safe and will not interfere with your treatment. Many pain medications can lead to constipation, which may make your pain worse. Your doctor can prescribe medications that help to avoid constipation.

Physical therapy, acupuncture and massage may also be of help in managing your pain. Consult with a member of your health care team before beginning any of these activities.

Managing Fatigue

Fatigue (extreme tiredness not helped by sleep) is one of the most common side effects of many cancer treatments. If you are taking a medication, your doctor may lower the dose of the drug, as long as it does not make the treatment less effective. If you are experiencing fatigue, talk to your doctor about whether taking a smaller dose is right for you.

There are a number of other tips for reducing fatigue:

- Take several short naps or breaks during the day.
- Take walks or do some light exercise, if possible.
- Try easier or shorter versions of the activities you enjoy.
- Ask your family or friends to help you with tasks you find difficult or tiring.

There are also prescription medications that may help, such as modafinil. Your health care team can provide guidance on whether medication is the right approach for your individual circumstances.



Communicating with Your Health Care Team

As you manage your MCC, it's important to remember that you are a consumer of health care. The best way to make decisions about health care is to educate yourself about your diagnosis and get to know the members of your health care team, including doctors, nurses, nurse practitioners, physician assistants, dietitians, social workers and patient navigators.

Here are some tips for improving communication with your health care team:

Start a health care journal. Having a health care journal or notebook will allow you to keep all of your health information in one place. You may want to write down the names and contact information of the members of your health care team, as well as any questions for your doctor.

Prepare a list of questions. Before your next medical appointment, write down your questions and concerns. Because your doctor may have limited time, ask your most important questions first and be as specific as possible.

Bring someone with you to your appointments. Even if you have a journal and a prepared list of questions or concerns, it's always helpful to have support when you go to your appointments. The person you bring may also think of questions to ask your doctor or remember details about your symptoms or treatment that you may have forgotten.

Write down your doctor's answers. Taking notes will help you remember your doctor's responses, advice and instructions. You can also ask the person who accompanies you to take notes for you, either in your journal or on a tablet or smartphone.

Record your visit if your doctor allows it. Recording the conversation with your doctor gives you a chance to hear specific information again or share it with family members or friends.

Incorporate other health care professionals into your team.

Your medical oncologist is an essential member of your health care team, but there are other health care professionals who can help you manage your diagnosis and treatment:

- Your primary care physician should be kept updated about your cancer treatment and any test results.
- Your dermatologist can be of help in minimizing any skin-related side effects that you may experience.
- Your local pharmacist is a great source of knowledge about the medications you are taking. Have all of your prescriptions filled at the same pharmacy to avoid the possibility of harmful drug interactions.
- Make sure your oncologist knows of any other medical conditions you have or any pain you are experiencing so that they can consult with your primary care physician or specialist as needed.

Remember, there is no such thing as over-communication.

CancerCare's Free Support Services and Programs

It can be very difficult to receive a diagnosis of MCC, and adjusting to the necessary changes in your life can be challenging.

CancerCare can help. We are a national nonprofit organization providing free, professional services to anyone affected by cancer. Our licensed oncology social workers can provide support and education, help in navigating the complicated health care system and offer information on support groups and other resources.

To learn more about how CancerCare helps, call us at 800-813-HOPE (4673) or visit www.cancercare.org.

You will likely also build your own personal support network composed of family and friends. In doing so, it's best to take some time to think about the people in your life and how they are best suited to help. Match the task to their strengths—ask a family member who loves to shop to pick up something for you at the store, or ask a friend who's a good listener to come over for a chat.



Frequently Asked Questions

Q: What causes MCC?

A: Researchers have discovered that a virus, Merkel cell polyomavirus, plays a role in causing many cases of MCC. This virus, which lives on the skin, doesn't cause any signs or symptoms. Given that this is a common virus and MCC is very rare (approximately 2,000 cases diagnosed in the United States annually), it's likely that MCC has other causes (including sun exposure), not all of which are completely understood.

Q: Even if the causes of MCC are not fully understood, are there risk factors that increase its likelihood?

A: There are factors that may increase the risk of MCC, which include:

- **Excessive exposure to ultraviolet light.** Although MCC tumors can develop anywhere on the body, most appear on skin surfaces frequently exposed to the sun.
- **Older age.** While MCC can occur at any age, it is most common in people over the age of 50.
- **A weakened immune system.** People with weakened immune systems, either due to medicines that are being taken or because of another health condition, are more likely to develop MCC.
- **History of other skin cancers.** MCC is associated with a history of other skin cancers, such as basal cell or squamous cell carcinoma.
- **Skin color.** MCC usually occurs in people with light-colored skin.

Q: Does it matter at what hospital or health care facility my MCC treatment takes place?

A. The effective treatment and ongoing management of MCC almost always involves a multi-disciplinary approach, including the specialties of surgery, dermatology, radiation oncology and medical oncology. As MCC is rare, being treated by a team that is expert in the management of MCC is important. Resources can be found at www.merkelcell.org/resources/find-a-specialist.

Q. What is a treatment summary and why is important?

A: A treatment summary, sometimes called a “shadow chart,” is a document that you create and keep in your possession. Maintaining your own records allows you and your family members to have instant access to the specifics of your MCC diagnosis and treatment. A treatment summary should include:

- Your name and date of birth
- Date of diagnosis
- Prescribed therapy/therapies, including dates started and stopped and dosages when appropriate
- Dates and types of post-diagnosis testing, and the results of these tests
- Other medications and supplements you are taking
- Names, affiliations and contact information of all members of your health care team

Talk to your doctor or a member of your health care team about your intention to create a treatment summary, and ask what else they suggest be included. Take your treatment summary with you when you visit any doctor, not just your oncologist.

Notes

Resources

CancerCare®

800-813-HOPE (800-813-4673)
www.cancercares.org

American Cancer Society

800-227-2345
www.cancer.org

Cancer.Net

Patient information from
the American Society of
Clinical Oncology
888-651-3038
www.cancer.net

National Cancer Institute

800-422-6237
www.cancer.gov

CLINICAL TRIALS WEBSITES**EmergingMed**

www.emergingmed.com

National Cancer Institute

www.cancer.gov

Merkelcell.org

www.merkelcell.org

The Skin Cancer Foundation

212-725-5176
www.skincancer.org

Medicine Assistance Tool

www.medicineassistancetool.org

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