Merkel Cell Carcinoma

Epidemiology
Merkel Cell Carcinoma (MCC) is a relatively rare skin cancer, occurring in less than one person per every 100,000 people. It is much less common than melanoma, squamous cell carcinoma, and basal cell carcinoma, although it is becoming more frequent with time. Patients who develop these cancers are usually older, with an average age over 70 years. Less than five percent of cases occur in patients under 50 and these tumors are very rare in children. Almost all patients with MCC are white, and most tumors occur in the head and neck.

Risk Factors
Ultraviolet radiation from sun exposure or tanning booth use increases risk of developing these tumors, as does suppression of the immune system from AIDS or organ transplant. MCC is also linked to a virus called Merkel Cell Polyomavirus, which is found on the skin of about 80% of adults and in most people causes no problems.

Presentation
The most common way that people notice MCC is the development of a flesh-colored or bluish-red nodule on the skin. Almost 90% of these nodules are painless and in fact show no symptoms at all. Although these can grow somewhat quickly, they are often thought to be benign because they cause no symptoms. About one in three patients will develop a lump in the neck that represents spread from the tumor, and occasionally this is the only noticeable sign of disease. Less than ten percent of patients will have cancer spread to other parts of the body when they are first diagnosed.

Diagnosis
Diagnosis is usually made with a biopsy of the suspicious nodule or of a swollen lymph node. This determines what type of tumor is present and helps guide what type of treatment is most appropriate. Additional workup often involved imaging of the head and neck with a CT or MRI scan, and sometimes other imaging is helpful. A treatment team is assembled, usually consisting of a dermatologist, surgeon, radiation oncologist, and sometimes a medical oncologist.

Treatment
Surgery is usually a part of the treatment for MCC, as is radiation. For most patients, surgical removal of the tumor will be recommended. Surgeons will usually cut out a small cuff of normal
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tissue around the tumor to catch any microscopic spread. Any lymph nodes with cancer spread will usually be removed. Even if there are no lymph nodes with obvious spread, surgeons will usually sample one or more lymph nodes in the neck during surgery to help determine if cancer has any microscopic spread. The area where the tumor was taken from sometimes needs reconstruction with a skin graft or movement of surrounding skin. After surgery, many patients will benefit from radiation, which starts about one month after surgery and usually lasts for about six weeks. Chemotherapy or other types of medical therapies are not a typical part of treatment unless cancer has spread to other parts of the body or in other special circumstances.

Surveillance and Prognosis
After surgery, it is important to continue to see your treatment team, especially your dermatologist, to monitor for recurrent cancer and manage any treatment side-effects. Unfortunately, MCC is often aggressive, especially in patients with spread to lymph nodes or other parts of the body. Specific information on prognosis is very individualized and should be discussed with your treatment team.

Recommendations
Patients with risk factors for skin cancers, including significant sun exposure or history of sunburns, a history of skin cancers, or a weak immune system, should have their skin checked regularly by a dermatologist or primary care physician. Any abnormal looking spot on the skin, especially rapidly growing, should be seen quickly. MCC is rare and can be challenging to treat, so it is important to work with your dermatologist or primary doctor to develop a treatment team that can help you make the best decisions about your health.

Figure 2: Merkel Cell carcinoma on the mid-portion of a patient’s scalp. This is a round, nodular shape, and this growth is covered in crusting. The second image is after surgical removal and reconstruction with a skin graft. In this procedure the patient had lymph nodes sampled from the neck as well, in a procedure called sentinel lymph node biopsy. Photos courtesy of Kevin Emerick, MD