MERKEL CELL CARCINOMA

http://www.aocd.org

Merkel cell carcinoma (MCC) is a highly aggressive and rare skin cancer. MCC most commonly occurs in elderly Caucasians, and it is slightly more prevalent in men. The risk factors for MCC are similar to that of other skin cancers, specifically older age, fair skin, ultraviolet light exposure, male gender, and immunosuppression. People who are immunosuppressed, particularly those with HIV, chronic lymphocytic leukemia (CLL), and organ transplants, have an increased chance in developing MCC.

This skin cancer presents as a 0.5-5cm pink-red to violaceous-blue, firm, dome-shaped, bump. This bump rapidly grows typically under 3 months. The bump is generally asymptomatic not causing any pain or itching. Most tumors appear on sun-exposed skin with the head and neck region being the most common followed by the lower limbs and upper extremities. Like melanoma, MCC can also occur on sun-protected areas such as the buttock, vulva, abdomen, and thighs. This cancer has the ability to spread from the skin to other parts of the body especially to regional lymph nodes.

The mnemonic AEIOU was created to highlight the clinical features of Merkel cell carcinoma. If a patient case has three or more of these features, MCC should be considered and the tumor ought to be biopsied.

AEIOU:
Asymptomatic/lack of pain
Expanding rapidly (≤ 3 months)
Immunosuppression (e.g. HIV, CLL, organ transplant recipient)
Older than age 50
Ultraviolet light exposed skin

Wide surgical removal with negative borders is the treatment of choice for local disease. However due to the aggressive nature of MCC, the testing of regional lymph nodes for cancer (sentinel lymph node mapping), adjuvant chemotherapy and radiation are recommended.

Unfortunately despite aggressive treatment, high risk of recurrence, metastasis, and death are common. For a bump that is less than 2 cm the relative survival is 66% at 5 years, for a bump that is greater than 2 cm relative survival is 51% at 5 years. When the cancer spreads to local lymph nodes the relative survival of the patient is 39% at 5 years and when cancer spreads beyond the lymph nodes to other parts of the body the relative survival is 18% at 5 years. Majority of MCC’s recur within 3 years of diagnosis, so a skin and a lymph node examination is recommended, at least every 3 months for the first year, then every 6 months for the second year, and annually thereafter.