An unusual presentation of Merkel cell carcinoma
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Introduction
Merkel Cell Carcinoma (MCC) is a rare, neural-crest derived, highly aggressive cutaneous neuroendocrine carcinoma. We describe a case of MCC leading to rapid clinical deterioration and death in an elderly patient.

The initial clinical presentation of fulminating hepatic failure and the pathologic findings of marked bone marrow involvement without an identifiable primary site make this case a highly unusual presentation for MCC.

Case Presentation
87 year old male presented with acute worsening of chronic low back pain, subjective lower extremity weakness, and dyspnea on exertion for the past several weeks.

He had a known history of prostate cancer, treated only with the biologic agent bicalutamide for relief of lower urinary tract symptoms and no history of bony metastasis.

Exam was notable for a 10-lb. weight loss in the preceding 2 months, tender hepatosplenomegaly, and no focal neurologic deficits.

Labs were significant for new mild anemia (Hgb 12.3g/dL), thrombocytopenia (platelets 96K/uL), elevated liver transaminases (AST 702U/L and ALT 264U/L), and marked hypoalbuminemia (<0.6g/dL).

A right upper quadrant ultrasound showed abnormal hepatic echotexture, concerning for an infiltrative disorder. Infectious workup was negative.

Over the next several days, the patient’s anemia and thrombocytopenia worsened (Hgb 8.7g/dL, platelets 58K/uL), and peripheral blood smears showed leukoerythroblastosis, suggestive of a myelophthisic process in the bone marrow.

A bone marrow biopsy demonstrated marrow involvement by a metastatic carcinoma.

The patient’s liver and kidney function rapidly declined (AST 2521U/L and ALT 686U/L), and he began to bleed from his bone marrow biopsy site and upper GI tract.

Intravenous steroids were given for symptom palliation, and due to his rapid clinical deterioration and previously stated wishes, his family transitioned him to palliative measures only. He died six days after admission.

Recent studies implicate a novel oncogenic polyoma virus, present in 80% of MCC cases¹.

MCC often presents as a solitary, firm, colored, nontender papule, nodule, or tumor, usually on a sun-exposed location.

It demonstrates rapid growth over several months, and is locally aggressive, spreading to regional lymph nodes in >50% of patients.

Prognosis depends on stage of disease at diagnosis, but it has generally high mortality rates²:

Sentinel lymph node biopsy (SLNB) is a sensitive test for detecting spread to lymph nodes, and is recommended due to high rates of regional metastasis.

Treatment almost always involves excision at the primary site and adjuvant radiation, both at the tumor site and at the draining lymph node bed.

Chemotherapy (most commonly etoposide + carboplatin) can be useful for palliation, but is otherwise not used as adjuvant therapy due to high morbidity, mortality and generally poorer outcomes.

Recurrence rates are high, and most (90%) happen within 2 years of diagnosis. Sites of recurrence include skin (28%), lymph nodes (27%), liver (13%), lung (10%), bone (10%), brain (6%), bone marrow (2%), pleura (2%), and other.

MCC is considered incurable once disease has spread to viscera.

References