

## In transit metastases of a leg merkel cell carcinoma

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### Clinical Image



Figure 1: In transit metastases of a leg Merkel Cell Carcinoma in a 73-year-old patient with a history of Hypertension and Chronic Lymphocytic Leukemia

### Description

Merkel cell carcinoma (MCC) is a rare and highly aggressive skin cancer. Merkel cells are neuroendocrine mechanoreceptor responsible for transducing mechanical stimuli from the skin. Recently was found they are derived from pluripotent epidermal stem cells [1]. MCC is mainly a carcinoma of the elderly and immunosuppressed patients. Its incidence varies from 0.79 per 100,000 in the USA to 1.6 per 100,000 in Australia [2]. Two aetiologies are currently discussed for its tumourigenesis: MCC polyomavirus (80%) and UV-induced mutations [3]. Viral oncogenesis is mediated by the large and small T antigens of MCC polyomavirus which promote cell proliferation and survival. MCC caused by chronic UV exposure displays a distinct mutation profile and chronic UV exposure may be responsible for the immunosuppression, helping MCC to develop. Patients with CLL have a 34 to 48-fold increased risk of having a MCC [4].

MCC typically develops rapidly over weeks to months on a chronically sun-damaged skin as an uncharacteristic nodule. Our patient reported a history of a rapidly growing nodule over 6 months on her left leg with sudden onset of in-transit cutaneous metastasis along the lymphatic drainage (figure 1). The diagnosis is therefore made by histopathology - there are three histologic subtypes, though clinically

insignificant. In the immunohistochemistry panel, cytokeratin 20 is a fairly specific and sensitive marker for MCC, with a characteristic paranuclear dotlike positivity. But epithelial markers (such as AE1/AE3, CAM 5.2, pan-cytokeratin, epithelial membrane antigen, and Ber-EP4) and a wide range of neuroendocrine markers (neurofilaments, neuron-specific enolase, chromogranin, synaptophysin, bombesin, somatostatin, vasoactive intestinal peptide, and proconvertases) are usually positive [5].

Although chemo-responsive and radiosensitive in advanced disease the prognosis is poor. Disease-associated mortality is estimated to be between 33% and 46% [2]. Recent investigations with anti PD-L1/PD-1 antibodies have shown promising results [6].

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