An 80 year-old female who had been diagnosed with CLL in 1994 presented to our Hematology Clinic in November 2002 with a rapidly growing nodular lesion on her left thigh. In 1994, she was diagnosed with CLL stage 0 and was observed for the next four years without treatment. In November 1999, owing to disease progression in the form of bulky disease, she was treated with six cycles of Fludarabine. After the last cycle of Fludarabine, she developed immune hemolytic anemia which was treated with steroids. In August 2001 she again had symptomatic organomegaly in addition to persistent hemolytic anemia. This prompted treatment with Rituximab, Cytoxan and Decadron. After the first cycle her hemolytic anemia resolved and steroids were discontinued. Additional doses of Rituximab q weekly x 4 in March and November 2002 resulted in significant reduction in her organomegaly.

In November 2002, she complained of a red indurated nodular lesion on her left thigh. Over the next 4-6 weeks it grew rapidly into a 5 x 6 centimeter firm, red, nodular, non tender intracutaneous mass. During this time she also developed an 8 x 6 centimeter left inguinal lymph node. Her white cell count was 8.2x10^9/L with 5x10^9/L lymphocytes, her hematocrit and platelets were normal. Her CD4 count was low at 0.6x10^9/L and CD 8 count 0.65x10^9/L and ratio of 0.94. Chemistries revealed an elevated LDH and a high uric acid. Core biopsy of the skin lesion and the left inguinal node showed intermediate size cells with bluish nuclei, small nucleoli and scant cytoplasm. There was extensive epidermal invasion and a high mitotic rate. Immuno histochemical stains were negative for LCA, S-100 and positive for chronogranin A, synaptophysin, neurofilament, AE-1/3 and CK-20 that confirmed the diagnosis of MCC. She was treated with Carboplatin and Etoposide with minimal improvement and died 8 weeks after the diagnosis.

MCC is a rare cutaneous neoplasm of the elderly. It is also called Neuroendocrine cancer of the skin or small cell cancer of the skin. Immunosuppresion of various causes may play a role in the pathogenesis of MCC. There is a recent report of development and rapid dissemination of MCC following therapy with Fludarabine and Rituximab for relapsing follicular lymphoma. To the best our knowledge this is the first case report of MCC in a patient with CLL treated with Fludarabine and Rituximab.