Multiple Chloromas in a Patient with secondary Acute Myeloid Leukemia: an interdisciplinary approach

Weiglein T, Rexrodt P, Rieger L, Vehling-Kaiser U.
The Landshut Network for Oncology and Palliative Care

Abstract:
We present the case of a 75-year old female patient diagnosed with secondary acute myeloid leukemia (sAML) in October 2011. Over the course of the disease, the patient developed three successive Chloromas at rare sites (epidural, orbital, vaginal), which were all treated with radiation therapy achieving excellent results. The patient survived over two years with radiation and systemic therapy with a good quality of life and only mild side effects. We conclude that the role of radiation therapy in the management of Chloroma is important, especially in a palliative therapy; low doses of radiation up to 30 Gy have excellent results in disease control and symptom relief.

Introduction:
Chloromas, also known as myeloid sarcoma or granulocytic sarcoma, are a rare extramedullary manifestation of acute myeloid leukemia (AML) and are reported in up to 9.0% of all patients with AML. It is a tumoral lesion consisting of immature cells of granulocytic series. They are often associated with a poor prognosis. The most common locations of Chloroma are skin, soft tissue, bone, periosteum and lymph nodes. Intensive Cytarabine-containing remission-induction chemotherapies have been the most applied therapeutic strategies, but many old or multimorbid patients do not qualify for intensive treatments. The role of radiotherapy in the treatment of extra-medullary AML is yet not fully established, but the literature and our results show excellent local disease control and palliation of symptoms without significant toxicity [1-4].

Case presentation:
A 75-year old female patient diagnosed with essential thrombocythaemia in 2009 presented herself at our practice in October 2011 with B Symptoms and excess of blasts in the peripheral blood. The bone marrow investigations showed a 50% infiltration of sAML blasts with unfavorable cytogenetics and molecular markers. Due to high age and low performance score we began a palliative therapy with Azacytidine from October 2011 till March 2012. The patient achieved a partial remission. In September 2012, the patient had an extramedullary progression with intraspinal Chloroma at the thoracic spine with beginning myeloid compression. After Radiotherapy with 30Gy and re-induction with Azacytidine, the patient had a complete remission of the Chloroma, and again achieved remission of AML blasts. Azacytidine was continued till March 2013 when the patient had rapid infiltration of AML blasts in both orbital cavities and severe exophthalmos with impairment of seeing. Radiotherapy was immediately applied to both sites with 24 Gray, leading to rapid shrinkage of tumor-size and improvement of local symptoms. The systemic therapy was switched to oral purinethol without effect on blast counts and was switched to subcutaneous Cytarabine in April 2013. At this time point a third Chloroma was histologically diagnosed within the skin of the genital area. A third course of RT was begun showing local remission, but stopped due to systemic disease progression and deteriorating performance status. The patient died in July at home.

Conclusion:
1. Our patient demonstrates a rare case of multiple Chloroma at atypical sites of involvement, showing high sensitivity of extramedullary manifestations when irradiated at lower doses
2. Especially for older and comorbid Patients not suited for intensive treatment regimens, radiotherapy is a fast and effective treatment option with tolerable side effects.
3. Palliative treatment should not be withheld for these patients, even if presenting with adverse prognostic factors such as chloroma or comorbidities
4. Low doses of radiation have excellent results in disease control and symptom relief
5. The role of radiation therapy in the management of Chloroma is important and many authors in the literature support this method