Aggressive Extramedullary Monocytic AML (AML-M5) With Leukemia Cutis in a Postpartum Woman: Challenges in Diagnosis and Management

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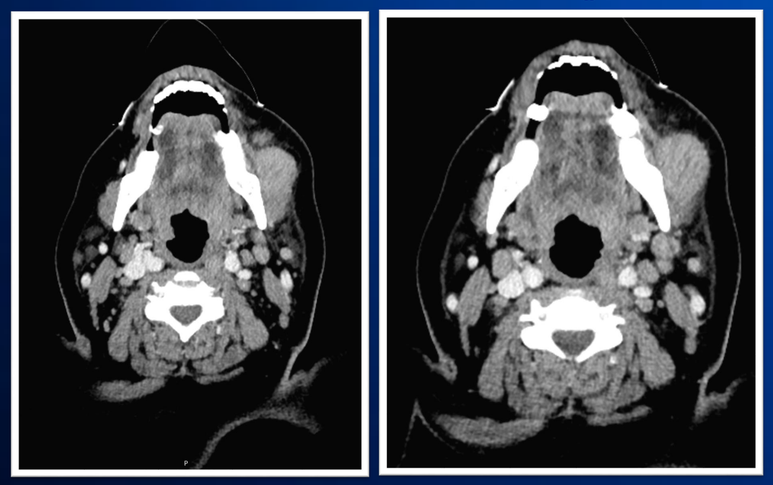
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Introduction

Extramedullary leukemia, also referred to as myeloid sarcoma, is an uncommon manifestation of acute myelogenous leukemia (AML) that frequently occurs alongside bone marrow involvement. [1] This condition is characterized by the infiltration of leukemic cells into extramedullary tissues, such as the skin, soft tissues, and lymph nodes, presenting as masses or nodules. In this case, we report a complex form of AML with monocytic differentiation, presenting with both myeloid sarcoma and leukemia cutis. This case underscores the diagnostic challenges of atypical presentations of AML.

Case Presentation

A 36-year-old postpartum female, 6 months after delivery, presented with a 3-month history of progressively enlarging nodules on the left side of her face, initially misattributed to a dental infection following the extraction of two teeth. Despite dental intervention, the nodules persisted and expanded, accompanied by worsening fatigue, shortness of breath, jaundice, and easy bruising in the week leading Up to her hospital admission. Given the persistent and multifocal nature of the facial masses, along with







Images showing Leukemia Cutes on back and face, lesions caused by infiltration of skin cells by leukemia cells

systemic symptoms, the initial differential diagnosis included chronic infections such as osteomyelitis, granulomatous diseases like sarcoidosis, and neoplastic conditions like lymphoma or metastatic carcinoma.

Upon admission, physical examination revealed hard, non-tender nodules on the left cheek, significant left cervical lymphadenopathy, scleral icterus, and widespread bruising. Laboratory workup showed pancytopenia, with significant anemia, thrombocytopenia, and neutropenia. Elevated liver enzymes and bilirubin suggested hepatic involvement, raising the possibility of systemic disease. Imaging studies, including CT and MRI of the head and neck, revealed multifocal masses in the facial soft tissues, tonsils, and extensive cervical lymphadenopathy, raising strong suspicion for a neoplastic process. [Figures 1,2] Given the rapid progression and severity of her symptoms, a hematologic malignancy became the primary concern. A core biopsy of the cheek mass was performed, revealing diffuse infiltration of atypical myeloid cells, consistent with extramedullary AML with monocytic differentiation. This was corroborated by a bone marrow biopsy, which showed a hypercellular marrow with a high blast count, confirming the diagnosis of AML. Chromosomal analysis revealed a t(10;11) translocation, consistent with an aggressive form of AML. The patient started on induction chemotherapy with cytarabine and idarubicin.

Despite initial treatment, the patient developed new cutaneous lesions in April 2024, presenting as nodules and plaques across the skin. Biopsy of these lesions confirmed leukemia cutis, indicating persistent extramedullary disease.

Discussion

This case highlights the atypical presentation of extramedullary acute AML with monocytic differentiation in a young postpartum female. The patient's presentation, including facial nodules initially misdiagnosed as a dental infection, delayed the correct diagnosis. Typically, AML primarily affects the bone marrow and blood, but in this case, the disease presented with prominent facial masses, initially misdiagnosed as a dental infection, and later progressed to involve the skin as leukemia cutis.

The presence of pancytopenia, jaundice, and systemic symptoms eventually prompted a more thorough evaluation, including imaging and biopsies, which confirmed the diagnosis of AML with extramedullary involvement.

Leukemia cutis occurs in 10-15% of patients with AML. [2] Its presence is usually associated with poor prognosis. Myeloid sarcoma occurs in 2.5-9.1% of patients with AML. The most common sites of involvement are lymph nodes (22.7%), soft tissue (14.2%), and the spinal canal (10.6%). [3]

The importance of considering extramedullary leukemia in patients with unusual masses especially accompanied by systemic signs is highlighted in this case. Comprehensive diagnostic workup was critical in identifying the underlying AML with monocytic differentiation. The subsequent development of leukemia cutis after chemotherapy further illustrates the aggressive nature of the disease and the need for continuous monitoring for relapse or refractory disease, which may require more intensive treatment approaches.

References

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