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Bilateral orbital myeloid sarcoma preceding acute myeloid leukemia in an adult: Case report and review of the literature

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Introduction: Acute myeloid Leukemia is typically a disease of the older population presenting mostly in the fifth decade of life. Myeloid sarcoma is rare presentation of acute myeloid leukemia previously well reported in children and younger population. We present an unusual case of retro-orbital myeloid sarcoma as an initial presentation of acute myeloid leukemia in a 43 year old Caucasian male.

Case Report: In the present case, a 43-year-old male with prior unsuccessful retro-orbital masses biopsies presented with pancytopenia, further studies revealed the diagnosis of AML. The presence of retro orbital masses preceding AML is a rare, to our knowledge since 1993, a total of 11 cases of granulocytic sarcoma preceding AML in adults have been reported, most of them presenting with the cytogenic (8:21) feature with fair prognosis, in the present case we describe a very aggressive case of myelomonocytic leukemia positive for CD34 and CD117 and rearrangement of chromosome 11q23 involving MLL gene with fatal outcome. Differential diagnosis in adults who present with similar symptoms is broad and require a high index of suspicion; in a recent review performed by Priego 2012, differential diagnosis should include inflammatory/metabolic disease (orbital inflammatory pseudo tumor, thyroid orbitopathy, sarcoidosis) and neoplasm (lacrimal tumors, lymphoma and metastasis); however, the clinical behavior and response to therapy seem not influenced by age, sex, anatomic site, de novo presentation or clinical history related to AML, MDS or MPN, histotype, phenotype or cytogenetic findings.

Conclusion: Granulocytic sarcoma or myeloid sarcoma is an uncommon malignant neoplasm associated with myeloid leukemia, the differential diagnosis in this age is broad and diagnosis is a challenge, therefore a multidisciplinary approach, an appropriate clinical exam and history accompanied by a high index of suspicion are needed for proper diagnosis and treatments to avoid fatal outcomes.

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