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TENTH ANNUAL MEETING

Association Between Myeloid Malignancies with MECOM Rearrangement and Ocular Adnexal Leukemic Infiltration

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Abstract

Context: Leukemic infiltration of the eye and ocular adnexal site is rare with unknown risk factors and clinical outcomes.

Objective: To describe the clinical characteristics of extramedullary ocular adnexal (OA) leukemia in patients with myeloid malignancies.

Design: In a retrospective analysis, we screened all patients treated at our center between 1993 and 2022. We identified 50 patients with myeloid malignancies who presented with vision changes with available magnetic resonance imaging of the orbits.

Results: Among all 50 patients, 38 had their symptoms attributed to an infection, 2 to retinal hemorrhage with thrombocytopenia, 1 to chronic graft-vs-host disease, and 1 to dacrocystocele. The remaining 8 patients (7 AML, 1 MDS) had leukemic infiltration of the eye and OA, diagnosed by imaging (7/8) or biopsy (1/8). The median age was 47 years (18–82), 6 (75%) were females. The optic nerve was most frequently affected in 7/8 patients, followed by infiltration of the cranial nerve (CN) III and intraconal soft tissues in 2/8 patients, each. Infiltration of the retina, lateral rectus muscle, CN IV, CN VI, and extraconal soft tissue was seen in 1/8 patient, each. Spinal fluid evaluation identified no evidence of leukemia in all 8 patients. Interestingly, 4/8 patients (50%) had abnormalities in chromosome 3q26 or *MDS1* and *EVI1* complex locus (MECOM) rearrangements, 2 (25%) had Lysine Methyltransferase 2A (*KMT2A*) rearrangement and the remaining two had trisomy 8 and a diploid karyotype respectively. Patients with *KMT2A* rearrangement also had other sites of extramedullary disease, while orbital adnexal involvement was the only extramedullary leukemia site in those with MECOM rearrangements. Intrathecal chemotherapy and concurrent local radiation were given to 6/8 patients while the other 2 received either intrathecal chemotherapy or radiation. Symptoms improved or resolved following these measures in 4/8 patients. These visual findings were detected during salvage therapy (median 4, range 1-9 lines). Overall survival following appearance of these findings was short with a median of 5.8 months (range: 0.4 –17.5).

Conclusion: Leukemic infiltration of the orbital adnexa in myeloid malignancies is rare and associated with an inordinately high proportion of MECOM rearrangements in addition to adverse outcomes.

Background

- Leukemic infiltration of the eye and ocular adnexal (OA) site is rare with unknown risk factors and clinical outcomes.

Objective

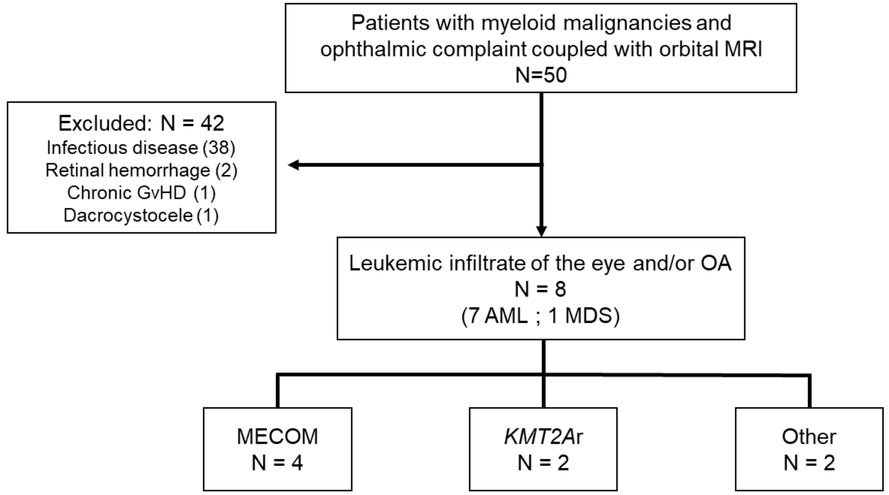
- Describe the clinical characteristics and outcomes of extramedullary OA leukemia in patients with myeloid malignancies.

Design

- Retrospectively screened all patients treated at our center between 1993 and 2022.
- Identified 50 patients with myeloid malignancies who presented with vision changes with available magnetic resonance imaging (MRI) of the orbits.

Results

Patient selection



Patient characteristics

- Median age of patients was 47 years (range: 18 – 82) ; 6 (75%) were females.
- The visual findings were detected during salvage therapy (median 4, range 1-9 lines of therapy).
- Patients with *KMT2A* rearrangement also had other sites of extramedullary disease.
- Patients with MECOM rearrangement had no other sites of extramedullary leukemia.
- Cerebrospinal fluid analysis was negative in all patients.

Affected OA structures

- The optic nerve in 7 patients (Fig A).
- The oculomotor nerve in 2 patients.
- The intraconal soft tissue in 2 patients (Fig B).
- The eye (Fig C), trochlear nerve, abducens nerve, lateral rectus muscle in 1 patient, each.







Treatment and outcomes

- 6 patients received intrathecal chemotherapy and concurrent local radiation.
- 1 patient received only local radiation; 1 patient received only intrathecal chemotherapy.
- Only 4 out of 8 patients experienced symptom improvement.
- The median overall survival following appearance of these findings was 5.8 months

Conclusion

- Leukemic infiltration of the OA in myeloid malignancies is rare and is associated with an inordinately high proportion of MECOM rearrangements.
- Leukemic infiltration of the OA is associated with adverse outcomes

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