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Case Report

Bilateral Orbital Involvement with Severe Visual Impairment and Anterior Staphyloma in a 13-year-old boy with Acute Myeloid Leukaemia: A Case Report

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Abstract

Background: Acute myeloid leukaemia (AML) is a haematological malignancy that can involve extramedullary tissues, including the orbit and ocular structures. Orbital leukemic infiltration is an uncommon but potentially vision-threatening manifestation, particularly in paediatric patients.

Case Presentation: A 13-year-old male with a known history of acute myeloid leukaemia presented with gradually progressive swelling of the eyelids in both eyes over eight months. The patient had previously received four doses of chemotherapy and radiation therapy and was undergoing oral metronomic treatment. Ophthalmological examination revealed severely reduced visual acuity, limited to hand movement perception (HM+) in both eyes. Anterior segment examination documented bilateral localized anterior staphyloma. B-scan ultrasonography revealed vitreous echoes, structural ocular changes, and orbital findings suggestive of leukemic infiltration. Flash visual evoked potential testing was also performed to assess visual pathway function.

Diagnosis: Based on the clinical history, ophthalmological examination, and ultrasonographic findings, bilateral orbital leukemic involvement was clinically assessed in association with localized anterior staphyloma.

Management and Outcome: The patient underwent multidisciplinary ophthalmological evaluation, including recommendations for oculoplastic and corneal consultations. A guarded visual prognosis was explained, and further ocular surgical intervention was considered in relation to the patient's systemic condition. Subsequent treatment response and visual outcome were not documented in the available records.

Conclusion: This case highlights the importance of comprehensive ophthalmic evaluation in children with acute myeloid leukemia presenting with persistent eyelid swelling and severe visual impairment. Early recognition of suspected orbital leukemic infiltration, appropriate ocular imaging, and coordinated multidisciplinary management are essential for clinical assessment and treatment planning.

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INTRODUCTION

Acute myeloid leukemia (AML) is an aggressive hematological malignancy characterized by the abnormal proliferation of immature myeloid cells in the bone marrow. While AML predominantly affects the hematopoietic system, it can also involve various ocular structures either by direct leukemic infiltration or secondary hematological abnormalities. [1]

Ocular manifestations of AML include orbit, eyelids, conjunctiva, retina, choroid and optic nerve.[1] Orbital infiltration, sometimes as myeloid sarcoma (chloroma), is a rare extramedullary manifestation that may lead to eyelid swelling, proptosis, restricted ocular movements and visual impairment. These presentations have also been described in the pediatric population. [2,3]

Anterior staphyloma is a rare and serious ocular condition in which uveal tissue protrudes through a weakened or defective corneoscleral wall. It may develop after corneal trauma, infection or ulceration and can cause significant structural damage and visual impairment. [4]

Case presentation

A 13-year-old male patient, a known case of acute myeloid leukaemia (AML) undergoing treatment for the past year, presented with complaints of swelling in both eyes and gradually progressive diminution of vision bilaterally for approximately 10 months. The patient was receiving oral metronomic therapy and had previously undergone four doses of chemotherapy along with radiation therapy as part of his treatment for AML.

Ocular Examination

Visual Acuity

EYE	**Distance Vision**
OD	HM+
OS	HM+
EYE	**AR Value**
OD	No Flash
OS	No Flash

Refraction	**DISTANCE**			
EYE	Dsph	Dcyl	Axis	BCVA
OD	-	-	-	HM+
OS	-	-	-	HM+

External Examination

Examination	**OD**	**OS**
Lid	Sutured lid tear	Normal
Conjunctiva	Normal	Normal
Cornea	Anterior staphyloma, Cornea bulge out	Anterior staphyloma, Cornea bulge out
Anterior Chamber	Not documented	Not documented
Iris	Not documented	Not documented
Pupil	Not documented	Not documented
Lens	Not documented	Not documented
Ocular Movement	Not documented	Not documented



Figure 1: Clinical photograph of both eyes (OU) showing bilateral anterior staphyloma, whitish corneal opacification, with prominent corneal involvement in both eyes.
Image Courtesy: Image captured by the author

Ophthalmic Examination

On ophthalmic examination, visual acuity was reduced to hand movement perception (HM+) in both eyes. External examination revealed a previously sutured eyelid tear in the right eye (OD), while the left eyelid (OS) was normal. The conjunctiva appeared normal bilaterally. Corneal examination demonstrated localized anterior staphyloma in both eyes. Findings related to the anterior chamber, iris, pupil, lens, and ocular movements were not documented in the available clinical records.

Posterior Segment Examination

Posterior segment evaluation indicated that the retina was attached in both eyes, and the optic nerve heads appeared grossly normal. B-scan ultrasonography revealed findings suggestive of bilateral orbital infiltration associated with the patient's underlying acute myeloid leukemia.

Clinical Diagnosis

Based on the patient's medical history, ophthalmological examination, and ultrasonographic findings, the following clinical diagnoses were recorded:

1. Bilateral localized anterior staphyloma.
2. Bilateral orbital leukemic infiltration.
3. Known case of acute myeloid leukemia (AML).

Management

- Multidisciplinary management in coordination with the haematology-oncology team for underlying AML.
- Comprehensive ophthalmic evaluation, including B-scan ultrasonography and VEP, to assess ocular and visual pathway involvement.
- A corneal consultation was also recommended for further assessment of bilateral localized anterior staphyloma and to determine the need for possible surgical intervention.
- The patient's family was counselled regarding the guarded visual prognosis. Further ocular surgical management was to be considered based on the patient's overall systemic condition and underlying acute myeloid leukemia (AML).

DISCUSSION

Acute myeloid leukemia (AML) is a haematological malignancy that can affect both ocular and orbital structures

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through direct leukemic infiltration or secondary hematological complications. Although it is uncommon, in certain cases it may present with eyelid swelling, proptosis and progressive visual impairment, particularly in pediatric patients.

In the following case, a 13-year-old boy with a known history of AML is presented with progressive bilateral eyelid swelling and severe diminution of vision over approximately 8–10 months. Examination revealed hand-movement vision in both eyes, accompanied by bilateral localized anterior staphyloma. These findings indicate significant bilateral ocular involvement and structural abnormalities.

B-scan ultrasonography revealed vitreous echoes, anterior staphylomatous changes and orbital findings suggestive of leukemic infiltration. The widening of the superior orbital shadow with internal echoes raised suspicion of bilateral orbital involvement. These findings highlight the importance of ultrasonography in evaluating orbital and posterior segment structures when direct clinical assessment is limited by severe visual impairment and anterior segment abnormalities.

An interesting finding was the Flash VEP response, which demonstrated identifiable P2 components in both eyes, with recorded latencies of 92.8 ms in the right eye and 100.3 ms in the left eye. Despite the severely reduced visual acuity, these recordable responses suggest preserved electrophysiological responsiveness to flash stimulation. However, the presence of VEP responses does not necessarily indicate normal visual function or guarantee visual recovery.

The coexistence of bilateral orbital findings suggestive of leukemic infiltration and bilateral anterior staphyloma in a young patient with AML makes this case clinically noteworthy. It emphasizes the importance of comprehensive ophthalmological assessment and a multidisciplinary approach involving ophthalmologists, oculoplastic specialists, corneal specialists and the treatment hematology team.

CONCLUSION

This case highlights the importance of recognizing ocular and orbital manifestations of acute myeloid leukemia in pediatric patients. Bilateral eyelid swelling, severe visual impairment and anterior staphyloma warrant detailed ophthalmological evaluation. B-scan ultrasonography and Flash VEP provide useful additional information regarding ocular structures and visual pathway responses. Early identification, appropriate investigations and multidisciplinary management are essential for clinical assessment and treatment planning. Given the severity of visual impairment and underlying systemic malignancy, the visual prognosis remains guarded.

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