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Thalassemia and Bilateral Acute Anterior Uveitis

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Abstract

Introduction: *Thalassemias are a group of genetic disorders characterized by a reduced synthesis of alpha or beta chains of hemoglobin (Hb). Hemoglobin, the oxygen-carrying component of red blood cells, consists of two proteins: alpha and beta. When the body does not produce enough of either protein, red blood cells form incorrectly and cannot carry sufficient oxygen, leading to anemia that starts in early childhood and persists throughout life.*

Materials and Methods: *A 13-year-old female was admitted with complaints of low vision, pain, redness (OU VA 20/400, BCVA 20/200, IOP OU -13 mmHg with GT) to the Cornea-Uveitis department at the S V Malayan Eye Center. The patient was referred by a pediatrician /hematologist. Two months ago, she was diagnosed Hb S beta thalassemia, which was accompanied by sudden vision loss. Two -month treatment. topical steroids eye drops q. i. d., cycloplegic eye drops t.i.d., no positive dynamics were observed. At the ocular examination, her BCVA 20/200 in both eyes. The patient was complaining of blurred vision in both eyes. On examination, she had granulomatous KPs on the lower surface of the cornea, 3+ cells in the anterior chamber, 2+ cells in the vitreous, optic disk hyperemia and swelling (obscuration of the nasal border of the disc.). All possible laboratory tests (ACE, RF, HLA -B27, HLA-B51, ANA, ANCA, Syphilis serology, HSV, VZV, CMV, Radiologic testing) were performed to diagnose uveitis, none of them were positive. Visual field (formal/ typically automated) testing and optical coherence tomography were performed and Stage-1 Early Papilledema was diagnosed. At the same time, under the supervision of a hematologist, prescribed topical steroids eye drops., cycloplegic eye drops every 2 hours, with multiple subtenon injections of Diprospan 1.0 (Betamethasone). Positive dynamics were observed after two weeks, and an excellent result was recorded after one month: uveitis remission, the patient BCVA was 20/20.Papilledema remained relatively stable (slight positive dynamics were observed in the layer of peripapillary nerve fibers).*

Discussion: *Our case report demonstrates a rare case of presumed Thalassemia associated bilateral acute anterior uveitis associated with papilledema in a 13 -year-old girl.The presence of papilledema was due to Thalassemia disease, and the presence of uveitis remains unexplained or can be not related .Considering our results, we aim to keep this case under dynamic control and report on further development, as conservative treatment (intravitreal injections, topical steroids, and cycloplegic drops) resulted in uveitis remission, but papilledema remained stable.*

Keywords: Uveitis, Papilledema, Thalassemia, Retina, Galucoma

Introduction

Thalassemias are genetic blood disorders characterized by defective hemoglobin synthesis, leading to chronic anemia. Hemoglobin consists of two proteins, alpha and beta, and an imbalance in their production results in structurally abnormal red blood cells with reduced oxygen-carrying capacity.

Optic nerve swelling, or papilledema, is known to occur as a reaction to intraocular inflammation. It has been associated with multiple types of uveitis, including anterior uveitis, intermediate uveitis, and panuveitis. However, inflammatory optic nerve changes in uveitis patients remain poorly documented. This case report presents a 13-year-old girl diagnosed with Hb S beta thalassemia and bilateral acute anterior uveitis with associated papilledema.

Materials and Methods

A 13-year-old female presented with low vision, pain, and redness in both eyes. Clinical findings at presentation included:

- **Visual Acuity:** 20/400 (OU), Best Corrected Visual Acuity (BCVA) 20/200 (OU)
- **Intraocular Pressure (IOP):** 13 mmHg (OU)
- **Slit-Lamp Examination:** Granulomatous keratic precipitates, 3+ cells in the anterior chamber, and 2+ cells in the vitreous
- **Fundoscopic Examination:** Optic disc hyperemia and swelling (early-stage papilledema)

Initial treatment with topical steroids (four times daily) and cycloplegic eye drops (three times daily) failed to produce improvement. Additional systemic and infectious workups (ACE, RF, HLA-B27, HLA-B51, ANA, ANCA, syphilis serology, HSV, VZV, CMV, radiologic imaging) were all negative.

Following consultation with a hematologist, the patient was started on an intensified regimen:

- **Topical steroids:** Increased frequency
- **Cycloplegic eye drops:** Every 2 hours
- **Subtenon injections:** Diprospan 1.0 mg (Betamethasone)

After two weeks, visual function improved significantly, and by one month, BCVA recovered to 20/20. Uveitis fully resolved, but papilledema persisted with slight improvement.

Results and Discussion

Table 1: The clinical presentation and response to treatment are summarized

Parameter	Initial Presentation	Post-Treatment Outcome
Best Corrected Visual Acuity (BCVA)	20/400 (OU) → 20/200 (OU)	20/20 (OU)
Intraocular Pressure (IOP)	13 mmHg	Stable
Anterior Chamber Cells	3+ cells	Resolved
Vitreous Cells	2+ cells	Resolved
Optic Disc Findings	Hyperemia, swelling	Papilledema stable

This case underscores the need for close monitoring of patients with thalassemia who develop ocular manifestations. While papilledema can be linked to the underlying hematologic disorder, the etiology of uveitis remains unexplained. Possible mechanisms could involve immune dysregulation or secondary inflammatory responses.

Conclusion

This report presents a rare instance of bilateral acute anterior uveitis with papilledema in a patient with Hb S beta thalassemia. While the uveitis resolved with aggressive corticosteroid therapy, papilledema persisted, necessitating ongoing follow-up. Further research is needed to understand any potential relationship between thalassemia and ocular inflammation.

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