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Case Report

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An Unusual Cause of Optic Atrophy in Childhood

V Swetha E Jeganathan[1,2], Ala Noaman[3] and Andrew Blaikie[4,5]*

[1]*W.K. Kellogg Eye Center, Michigan Medicine, Ann Arbor, USA*

[2]*Department of Biomedical Engineering, University of Michigan, Ann Arbor, USA*

[3]*University of Edinburgh, Edinburgh, Scotland*

[4]*Department of Ophthalmology, Queen Margaret Hospital, Dunfermline, Scotland*

[5]*Global health Implementation, University of St Andrews School of Medicine, St Andrews, Scotland*

***Corresponding Author:** Andrew Blaikie, Senior Lecturer, Global Health Implementation, University of St Andrews School of Medicine, St Andrews, Scotland.

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Abstract

Purpose: To report a rare and interesting cause of bilateral optic atrophy secondary to hemolytic uremic syndrome (HUS).

Case Report: A 6-year-old asymptomatic child was referred with bilateral pale optic discs. She was otherwise healthy, with no known family history of any eye problems. On detailed questioning, it was revealed that she had HUS secondary to *E. coli* diarrheal infection at the age of 2.5 years, with renal failure that necessitated treatment with dialysis. Retinal nerve fiber analysis confirmed poor reserves with fewer axons in both eyes. The patient was followed up and discharged when there were no signs of progression.

Conclusion: Ocular involvement in HUS although rare, depicts a severe form of HUS, and may develop as sequelae to ischemic or toxic insults. Ophthalmology referral is required especially in preverbal patients with HUS to exclude potential vision-endangering consequences.

Key words: *Optic atrophy; Hemolytic uremic syndrome; Ophthalmology; Children; Dialysis*

Abbreviations: HUS: Hemolytic Uremic Syndrome; OCT: Optical Coherence Tomography;

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Introduction

We present a very unusual cause of optic atrophy that was identified on detailed clinical history.

Case Report

A 6-year-old female patient was referred by the optometrist to our local Ophthalmology department with bilateral pale optic discs. The patient did not report any ophthalmological problems. She denied any headaches, photophobia, malaise or night blindness, and did

not have any problems with contrast sensitivity. Her birth history was unremarkable. On thorough questioning, it was revealed that she had hemolytic uremic syndrome (HUS) secondary to E. coli diarrheal infection at the age of 2.5 years, with renal failure that necessitated a short period of hemodialysis. She was otherwise healthy, and there was no family history of any eye related problems. Her visual acuity was 20/40 in the right eye and 20/20 in the left. She had no refractive errors. There was no afferent pupil defect, and her intraocular pressures were normal. Dilated fundoscopy showed pale, cupped discs in both eyes, which was worse in her right eye [Figure 1a and 1b].

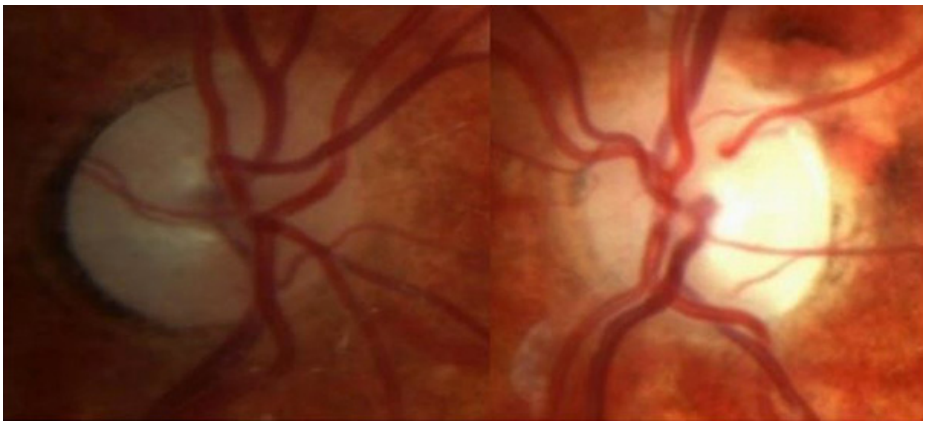

Figure 1a and 1b: *Bilateral optic atrophy (Note: different exposures).*

There was no evidence of retinal ischemia or neovascularization. Her parents and siblings had normal eye examinations. Her optical coherence tomography (OCT) confirmed a flatter papillo-macular bundle [Figure 2] with loss of foveal dip in the right eye [Figure 3]. OCT of her left eye was better in appearance [Figures 2 and 3].

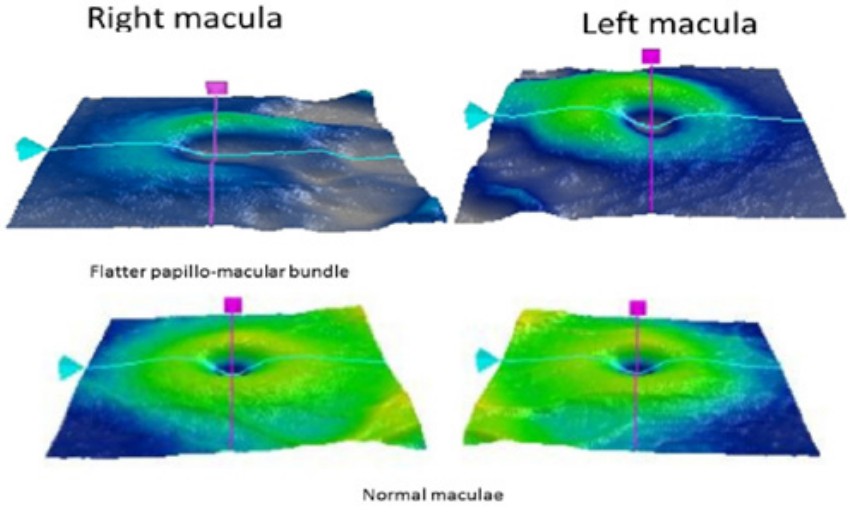

Figure 2: *Macular OCT findings.*

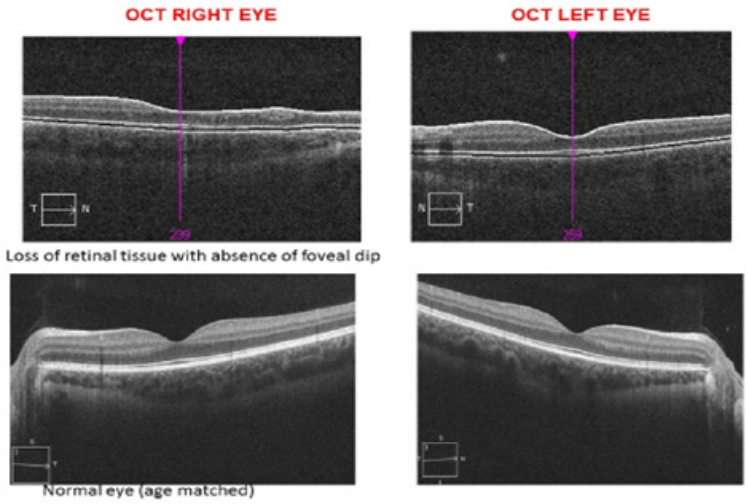

Figure 3: *Macular OCT comparing both eyes, with age-matched normal.*

Retinal nerve fiber analysis supported poor reserves with fewer axons in both eyes [Figure 4]. The patient had serial examination by the same Ophthalmologist (AB). Her visual acuity, optic disc appearance, and OCT findings remained unchanged, and the patient was discharged from the eye clinic.

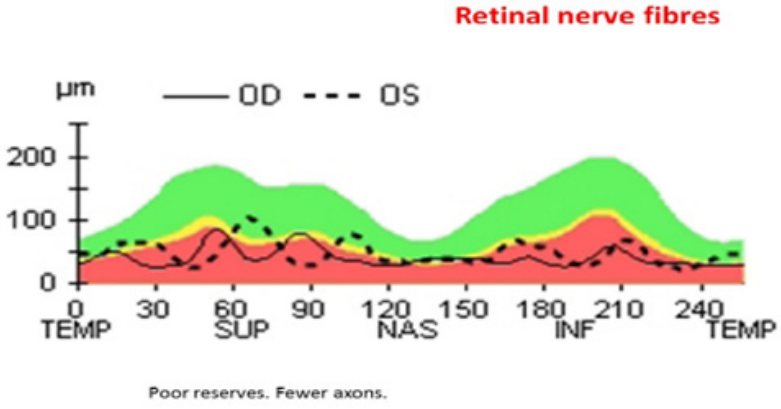

Figure 4: *Retinal nerve fiber examination of both eyes.*

Discussion

Hemolytic-uremic syndrome, first defined in 1955, is a predominantly childhood disease characterized by a triad of microangiopathic hemolytic anemia, consumptive thrombocytopenia, and renal insufficiency. [1] HUS is the most common cause of acquired acute renal failure in childhood, with a 5 to 10% mortality rate. [2] The overall incidence of HUS in the United States is estimated at 2.1 cases per 100,000 persons/year, with a peak age incidence between six months and four years, and its incidence is increasing worldwide. [3] HUS is usually preceded by a prodrome of bloody diarrhea. Several causative agents have been implicated – the most common is Shiga-like toxin-producing *E. coli HUS* (*STEC-HUS*), triggered by *E. coli O157:H7*. [4] Most children with post diarrheal HUS, have a good chance of spontaneous recovery, with hospital observation and hemodialysis where indicated. Acute renal failure occurs in up to 70% of patients with STEC-HUS, although almost 85% recover their renal function. [2]

We report an unusual case of bilateral optic atrophy in pediatric case with HUS following *E. coli* infection. Ocular involvement in HUS although rare, depicts a severe form of HUS. Purtscher retinopathy, subhyaloid hemorrhage, chorioretinitis, ischemic signs and serous

retinal detachment are serious eye conditions that are infrequently present in early stages of disease, and may be missed by the general pediatrician. [5-7] Bilateral optic atrophy is seen only in exceptional cases and may develop as sequelae to ischemic or toxic insults. [8,9] Anemia, hypoproteinemia, and intradialytic hypotension are known predisposing risk factors for anterior ischemic optic neuropathy. [9] However, the elevated blood viscosity and hematocrit during dialysis, may result in reduced blood flow and further increased risk of ischemia. Blindness from uremic optic neuropathy may occur, due to compromised vascular circulation, with potential reversibility with immediate dialysis. [10]

Conclusion

Ophthalmology referral is warranted especially in preverbal patients with HUS to exclude potential sight- threatening ramifications. Ophthalmologists also need to be vigilant in associating the patient's ocular findings (which may be an early presentation of HUS) with serious underlying systemic disease such as vasculitis.

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