American Journal of Ophthalmology Case Reports 17 (2020) 100599

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.elsevier.com/locate/ajoc

Case report

Oral acetazolamide as a medical adjuvant to retinal surgery in optic disc pit maculopathy in a pediatric patient

Carla J. Osigian*, Daniel Gologorsky, Kara M. Cavuoto, Audina Berrocal, Victor Villegas

Bascom Palmer Eye Institute, Department of Ophthalmology, University of Miami Miller School of Medicine, Miami, FL, USA

ARTICLE INFO

Keywords:
Optic disc pit
Optical coherence tomography
Intracranial pressure
Acetazolamide

ABSTRACT

Purpose: To describe a novel combination of surgical and medical management for the treatment of optic disc pit maculopathy.

Observations: A 12-year-old obese girl with bilateral optic disc pits presented with decreased vision in the left eye (20/400). On dilated fundus examination, she was found to have a macula-involving serous retinal detachment. Pars plana vitrectomy, posterior hyaloid peel, internal limiting membrane peel, fluid-air exchange, temporal juxtapapillary endolaser, and C_3F_8 tamponade were performed. Postoperatively, there was persistence of subretinal fluid, so oral acetazolamide and a weight loss regimen were started. After 3 months of medical treatment, the subretinal fluid decreased significantly and visual acuity improved to 20/60.

Conclusion and Importance: Currently, the mechanisms leading to optic disc pit maculopathy remain a topic of debate. As optic disc pits may provide a conduit between the subarachnoid and subretinal spaces, the reduction of intracranial pressure with the use of systemic carbonic anhydrase inhibitors may play a role in decreasing the subretinal fluid associated with select cases of optic disc pit maculopathy.

1. Introduction

Optic disc pits are rare congenital defects considered to be part of the spectrum of congenital cavitary optic disc anomalies. The pit usually presents as a solitary, oval, gray-white depression, most commonly located in the inferotemporal segment of the optic disc. Optic disc pits are typically sporadic and unilateral but may be bilateral in up to 15% of patients.[1] Vision loss is not due to the pit itself but rather to macula-involving serous retinal detachments or cystoid macular edema. The source of the fluid is unclear but is thought to originate from the vitreous or the cerebrospinal fluid.[2] Currently, the management of optic pit maculopathy remains a topic of debate. One-quarter of cases may resolve with conservative management, however the chronicity of retinal fluid results in poor visual outcomes, making surgical intervention the preferred treatment course.[3] We report a case of ODP-M treated with a combination of surgical and medical therapy with favorable anatomic and visual outcomes.

2. Case report

An 86kg 12-year-old girl presented to the ocular emergency room with decreasing vision in her left eye. Her past medical history was significant for polydactyly, flat feet, and profound left-sided deafness. On examination, her vision was 20/20 in the right eye and 20/400 in the left. Intraocular pressures (IOP) were 13 mm Hg in both eyes. Pupils were round and reactive to light. The anterior segment examination was unremarkable. Dilated fundus examination and spectral domain optical coherence tomography (SD-OCT) imaging of the right eye revealed an inferotemporal optic disc pit without subretinal fluid. The left eye showed an inferotemporal optic nerve pit with subretinal fluid tracking from the optic nerve through the macula, as well as a collection of subretinal fluid immediately nasal to the optic nerve (Fig. 1A and B). There was no evidence of papilledema. Magnetic resonance imaging of the brain and orbits was unremarkable. The patient underwent a 25 gauge pars plana vitrectomy, posterior hyaloid peel, and temporal juxtapapillary internal limiting membrane peel. Fluid-air exchange was performed, but was limited due to fluid influx from the optic disc pit. Endolaser was then applied to the temporal edge of the optic nerve head, followed by an air-gas exchange with 18% C_3F_8 tamponade. The first postoperative images showed reduced fluid, however two months postoperatively, SD-OCT images demonstrated significant recurrence of subretinal fluid (Fig. 2). The patient was placed on 500 mg acetazolamide extended-release tablets three times daily, was started on a weight loss regimen, and was referred for evaluation to a pediatrician and an endocrinologist. Subsequent evaluations were unremarkable for genetic or medical conditions. Three

* Corresponding author. MD Bascom Palmer Eye Institute University of Miami Miller School of Medicine, 900 NW 17th Street, Miami, FL, 33136, USA.
E-mail address: cjo39@med.miami.edu (C.J. Osigian).

https://doi.org/10.1016/j.ajoc.2020.100599
Received 16 April 2018; Received in revised form 1 October 2019; Accepted 10 January 2020
Available online 16 January 2020

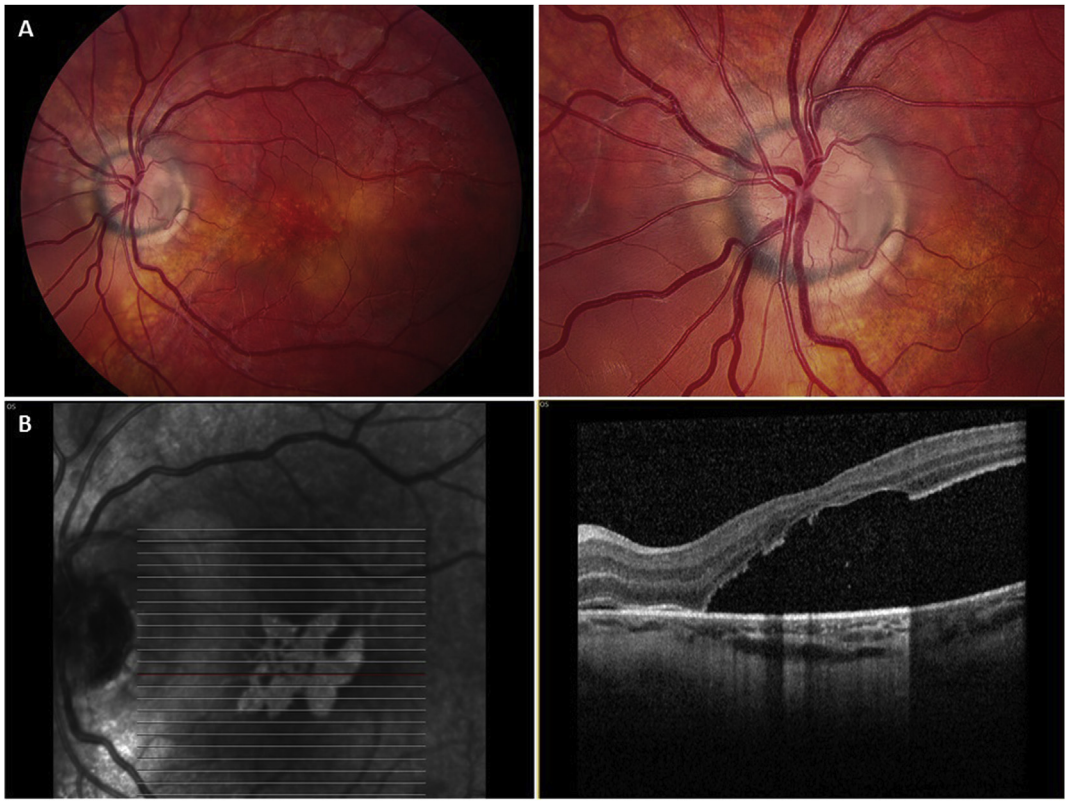

Fig. 1. A) Fundus photographs of the left eye showing an inferotemporal optic nerve pit with subretinal fluid in the macula and nasal to the optic nerve. B) Optical coherence tomography imaging of the left eye demonstrating subretinal fluid tracking from the optic nerve through the macula.

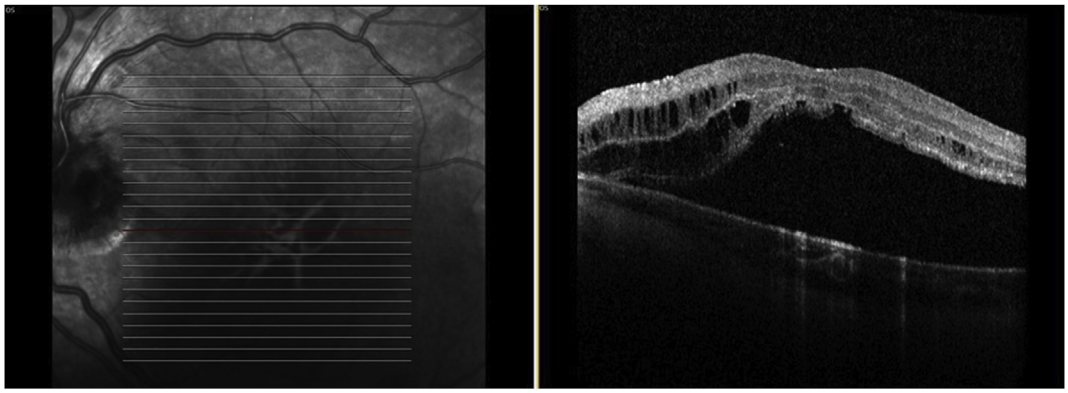

Fig. 2. Optical coherence tomography images two months after surgery showing significant persistence of subretinal fluid.

months later, the patient had a 25 kg decrease in weight and fundus examination as well as SD-OCT images showed a significant decrease in subretinal and intraretinal fluid (Fig. 3A and B), with an improvement in the vision of the left eye to 20/60.

3. Discussion

Optic disc pits are relatively rare with an incidence of about one in 11,000 people.[1] Histologically, they consist of a herniation of dysplastic retina through the lamina cribrosa into the subarachnoid cavity, creating an anomalous communication between the intraocular and extraocular space.[4]

Visual deterioration most commonly occurs when optic disc pits are associated with maculopathy, ranging from 25 to 75% of cases.[5] ODP-M presents either as macular edema, schisis-related macular detachment, or macular pigmentary changes. There is no known trigger for the development of maculopathy. The nature and origin of the intraretinal or subretinal fluid is controversial, but is thought to originate from the cerebrospinal fluid, vitreous, vessels at the base of the pit, or leakage from the choroid.[2]

The mechanism of ODP-M is also unclear. Intraocular factors such as vitreous liquefaction or traction or extraocular factors such as pressure gradients have been thought to play a role in the pathogenesis.[1,2] As vitreous liquefaction and traction were not present in our patient, this leads to the concept of a pressure gradient between the subarachnoid space and the eye as a possible mechanism affecting intra- and sub-retinal fluid accumulation. Cerebrospinal fluid has been proposed to enter the intra- and sub-retinal spaces from the subarachnoid cavity through the herniated optic disc pit defect. Several studies demonstrated evidence of communication between the subarachnoid space and the subretinal space through SD-OCT imaging.[6,7] Other authors have reported intracranial migration of silicone oil from an eye with an optic disc pit after vitrectomy with silicone oil tamponade, supporting this etiology.[8] With an optic pit acting as a communication between the

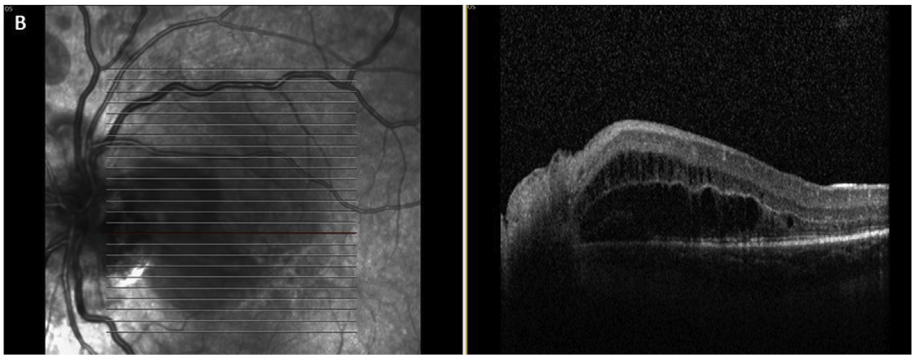

Fig. 3. A) Postoperative fundus photograph of the left eye three months after initiation of acetazolamide and weight loss regimen showing complete resolution of the subretinal fluid in the nasal aspect of the retina, good laser take in the juxtapapillary temporal retina, and some residual microcystic macular edema. B) Optical coherence tomography images three months after initiation of acetazolamide and weight loss regimen, showing significant decrease in subretinal fluid.

eye and the subarachnoid space it is possible that when the ICP rises, fluid is shifted into the eye, dissecting under or within the retina.[9] A lumbar puncture was not performed in our patient prior to treatment. However, since a relationship between increased ICP and obesity has been established,[10] pressure gradients can be considered to be potentially related to retinal fluid accumulation in an obese patient.

As the mechanism of disease is still poorly understood, treatment of optic disc maculopathy remains controversial. The timing, surgical procedure, and long-term outcomes continue to be a topic of debate. There are no guidelines regarding the management of ODP-M in pediatric patients, as the literature is limited to a small number of cases.[1,2] Although PPV is the predominant approach, numerous other techniques and modifications have been described, including laser photocoagulation at the temporal disc margin, intravitreal gas injection, and macular buckling surgery. A recent multicenter retrospective study that evaluated patients with optic disc pit maculopathy that underwent vitrectomy surgery found that the mean time for subretinal fluid resolution was over 12 months in patients with positive outcomes.[11] Therefore, acetazolamide may have a benefit as an adjuvant therapy in order to decrease the time to achieve subretinal fluid resolution as seen in our case. The adjuvant use of acetazolamide in ODP-M has only been previously described in an isolated case report,[12] and was used preoperatively as compared to its use post-operatively in our case. Carbonic anhydrase inhibitors produce acidification of the subretinal space, which induces the resorption of subretinal fluid and improves retinal adhesiveness.[13] Acetazolamide also inhibits the carbonic anhydrase enzyme in the choroid plexus, resulting in decreased cerebrospinal fluid production and pressure.[14] Assuming macular fluid originates from the pressure gradient between the subarachnoid space and the eye, lowering the ICP could decrease the accumulation of macular fluid. Additionally, acetazolamide also decreases IOP, which might affect the pressure gradient at the lamina cribrosa. Overweight individuals undergoing significant weight loss may demonstrate a greater decrease in the ratio of ICP to IOP, similar to the mechanism in idiopathic intracranial hypertension. Given our patient's obesity, we suspect the adjuvant use of acetazolamide in her specific case assisted in a faster decrease of subretinal fluid after surgery.

Although in our patient a notable decrease in postoperative subretinal fluid was noted after initiation of acetazolamide, it remains unclear if this improvement was due to 1) acetazolamide, 2) weight loss, 3) delayed response to surgical intervention, 4) natural course of the disease, or 5) a combination of all of these. We therefore propose the use of acetazolamide as an adjuvant therapy along with previously described surgical interventions.

4. Conclusions

Acetazolamide may be beneficial in some cases of optic disc pit maculopathy. However, studies including a larger population should be considered to demonstrate the efficacy of this drug prior to recommending it as a routine adjuvant to surgical intervention for the treatment of these patients.

Patient consent

The patient provided written consent for publication of personal information including medical record details and photographs.

Funding

No funding or grant support

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare no financial interest or sources of support.

Acknowledgements

None.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ajoc.2020.100599.

References

1. Georgalas I, Ladas I, Georgopoulos G, Petrou P. Optic disc pit: a review. *Graefe's Arch Clin Exp Ophthalmol (Albrecht Von Graefes Arch Klin Exp Ophthalmol)*. 2011;249:1113–1122.
2. Moisseiev E, Moisseiev J, Loewenstein A. Optic disc pit maculopathy: when and how to treat? A review of the pathogenesis and treatment options. *Int J Retin Vitr*. 2015;1:1–9.
3. Sobol WM, Blodi CF, Folk JC, Weingeist TA. Long-term visual outcome in patients with optic nerve pit and serous retinal detachment of the macula. *Ophthalmology*. 1990;97:1539–1542.
4. Ferry AP. Macular detachment associated with congenital pit of the optic nerve head. pathologic findings in two cases simulating malignant melanoma of the choroid. *Arch Ophthalmol*. 1963;70:346–357.
5. Brown GC, Shields JA, Goldberg RE. Congenital pits of the optic nerve head. II. Clinical studies in humans. *Ophthalmology*. 1980;87:51–65.
6. Krivoy D, Gentile R, Liebmann JM, et al. Imaging congenital optic disc pits and associated maculopathy using optical coherence tomography. *Arch Ophthalmol*. 1996;114(2):165–170.
7. Rutledge BK, Puliafito CA, Duker JS, Hee MR, Cox MS. Optical coherence tomography of macular lesions associated with optic nerve head pits. *Ophthalmology*. 1996;103(7):1047–1053.
8. Kuhn F, Kover F, Szabo I, Mester V. Intracranial migration of silicone oil from an eye with optic pit. *Graefe's Arch Clin Exp Ophthalmol (Albrecht Von Graefes Arch Klin Exp Ophthalmol)*. 2006;244(10):1360–1362.
9. Jain N, Johnson MW. Pathogenesis and treatment of maculopathy associated with cavitary optic disc anomalies. *Am J Ophthalmol*. 2014;158:423–435.
10. Berdahl JP, Fleischman D, Zaydlarova J, Stinnett S, Allingham RR, Fautsch MP. Body mass index has a linear relationship with cerebrospinal fluid pressure. *Investig Ophthalmol Vis Sci*. 2012;53:1422–1427.
11. Abouammoh MA, Alsulaiman SM, Gupta VS, et al. Pars plana vitrectomy with juxtapapillary laser photocoagulation versus vitrectomy without juxtapapillary laser photocoagulation for the treatment of optic disc pit maculopathy: the results of the KKESH International Collaborative Retina Study Group. *Br J Ophthalmol*. 2016;100:478–483.
12. Martins D, Bacalhau C, Santos MF. Etiology and treatment of serous macular detachment associated with optic pit – clinical case. https://www.evrs.eu/etiology-and-treatment-of-serous-macular-detachment-associated-with-optic-pit-clinical-case/; 2012, Accessed date: 1 July 2017.
13. Wolfensberger TJ. The role of carbonic anhydrase inhibitors in the management of macular edema. *Doc Ophthalmol Adv Ophthalmol*. 1999;97:387–397.
14. Supuran CT. Acetazolamide for the treatment of idiopathic intracranial hypertension. *Expert Rev Neurother*. 2015;15:851–856.