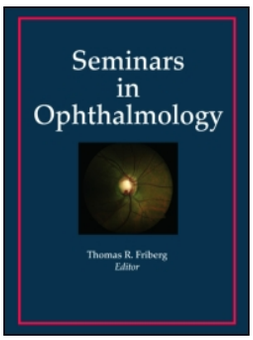

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ORIGINAL ARTICLE

Bilateral Macular Edema: A New Ocular Feature of Dandy–Walker Syndrome

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ABSTRACT

To describe a case of bilateral cystoid macular edema in a patient with Dandy–Walker syndrome. An 18-year-old male was referred to our tertiary referral center for evaluation of his decreased visual acuity. Detailed ophthalmic examination and imaging revealed the presence of bilateral cystoid macular edema, which was successfully treated with intravitreal triamcinolone injections (2 mg in 0.05 ml). Recurrence of macular edema developed after a period of approximately four months. This is an unusual ophthalmic manifestation of Dandy–Walker syndrome. Cystoid macular edema should be included in the differential diagnosis of subjects with Dandy–Walker syndrome presenting with decreased vision. The pathogenetic mechanism for the development macular edema in this case is not clear. Intravitreal triamcinolone is an effective treatment, but edema was recurrent in our case. Other approaches (such as oral Acetazolamide or intravitreal Anti-VEGF) have to be considered as well.

Keywords: Dandy–Walker syndrome, intravitreal triamcinolone, macular edema

We present the first case of bilateral cystoid macular edema in a patient with Dandy–Walker syndrome.

Dandy–Walker complex represents various posterior fossa malformations. The complex is characterized by a continuum of posterior fossa cystic abnormalities with varying degrees of vermian agenesis. There is a broad spectrum of severity ranging from absence of symptoms to severe disabilities or even death. The most commonly encountered malformation is the one initially described: the Dandy–Walker malformation. This is characterized by enlargement of the posterior fossa, agenesis of the cerebellar vermis and cystic dilation of the fourth ventricle. The malformation has been associated with intra- and extracranial anomalies, hydrocephalus and developmental problems.[1] Several ophthalmic manifestations have been associated with Dandy–Walker complex. These include nystagmus, cataract, choroidal or iris coloboma, microphthalmia, palpebral ptosis, hypertelorism, strabismus, and optic neuropathy due to hydrocephalus.[1–4] To our knowledge, this is the first report to describe macular edema in association with Dandy–Walker syndrome.

An 18-year-old male was referred to our eye clinic for ophthalmic examination and assessment of his decreased visual acuity. He had a history of Dandy–Walker complex and he complained of decreased visual acuity during the last three months. Previous MRI imaging revealed the absence of cerebellar vermis in association with cystic dilation of the fourth ventricle (Figure 1). He had no history of hydrocephalus or previous ophthalmic pathology. His overall medical history, other than the malformation, was unremarkable and he was not on any ophthalmic or systemic treatment.

Best-corrected visual acuity was 20/30 in both eyes with anterior and posterior segment examination showing no signs of inflammation. Fundoscopy revealed abnormal foveal reflex in both eyes and imaging with Spectral Domain Optical Coherence Tomography (Spectralis OCT–Heidelberg Engineering, Germany) confirmed the presence of bilateral small intraretinal cysts at the macula with no evidence of vitreoretinal traction. Fundus fluorescein angiography showed normal filling of the retinal vessels and leakage at the macula with no signs of inflammatory vasculitis. The patient was commenced on topical brinzolamide 1% eye drops b.d., which was considered a favorable, less invasive treatment for this case.

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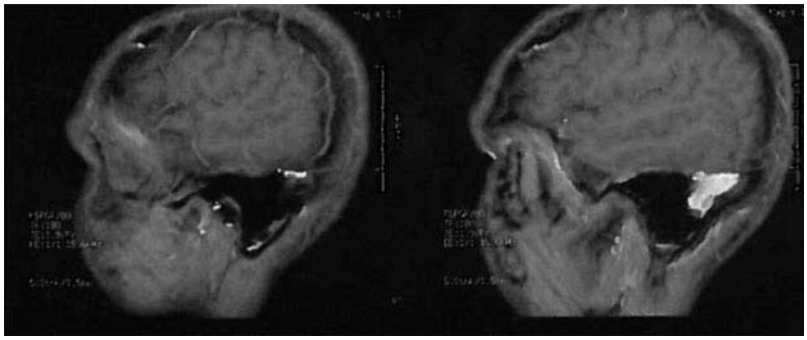

FIGURE 1. MRI imaging revealing the absence of cerebellar vermis in association with cystic dilation of the fourth ventricle.

Six weeks later, his best-corrected VA had decreased to 20/40 in his right and left eye and OCT showed an increase of central macular thickness and of the size of intraretinal cysts (Figure 2). Risks and benefits of potentially more invasive treatments were discussed and it was decided to proceed with intravitreal triamcinolone injections (0.05 cc of 40 mg/ml) in both eyes under general anaesthesia due to poor cooperation of the patient.

Best-corrected visual acuity improved to 20/25 in both eyes and central macular thickness decreased during the follow-up visits, one and two months after the injections. However, intraretinal cysts recurred four months after the injections, necessitating further intervention.

Clinical and imaging findings in our case did not reveal the exact etiology of inner and/or outer blood-retinal barrier impairment. The underlying mechanism for posterior fossa cyst development in Dandy–Walker patients has not been defined either. The simultaneous development of macula and central nervous system cysts could suggest similar mechanisms at cellular level.

Macular edema is the final common pathway of several intraocular and systemic insults. The pathogenesis of macular edema involves increased vascular permeability, increased blood flow, dysfunction of the RPE barrier/pump, tractional stress, drug reactions, or intraretinal fluid migration from optic nerve head abnormalities. The mechanism responsible for macular edema development in our case remains unclear. Macular edema has been associated with other neurological diseases (e.g., multiple sclerosis) as well, but the mechanism responsible in these cases can be usually identified.[5]

This report of macular edema in a patient with Dandy–Walker malformation broadens the spectrum of ocular findings that may be encountered in this disorder. Macular edema should be included in the differential diagnosis of visual loss in individuals with Dandy–Walker syndrome. In view of the previously

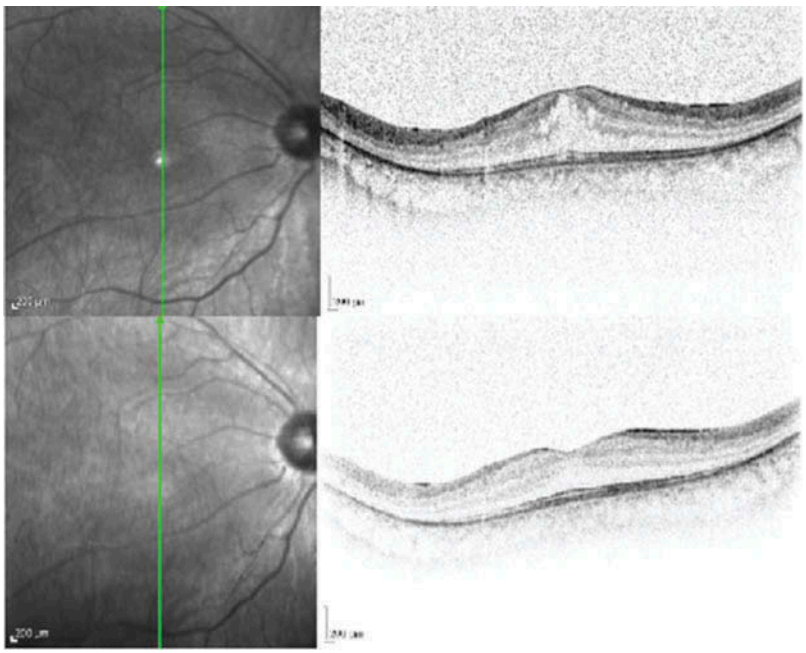

FIGURE 2. OCT showing an increase of central macular thickness and intraretinal cysts.

noted complication, periodic fundoscopy and imaging with optical coherence tomography are essential in order to effectively monitor the status of the macula.

DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of this article.

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