Intravitreal ranibizumab in the treatment of choroidal neovascularization secondary to morning glory syndrome in a child

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Abstract

Congenital optic nerve abnormalities may rarely cause choroidal neovascularization (CNV). This case report summarizes the clinical and therapeutic outcomes of a 7-year-old boy with unilateral CNV secondary to morning glory syndrome associated with acute visual acuity loss. The patient was successfully treated with a single intravitreal ranibizumab injection. One month after the injection the visual acuity increased and optic coherence tomography (OCT) showed a decrease in the intraretinal fluid around the CNV. The patient was then called for monthly follow-up visits. No further treatment was needed for the next 12 months after the first treatment. There was no complication related to the injection.

Keywords: Choroidal neovascularization, Morning glory syndrome, Ranibizumab

Introduction

Morning glory syndrome (MGS) is an uncommon and unilateral congenital anomaly which was coined by Kindler in 1970. The appearance of the anomaly was so similar to the tropical morning glory flower; therefore, this name was chosen. It is a funnel-shaped excavation of the posterior pole involving the optic disk. The excavation is filled with a white tuft of glial tissue and surrounded by a pigment ring. The number of retinal vessels is increased, and they appear to arise from the periphery of the disk, and tend to run to the peripheral retina. Choroidal neovascularization (CNV) secondary to congenital optic nerve abnormalities is a rare cause of visual loss in children. In this case report we aimed to present the clinical and therapeutic outcomes of a 7-year-old boy with unilateral CNV secondary to MGS.

Case report

A 7-year-old boy was admitted with the complaint of decreased visual acuity since 5 days in the right eye. Systemic evaluation was not notable; therefore, cranial magnetic resonance imaging (MRI) was not obtained. He had a history of amblyopia in his right eye. His family history was not significant. On ocular examination, best corrected visual acuity (BCVA) was 20/100 in the right eye and 20/20 in the left eye. Slit lamp examination of the anterior segment did not show any abnormality and intraocular pressure was within normal limits.
normal limits in both eyes. Fundus examination revealed a morning glory disk and a yellowish subretinal lesion at the temporal margin of the optic disk in the right eye (Fig. 1a). The fundus examination was not notable in the left eye. Optical coherence tomography (OCT) revealed a peripapillary choroidal neovascularization (CNV) associated with intraretinal fluid at the temporal margin of the optic disk in the right eye (Fig. 1). Fluorescein angiography (FA) showed the staining of the peripapillary CNV and a mild leakage (Fig. 2). Based on ocular findings and the history, the patient was diagnosed as peripapillary CNV secondary to morning glory syndrome. Informed consent for intravitreal ranibizumab injection was obtained from the parents, and 0.5 mg/0.05 ml intravitreal ranibizumab injection was then performed under general anesthesia. There was no complication related to the injection. After 4 weeks of treatment, BCVA improved to 20/50, and intraretinal fluid decreased significantly (Fig. 1b). The patient was then called for monthly follow-up visits, and the peripapillary CNV did not show any kind of activity (subretinal fluid, intraretinal fluid, visual loss, hemorrhage) for the next 12 months.

Discussion

The embryogenesis of MGS is not well understood yet. Previous studies have suggested that MGS was a primary mesenchymal abnormality and it was proposed to be the result of a faulty closure of the posterior scleral wall and the poor development of the lamina cribrosa. MGS was often associated with persistent hyperplastic primary vitreous (PHPV). Fei et al. reported that the coexistence of PHPV in a significant percentage of patients with MGS, might suggest a potential common genetic link between the two disorders. They added that compared with MGS or PHPV alone, the combination of the two conditions might manifest with higher incidence of more severe complications. The same study reported that PAX6 mutations that have also been identified in patients with optic nerve malformations, including coloboma, MGS, optic nerve hypoplasia/aplasia and PHPV might play an important role on the pathogenesis. However some other reports revealed that not all patients with MGS have the PAX6 mutation. Another common co-existing disorder with MGS is basal encephalocele. Sasani et al. reported that the presence of ophthalmologic findings such as strabismus in association with anomalies of optic nerve should always bring in mind the possible presence of an unrecognized skull base midline defect and encephalocele. The systemic associations of MGS are usually uncommon, and if present cranial anomalies are associated with mid-facial anomalies such as hypertelorism and flat nasal bridge. Mid-facial anomalies, basal encephalocele, and midline brain malformations are coined as frontonasal dysplasia. As our patient did not show any abnormality in clinical examination we did not obtain cranial MRI from him.

Strabismus may be the first symptom and serous retinal detachment may develop in up to 30% of patients with MGS. However CNV secondary to MGS is very rare. There is no established treatment regimen yet. Anti-VEGF therapy is currently indicated for the treatment of CNV, macular edema associated with vascular occlusion or diabetes mellitus and retinopathy of prematurity. Only a few cases of CNV secondary to MGS is reported in the literature. Two of the reported cases who were in adulthood were treated...
with intravitreal anti-VEGF agents. The first case was reported to be a 51-year-old male and received 2 intravitreal bevacizumab injections during the 6 month follow-up period. The visual acuity of the patient was reported to be increased from 0.1 to 0.6 in decimals. The second case was a 63-year-old woman who received three consecutive monthly ranibizumab injections and did not require any additional injections for the next 24 months. The visual acuity of the patient was reported to increase from 0.7 LogMAR to 0.1 LogMAR and remained stable for the next 24 months. In the current report we used a single intravitreal injection of ranibizumab for the treatment of peripapillary CNV secondary to MGS in a 7-year-old boy and no recurrence was observed during the 12 month follow-up period.

Conclusion

To our knowledge this is the first case of CNV secondary to morning glory syndrome in a child treated with ranibizumab. Peripapillary CNV secondary to optic nerve abnormalities may be successfully treated with intravitreal ranibizumab injections. This report adds to the evidence base accumulating for the use of ranibizumab in a case of CNV secondary to MGS. Flexible treatment regimens should be advised in pediatric patients rather than monthly injections in these settings.

Conflict of interest

The authors declared that there is no conflict of interest.

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