Retinal astrocytoma with exudative retinal detachment treated with photodynamic therapy in a young girl with tuberous sclerosis

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Abstract:
Around 50% of patients with tuberous sclerosis have the manifestation of retinal astrocytomas. Symptomatic retinal astrocytomas are very rare, with no consensus on optimal treatment. A 7-year-old patient with tuberous sclerosis presented with progressive blurred vision in the right eye for more than half a year. On examination, best-corrected visual acuity was 20/30 in the right eye and 20/20 in the left eye. Dilated fundus examination of the right eye showed a well-circumscribed, elevated, opaque lesion, with surrounding lipid exudate, and retinal detachment involving the fovea. Spectral-domain optical coherence tomography of the right eye revealed an intraretinal lesion with adjacent subretinal fluid. A diagnosis of retinal astrocytoma with exudative retinal detachment was made. The patient was treated with verteporfin photodynamic therapy (PDT) on the lesion in the right eye. After 1 month, complete resolution of subretinal fluid was observed. At 21-month follow-up, the right eye vision was stable at 20/20, without ocular or systemic adverse events. In conclusion, PDT had a good safety profile in a cooperative pediatric patient, and was able to induce regression of astrocytoma as well as resolution of exudation with excellent visual outcome.

Keywords:
Astrocytoma, retinal detachment, tuberous sclerosis

Introduction
Retinal astrocytomas are composed of foci with dystrophic calcification and several types of glial cells at the retinal nerve fiber layer. Around 50% of patients with tuberous sclerosis present retinal astrocytomas. Despite its benign histopathology, progressive growth, exudation, vitreous hemorrhage, secondary retinal detachment, neovascular glaucoma, and spontaneous scleral perforation have been reported. Herein, we present the case of a young tuberous sclerosis patient with retinal astrocytoma and exudative retinal detachment treated with verteporfin PDT.

Case Report
A 7-year-old girl with a history of tuberous sclerosis presented to our clinic with progressive blurred vision in the right eye for more than 6 months. The past medical history included excisional biopsy-proven subependymal giant cell astrocytoma in the left lateral ventricle and related infantile spasms at 4-month-old. On examination, best-corrected visual acuity was 20/30 in the
right eye and 20/20 in the left eye. Intraocular pressure and anterior segment examination were unremarkable. A dilated fundus examination of her right eye showed a well-circumscribed, elevated, opaque lesion, around 1.5 disc diameter, located superior to the superior vascular arcade with lipid exudate and exudative retinal detachment extending to the fovea [Figure 1a]; two other flat, smooth and translucent gray lesions were found in the left eye. Spectral-domain optical coherence tomography of the right eye revealed an intraretinal lesion causing disruption of retinal layers [Figure 1b], with surrounding subretinal fluid extending to the fovea [Figure 1c]. Fluorescein angiography showed a hyperfluorescent mass with vascular ingrowth located at 12 o’clock, with late leakage in the right eye [Figure 1d], as well as the presence of two hyperfluorescent patches with late staining in the left eye. Based on the history of tuberous sclerosis and the morphology of retinal lesions, retinal astrocytomas of both eyes were diagnosed. After explaining the pros and cons of intervention to the patient’s parents, PDT with verteporfin at standard dose, spots size 4200um, and 166 s was applied on the lesion in the right eye. At the 1-month follow-up, improvement of vision to 20/25 OD and complete resolution of subretinal fluid OD was observed. At 21 months [Figure 2] after intervention, there was no recurrence and right eye vision was stable at 20/20. No adverse events were noted.

**Discussion**

Spontaneous regression of symptomatic retinal astrocytoma within a few months has been reported,[6] however, the treatment was offered to our patient due to the history of worsening visual acuity in the right eye for more than 6 months. Regarding treatment options, laser photocoagulation was not chosen due to possible complications including laser-induced choroidal neovascularization and thermal destruction of the retina.[11] In previous literature, six cases of symptomatic retinal astrocytoma, including two patients with tuberous sclerosis, were treated with PDT.[12] The patients were between 18 and 68 years of age and had varying degrees of visual improvement after PDT (final visual acuity ranging from 20/800 to 20/20). Good anatomical outcome was achieved in all cases with complete resolution of subretinal fluid, without any complications. The success in previous cases encouraged us to choose PDT as the treatment for this patient.

Previous publications of PDT in the pediatric population have been limited to small case series or case reports, most of which were treatment of choroidal neovascularization.[7-10] Mimouni et al.[7] were the first to report on beneficial results after PDT in subfoveal choroidal neovascularization lesions in three young patients (aged 11–13 years). Although treatment regimens and disease entities differed, no serious pediatric ocular or systemic adverse events related to PDT have been reported in literature thus far.[7-10]

This is the youngest patient with retinal astrocytoma and exudative retinal detachment successfully treated with verteporfin PDT. PDT appears to be a safe and effective treatment modality for the management of retinal astrocytoma, and may be considered in cooperative pediatric patients complicated with nonresolving subretinal fluid.

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Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient’s guardian has given consent for the child’s images and other clinical information to be reported in the journal. The patient’s guardian understands that the child’s name and initials will not be published and due efforts will be made to conceal the child’s identity, but anonymity cannot be guaranteed.

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Conflicts of interest
The authors declare that there are no conflicts of interests of this paper.

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