Rubeosis Irides Complicating Acquired Astrocytoma of Solitary Optic Nerve Head; a Clinicopathologic Case Report

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Abstract

Purpose: To report a rare case of large vascularized astrocytoma on optic disc.

Case Report: A 12-year-old boy with history of decreased vision in the left eye since 5-6 months ago was referred to the ocular oncology service at Rassoul Akram Hospital with provisional diagnosis of retinoblastoma.

On examination, visual acuity was counting fingers at 50 cm, and a 3+ relative afferent pupillary defect response as well as an exodeviation was evident in the left eye. Fundus examination of the left eye revealed a large yellow-white mass with superficial vessels overhanging the optic disc. Right eye slit lamp exam and funduscopy were unremarkable.

No clinical manifestation of tuberous sclerosis was found.

Orbital CT scan, MRI and ultrasonography of the globe was done.

Enucleation was performed because of the development of iris neovascularization and progressive enlargement of the tumor. Histologic exam confirmed the diagnosis of astrocytoma.

Key words: Rubeosis Irides, Optic Nerve Head, Astrocytoma, Optic Disc, Iris Neovascularization.

Introduction

Although the optic nerve head or retinal astrocytic hamartoma which appears in early age considered as a benign congenital tumor that usually occurs as part of tuberous sclerosis (TS) or neurofibromatosis (NF), acquired astrocytoma appears typically as a white to fleshy pink vascularized mass, usually around the optic disc in otherwise healthy persons, which may show progressive growth and can produce secondary complications. It may enlarge slowly and affected patient becomes symptomatic whenever associated with retinal detachment, subretinal exudation and vitreous hemorrhage.

Histopathologically the tumor is composed of either typical mature retinal astrocytes or Muller cells.

Since the diagnosis is so difficult to establish clinically, most affected eyes have been managed by enucleation because of clinical diagnosis of retinoblastoma or melanoma. We herein report a case of optic nerve head acquired astrocytoma with no systemic association that simulated a retinoblastoma and complicating with rubeosis irides.

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Case Report

A 12-year-old boy was referred to the oncology service because of decreased vision in the left eye. Systemic evaluation of the patient was completely normal and there was no history of intraocular inflammation or trauma.

The best corrected visual acuity was 20/20 in the right eye and 50 cm, finger counting in the left eye.

Intraocular pressure, slit lamp exam and ocular motility were normal, except for left eye exodeviation. There was a 3 plus relative afferent pupillary defect in the left eye. Indirect ophthalmoscopy revealed one sessile, fleshy, elevated yellow-white mass approximately 9×8 mm in largest diameter with superficial vessels and the tumor was so overhanged on the posterior pole that the optic disc and macula were invisible (Figure 1). The right fundus was completely normal. B-Scan ultrasonography showed an elevated mass adjacent to the optic disc in the retina measured 9 mm in height and high internal reflectivity was evident in A-scan (Figure 2-a).

Computerized tomography scan and MRI revealed one hyperintense mass in the left eye on the optic nerve without any evidence of subependymal hamartomas (Figure 2-b and 2-c).

According to clinical and imaging findings, retinoblastoma, astrocytic hamartoma and retinal gliosis were considered as differential diagnosis and close follow-up was scheduled. On next visit, progressive tumor enlargement and development of iris neovascularization complicated the clinical picture of the patient. The clinical findings were discussed with his family and finally enucleation was done. In macroscopic examination a well defined homogenous grayish-pink mass attached to optic disc was evident (Figure 3-a).

Microscopic sections of the mass displayed fascicles of elongated tumoral cells as well as aggregates of plump polyhedral forms with abundant eosinophilic cytoplasm and eccentrically placed vesicular nuclei possessing prominent nucleoli (Figure 3-b). In immunoperoxidase stains these cells were positive for glial fibrillary acidic protein (GFAP) and negative for epithelial membrane antigen as well as synaptophysin (Figure 3-c). The outer most rim of the tumor mass was covered by an attenuated multilayered rim of retinal cells.

Based on the histopathologic examination the diagnosis of retinal astrocytic hamartoma was made. Systemic examination did not reveal any evidence of NF and TS. One year after enucleation, the boy was doing well with neither tumor recurrence nor features of TS or NF.
Figure 2-b. Computed tomography scan showing a mass occupying the posterior segment of the left eye.

Figure 2-c. In MRI there is a round intraocular mass 9 mm in diameter, at the junction of optic nerve with intact wall and isosignal with white matter.

Figure 3-a. Pupillo-optic section of globe contains a homogenous grayish-pink mass arose from optic disc.

Figure 3-b. Positive immunoreactivity for GFAP.

Figure 3-c. Large eosinophilic tumoral cells with eccentrically placed vesicular nuclei and prominent nucleoli.
Discussion

Acquired retinal astrocytoma which can develop at any age should be differentiated from astrocytic hamartoma, and reactive gliosis. As a rule, patients with reactive gliosis have a history of prior ocular inflammation or trauma, and the involved eye is more degenerated.\textsuperscript{1,2}

Retinal astrocytic hamartoma, appearing solitary or multifocal, is typically a benign tumor in association with TS (Bourneville’s disease) or NF (Von Recklinghausen’s syndrome).\textsuperscript{1,2,4}

However, acquired retinal astrocytoma occurs sporadically in patients without any evidence of systemic TS and NF or before ocular inflammation and trauma.

The most common type of astrocytic hamartomas is a fairly flat, soft, semi-translucent lesion of the nerve fiber layer and usually remains stable. However, progressive calcification of the mass can change the clinical picture of the mass like a mulberry in some cases.\textsuperscript{6}

Fewer than 20 cases of solitary lesion unassociated with phakomatosis have been reported in the literature (Table 1).\textsuperscript{6}

According to our best knowledge, only one case of progressive retinal astrocytoma with total retinal detachment and proliferative retinopathy has been reported by Bornfeld et al\textsuperscript{8} in which the clinical picture was complicated by rubeosis irides. Since there was no significant subretinal fluid in our case, we presume that iris neovascularization has developed following some intratumoral necrosis or some retinal ischemia due to compressive optic neuropathy. As shields et al\textsuperscript{6} have emphasized the lack of evidence for calcification in acquired astrocytoma, histopathologic evaluation of the tumor as well as CT-scan and B-scan echography of the patients did not depicted any focus of calcification in the tumor. In summary we present a case of acquired retinal astrocytoma in an otherwise healthy boy which complicated by rubeosis irides.

In addition, we are aware that in such case we should consider longer follow-up for developing any systemic manifestation of TS in the first or second decades of the patient’s life.

### Table 1. Review of published cases of retinal and optic disc astrocytoma not associated with tuberous sclerosis or neurofibromatosis.

<table>
<thead>
<tr>
<th>Authors</th>
<th>Date</th>
<th>Age</th>
<th>Race</th>
<th>Gender</th>
<th>Tumor location</th>
<th>Tumor course (mos)</th>
<th>Preoperative diagnosis</th>
<th>Treatment</th>
<th>Pathologic diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Marks et al</td>
<td>1939</td>
<td>6 yr/W/M</td>
<td>Retina juxtapapillary</td>
<td>Growth (62)</td>
<td>NA</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
<td></td>
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<tr>
<td>Boles et al</td>
<td>1958</td>
<td>6 yr/W/F</td>
<td>Retina juxtapapillary</td>
<td>Growth (12)</td>
<td>NA</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
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<tr>
<td>Foos et al</td>
<td>1965</td>
<td>14 yr/W/F</td>
<td>Retina juxtapapillary</td>
<td>Growth (204)</td>
<td>Retinal angioma</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
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<tr>
<td>Cleasby et al</td>
<td>1967</td>
<td>20 mo/W/M</td>
<td>Retina juxtapapillary</td>
<td>NA</td>
<td>Retinoblastoma</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
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<tr>
<td>Jordano et al</td>
<td>1974</td>
<td>9 yr/F</td>
<td>Retina juxtapapillary</td>
<td>Growth (8)</td>
<td>Tuberculoma</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
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<tr>
<td>Reeser et al</td>
<td>1978</td>
<td>6 yr/W/M</td>
<td>Retina juxtapapillary</td>
<td>Growth (84)</td>
<td>Astrocytoma</td>
<td>Enucleation + external radiation</td>
<td>Astrocytoma</td>
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<td></td>
</tr>
<tr>
<td>Ramsay et al</td>
<td>1979</td>
<td>41 yr/W/M</td>
<td>Retina juxtapapillary</td>
<td>NA</td>
<td>Retinal angioma</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
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<td></td>
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<tr>
<td>Jakobiec et al</td>
<td>1983</td>
<td>5 mo/M</td>
<td>Retina juxtapapillary</td>
<td>NA</td>
<td>Retinoblastoma</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
<td></td>
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<tr>
<td>Ulbright et al</td>
<td>1984</td>
<td>13 mo/F</td>
<td>Retina juxtapapillary</td>
<td>NA</td>
<td>Retinoblastoma</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
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<tr>
<td>Arnold et al</td>
<td>1985</td>
<td>16 yr/W/F</td>
<td>Retina juxtapapillary</td>
<td>Growth (7)</td>
<td>NA</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
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<tr>
<td>Bornfeld et al</td>
<td>1987</td>
<td>56 yr/W/M</td>
<td>Retina juxtapapillary</td>
<td>Growth (4)</td>
<td>Choroidal melanoma</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
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<tr>
<td>Sharma et al</td>
<td>1991</td>
<td>2 yr/F</td>
<td>Optic disc</td>
<td>NA</td>
<td>Retinoblastoma</td>
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<td>Astrocytoma</td>
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<tr>
<td>Lee et al</td>
<td>1996</td>
<td>3 yr/F</td>
<td>Retina periphery</td>
<td>NA</td>
<td>Retinoblastoma</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
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<tr>
<td>Shields et al</td>
<td>2002</td>
<td>35 yr/W/M</td>
<td>Retina juxtapapillary</td>
<td>Growth (24)</td>
<td>Choroidal melanoma</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
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<tr>
<td></td>
<td></td>
<td>14 yr/W/F</td>
<td>Retina juxtapapillary</td>
<td>Growth (37)</td>
<td>Astrocytoma</td>
<td>Enucleation</td>
<td>Astrocytoma</td>
<td></td>
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Authors have no financial relationship with the manufacturer of any product discussed in this manuscript.
References