males and five were females, and all were aged 5 to 13 years (6.5 ± 2.1 years). All patients had unilateral eye disease and had been misdiagnosed for bilateral squint or amblyopia for years. All eyes (5 eyes were esotropia and 3 eyes were exotropia) were associated with refractive errors and were able to make normal eye movements. All patients had normal intraocular pressure, and denied any family history; no other abnormalities were detected. Five patients could not cooperate with the fundus examination and one patient had lens opacities. B ultrasound confirmed that all eight patients had retinal detachment and optic disc dysplasia. During the surgery, all were confirmed to have MGS, MH, PVD, and subretinal proliferation. Two cases had a funnel-shaped bulge. Large excavated discs (about 1.5PD in diameter) were present, with retinal vessels radiating from the periphery. A tuft of translucent fibrous tissue covered the central part of the excavation and peripapillary chorioretinal pigmentary change was evident. The macular area with its xanthophyll pigment was displaced to the temporal margin of the excavation, while the papillary region was filled with acoustically dense material reminiscent of a (withering) flower and surrounded by a broad elevated, yellowish-white zone of chorioretinal pigmentary disruption. No central artery or vein was clearly distinguished; the blood vessels appeared to arise from the peripheral disc substance and course radially over the peripapillary annulus toward the retinal periphery. The vessels appeared to increase in number and to run an abnormally straight course, while tending to branch at acute angles. Serious macular detachment was involved and macular holes were detected in the abnormal expansion excavation of the optic disk.

Methods

All patients underwent standard 3-port pars plana vitrectomy surgery (20G for three cases and 23G for five cases), seven transparent lenses were well-preserved and one patient with cataract was treated using phacoemulsification. The epiretinal and subretinal membranes were completely peeled during the surgery; subretinal proliferation could be cut through the macular hole. The internal limiting membrane peeling and the gas/liquid exchange process were followed by flattening of the peripapillary retina with heavy liquids. Joint photocoagulation in the abnormal expansion excavation of the optic disk and silicone oil tamponade were also performed. The laser should be directed obliquely away from the posterior pole to avoid the macular area.

Results

Table 1 shows the clinical data for the patients. All eyes achieved anatomical resolution. After follow-ups ranging from eight months to four years, the visual function for all patients was improved by postoperative refractive correction associated with vision training. Best corrected visual acuity reached 0.01 to 0.2 at the final follow-up. No retinal detachment recurred and no silicone oil flowed into the subretinal space. The silicone oil was successfully removed postoperatively after 0.5 years in five cases and after 1-1.5 years in three cases. No recurrence of retinal detachment occurred during the follow-ups and the MHs closed in five cases (5/8, 62.5%).

Table 1 Basic data of patients in this study

case	age	sex	cataract (+/-)	PVR (+/-)	silicone oil filling time	macular hole (+/-)	Follow-up Time postoperative	preoperative visual acuity	postoperative visual acuity (last follow-up)
1	5	male	-	+	5 m	-	4y	FC/30 cm	0.01
2	11	female	-	+	17 m	+	8m	FC/30 cm	0.01
3	6	male	-	+	7 m	+	2y	FC/30 cm	0.05
4	7	male	-	+	6.5 m	+	3y	0.05	0.2
5	6	female	-	+	14 m	-	2.5y	0.01	0.05
6	9	female	+	+	3 m	+	10m	0.02	0.1
7	7	male	-	+	11 m	-	1y	FC/30cm	0.1
8	13	female	-	+	4 m	+	4y	0.01	0.2

cataract(+/-) - + with cataract; - without cataract

macular hole(+/-) - + closed macular hole; - unclosed macular hole

PVR -+ with proliferative vitreoretinopathy; - without proliferative vitreoretinopathy

Case Report

Presumed acquired retinal astrocytoma in association with anterior uveitis

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Abstract

Purpose: To report a case of presumed acquired retinal astrocytoma in association with anterior uveitis.

Methods: A 29-year-old man presented with mutton fat keratic precipitates in the lower cornea, with complicated cataract and a circumscribed, solitary, yellowish-white retinal lesion in the right eye. Phacoemulsification with intraocular lens implantation was performed, with election to observe the lesion periodically. The lesion was followed for two years without any change in size, shape, and appearance. The anterior uveitis has not recurred at the time of writing.

Results: Systemic medical and laboratory evaluations, including chest computed tomography, cranial magnetic resonance imaging, and serum angiotensin converting enzyme level, were normal. The characteristic fundus, FA, OCT scan, ultrasound scan, and lack of other contributory laboratory findings strongly supported the diagnosis of acquired retinal astrocytoma in this patient.

Conclusion: We hypothesized that anterior uveitis may contribute to the growth and maintenance of retinal lesions. (***Eye Science 2013***; ***28***: –)

Keywords: acquired retinal astrocytoma; complicated cataract; anterior uveitis

Introduction

Acquired retinal astrocytoma is a rare benign tumor that occurs within the sensory retina as a yellowish-white mass. This tumor resembles a retinal astrocytic hamartoma, and it is rarely associated with tuberous sclerosis or neurofibromatosis[1]. Various ocular complications, including vitreous hemorrhage and seeding, subretinal hemorrhage and exudation, and even retinal detachment and secondary glaucoma have been reported in association with these tumors[2, 3].

However, no association has yet been reported between complicated cataract and anterior uveitis, to the best of our knowledge (MEDLINE search). Here, we describe an unusual case of presumed acquired retinal astrocytoma in association with complicated cataract and anterior uveitis.

Case report

A 29-year-old man from the outskirts of Tangshan HeBei province was referred to our department. He complained of decreased visual acuity, with pink eye in his right eye for the past three months. Visual acuity was 20/40 in the right eye and 20/20 in the left eye. Slit-lamp examination revealed two mutton fat keratic precipitates in the lower cornea of the right eye, with posterior subcapsular lens opacity (Figure 1). Aqueous rays and cells were absent. The iris and pupil were normal, with no peripheral anterior synechiae. The left eye was entirely normal. The intraocular pressure was 11 mmHg OD, and 12 mmHg OS.

Fundus examination of the right eye showed a circumscribed solitary, yellowish-white retinal lesion along the superotemporal arcade. Fluorescein angiography demonstrated filling of vessels in the arteriovenous phase, with diffuse late-phase hyperfluorescence. Autofluorescence showed hypofluorescence of the mass. Blocking of choroidal fluorescence due to a retinal component of the lesion was also observed. The OCT images revealed a retinal tumor of an intrinsic "moth-eaten" appearance localized within the nerve fiber layer (NFL) with the outer retinal layers intact (Figures 2). B-scan ultrasonography revealed that the mass was acoustically solid without calcifi-

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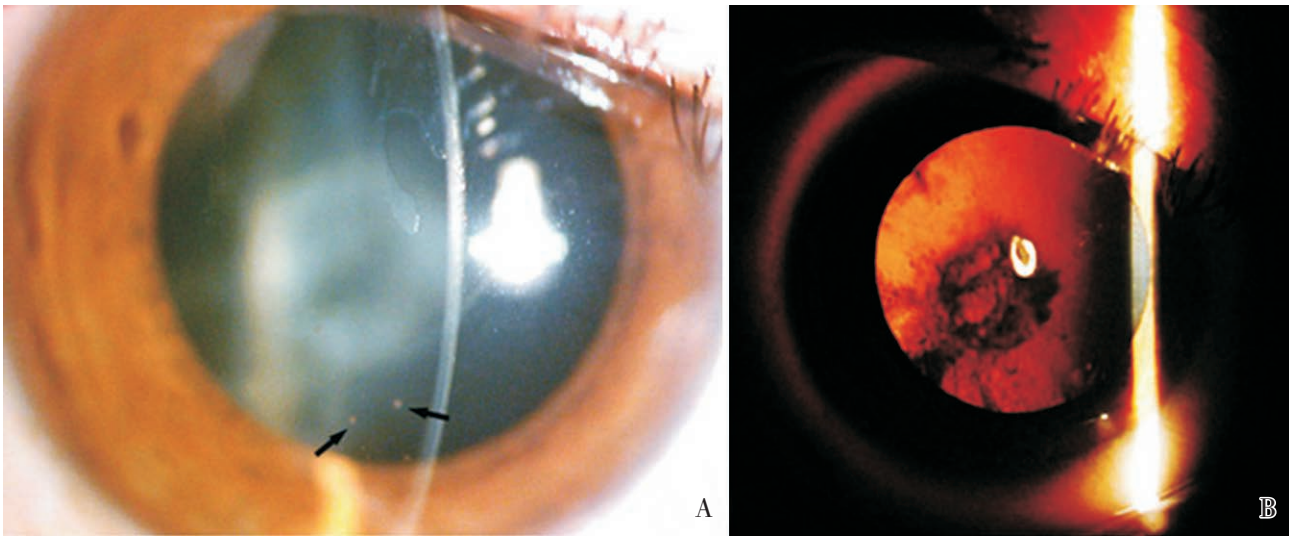

Figure 1 A, Keratic precipitates (arrows) are visible on the lower part of the corneal endothelium. B shows the posterior subcapsular lens opacity

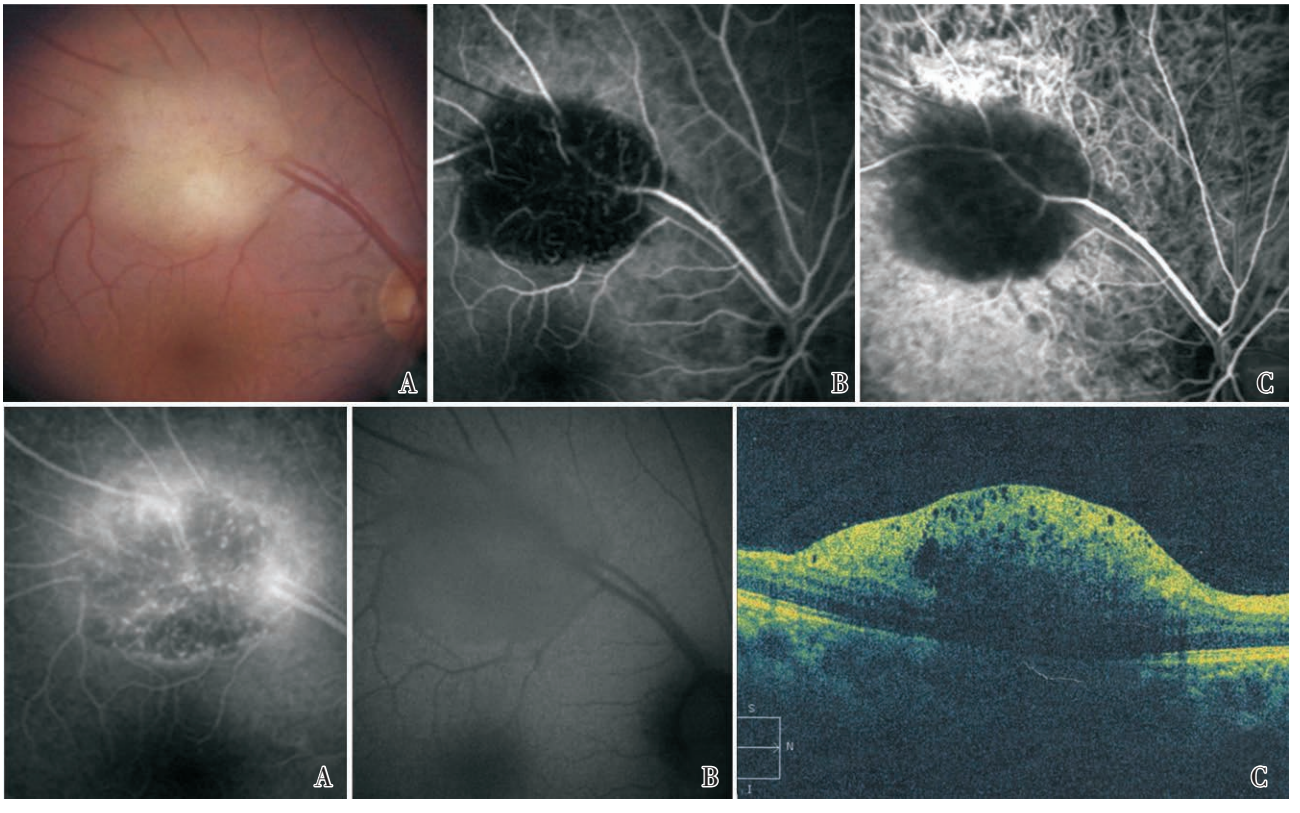

Figure 2 A, Color fundus photograph of the right eye showing a circumscribed yellowish-white retinal lesion. B, Fluorescein angiogram (arteriovenous phase) reveals fine intrinsic vessels. C, Indocyanin angiogram shows blocked choroidal fluorescence due to a retinal component of the mass. D, Late phase of the fluorescein angiogram showing diffuse hyperfluorescence. E, Autofluorescence shows hypofluorescence of the mass. F, OCT showing gradual transition from a normal retina to the hyperreflective intraretinal mass with loss of retinal anatomic organization. Note the moth-eaten, optically empty spaces and prominent optical shadowing posteriorly

cation (Figures 3).

Systemic medical and laboratory evaluations, including chest computed tomography, cranial magnetic resonance imaging, and serum angiotensin con-

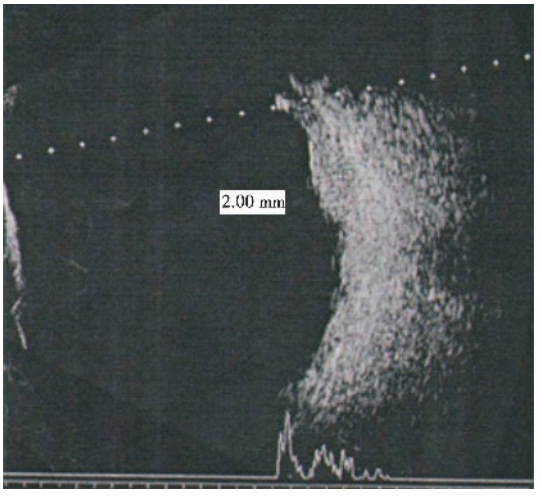

Figure 3 B-scan ultrasonograph shows an acoustically solid mass without calcification

verting enzyme level, were normal. A tuberculin skin test was negative.

No signs were evident of prior retinal trauma, inflammation, or posterior vitreous detachment. The patient was otherwise healthy, without previous eye trauma or infection. No systemic evidence was present for tuberous sclerosis or neurofibromatosis, and the family history had no incidence of tuberous sclerosis or neurofibromatosis. He gave no history of cat-scratch disease or contact with cats.

Slit lamp examination revealed a history of anterior uveitis as the patient had mutton-fat keratic precipitates and cataract formation associated with chronic inflammation. However, three months after the acute episode, the patient was asymptomatic with no redness, flare, or other signs of active inflammation detectable. We therefore diagnosed complicated cataract and presumed acquired retinal astrocytoma, as he had none of the clinical findings associated with tuberous sclerosis complex.

The patient underwent phacoemulsification with intraocular lens implantation and his visual acuity was 20/20 after surgery. Periodic lesion observation was elected as follow-up and no change in size, shape, of appearance was noted for two years. The anterior uveitis has not recurred at the time of writing.

Discussion

Endophytic retinal tumor and pseudotumors that originate from the glial cells comprise a spectrum of lesions including astrocytic hamartoma, acquired retinal astrocytoma, massive gliosis, and presumed solitary circumscribed retinal astrocytic proliferation (PSCRAP)[1–4].

Retinal astrocytic hamartoma presents in younger patients, and is frequently associated with central nervous system astrocytomas in the tuberous sclerosis complex or with neurofibromatosis[5,6]. Astrocytic hamartomas are generally stationary, but some lesions may show progression. Some studies have found that while smaller peripheral lesions remained stable, those located around the optic disc had a greater tendency to progress[7].

Acquired retinal astrocytomas, which some authors refer to as spontaneous retinal astrocytic hamartomas, are gelatinous, yellowish-white tumors that occur sporadically in patients without systemic tuberous sclerosis or neurofibromatosis. Compared with an astrocytic hamartoma, the acquired retinal astrocytoma is more likely a unilateral, unifocal lesion that generally lacks calcification and presents in older children or adults; otherwise, it is similar ophthalmoscopically[1,8]. Optical coherence tomography (OCT), ultrasound scans, and fluorescein angiography (FA) may aid in the diagnosis of this type of lesion.

A/B scanning will demonstrate focal calcification of the lesion and acoustic shadowing behind the lesion[9]. FA demonstrates delayed and gradually increasing hyperfluorescence throughout the angiography because a low number of astrocytic hamartoma vessels have abnormalities, including irregular swelling, enlargement, or extensive calcification[8]. OCT demonstrates an inner retina or full-thickness retinal disorder, with a gradual transition from a normal retina into a tumorous retina. The most striking finding on OCT was the presence of multifocal, round, confluent "moth-eaten" lucencies in the anterior of the mass, with posterior shadowing, which was presumed to represent foci of calcification or intratumor cavities[10].

Massive gliosis is typically a reactive process, usually occurring after posterior segment diseases such as congenital malformations, trauma, vascular disorders, and chronic inflammatory conditions re-

sulting in atrophic phthisis bulbi[4]. In massive gliosis, signs of previous retinal inflammation and retinal pigment epithelial proliferation are usually present.

The term presumed solitary circumscribed retinal astrocytic proliferation (PSCRAP) was first used by Shields[2] to describe an endophytic solitary white or yellow retinal mass located in the sensory retina. He described seven cases of a unique retinal lesion termed PSCRAP. Six lesions were stable after a median follow-up of six years, and one lesion gradually disappeared. All patients were asymptomatic and ultrasonography revealed no calcification. In OCT, PSCRAP does not demonstrate the "moth-eaten" appearance that characterizes most calcified astrocytic hamartomas and all lesions can show autofluorescence. However, autofluorescence showed hypofluorescence for the acquired retinal astrocytoma.

Other possible diagnostic options included retinoblastoma, sarcoidosis, and tuberculosis. Retinoblastoma is usually diagnosed in young patients who experience extensive tumor growth over time. In sarcoidosis, anterior segment disease occurs most commonly and usually takes the form of a chronic granulomatous uveitis, but involvement is usually bilateral. The levels of the serum markers angiotensin-converting enzyme and lysozyme were elevated and the chest imaging showed characteristic sarcoidosis findings of hilar lymphadenopathy and pulmonary nodules[11]. Tuberculosis is a well-known cause of acute or chronic granulomatous anterior uveitis. Posterior segment involvement of tuberculosis manifested as multifocal choroiditis, which can be divided into four groups: choroidal tubercles, choroidal tuberculoma, serpiginous-like choroiditis, and subretinal abscess[12].

When trying to diagnose this unusual case, we were faced with the dilemma that no histopathological evidence is available and we had to judge the case solely on clinical parameters. The characteristic fundus circumscribed solitary, yellowish-white retinal lesion, and the FA, OCT, and ultrasound scan results, together with the lack of other contributory laboratory findings, strongly support the diagnosis of acquired retinal astrocytoma in this patient.

In our patient, we excluded sarcoidosis and tuberculosis based on clinical manifestations and auxiliary examination results, but the pathogenesis of the anterior uveitis is still unknown. An association between anterior uveitis and acquired retinal astrocytoma has never been described before in the literature. This association is probably incidental because a pathogenetic relationship between them has never been established. Whether anterior uveitis contributes to the growth and maintenance of the presumed astrocytic hamartoma is still unknown and further studies are certainly required.

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