Chinese Medicine, **2011, 2, 16-19**
doi:10.4236/cm.2011.21003 Published Online March 2011 (http://www.SciRP.org/journal/cm)

Primary Pupillary Margin Cyst of the Iris Pigment Epithelium

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Received January 13, 2011; *revised February* 2, 2011; *accepted February* 10, 2011

Abstract

Purpose: Description of a patient with a solitary cyst of the pupillary margin iris pigment epithelium (IPE). Methods: A 63-year-old man referred a suspected iris-ciliary body melanoma in his left eye. Based on both clinical examination and ultrasound biomicroscopy, melanoma was considered unlikely. Surgery was undertaken to correct recurrent deterioration of vision due to movement of the lesion across the visual axis. Results: The lesion was excised completely. Ultrasound biomicroscopy and histopathological examination ruled out melanoma and allowed a final diagnosis of primary pupillary margin cyst of the IPE, characterized of pigmented epithelium, with no connective tissue or vessels. No recurrences or fresh lesions appeared during a one-year follow-up. Conclusions: Primary epithelial iris cysts are usually benign. Treatment is required only in symptomatic patients and those with an uncertain diagnosis. Ultrasound biomicroscopy is indispensable to confirm the clinical diagnosis, follow the clinical course and intervene if surgery is required.

Keywords: Melanoma, Iris Pigment Epithelium, Pupillary Cyst, Surgical Excision, Ultrasound Biomicroscopy

1. Introduction

Primary cysts in the anterior segment, observed during a routine eye examination, are divided into stromal and pigment epithelial cysts [1]. Primary stromal cysts are less common and mostly congenital [2]. Their differential diagnosis from malignant uveal melanoma may sometimes be difficult. Primary cysts are of neuroepithelial origin, whereas secondary cysts may result from intraocular implantation of surface epithelium following surgery or a penetrating trauma, or else from metastasis, parasites, or chronic myotic therapy [3]. Neuroepithelial cysts involve the iris pigment epithelium (IPE) and the ciliary body [4].

Primary cysts of the IPE are uncommon and spontaneous. They are defined as epithelial-lined structures that arise in the posterior iris layer [5]. Shields (1981) distinguishes four types, according to their position in relation to the iris [1]: central, at the pupillary margin; midzonal, between the pupillary margin and the iris root; peripheral, at the iridociliary sulcus; dislodged, in the anterior chamber or the vitreous cavity. The vast majority (76%) is peripheral [1].

This paper describes a case of symptomatic primary central IPE cyst initially mistaken for an iris melanoma. Stress is laid on ultrasound biomicroscopy (UBM) as a noninvasive way of determining the nature of a cyst, establishing its final diagnosis and following its course.

2. Case Report

A 63-year-old man presented with a two-month history of disturbed vision in his left eye and a suspected iris melanoma. In distant vision, his best corrected visual acuity (BCVA) was 20/20 in both eyes, but his left-eye disturbance was aggravated by strong daylight, which caused a severe temporary deterioration of vision (≤ 20/50). Slit-lamp examination showed a solitary, homogeneously pigmented mass hanging at the superior pupillary margin of the iris and at 1 o'clock in the anterior chamber (**Figure 1**). It slowly moved when the patient changed the position of his head. It was smooth, dark-brown and apparently cystic. The pupil was round, readily reacting to light and accomodation. During miosis, the lesion often obstructed the pupillary aperture. The slit lamp also revealed a normal left external eye,

cornea and conjunctiva, both before and after papillary dilatation, with no signs of inflammation in the anterior chamber. The right eye was normal. Examination with a Goldmann goniolens disclosed normal angular structures in both eyes. A Goldmann applanation tonometer showed an intraocular pressure of 14 mmHg in each eye. The fundi oculi were unexceptional. UBM of each eye was performed with a Humphrey 840 instrument, using a 50 MHz probe and an immersion technique. An immersion scleral shell was placed on the globe between the eyelids and filled with 2% methylcellulose. The probe was moved at right angles to produce radial and transverse sections. A 5 x 5 mm cyst with a thin, medium reflective wall and acoustically clear content (**Figure 2**) was observed in the left eye. The patient was duly informed that his cyst could be removed or kept under regular observation.

He chose surgery because of his disturbing visual symptoms. Surgery was then performed under topical anesthesia. A blade was used to make a 3-mm limbal section, followed by controlled entry into the anterior chamber. Downward rotation of the globe resulted in the spontaneous rupture and collapse of the cyst. It was then extruded through the limbal section (**Figure 3**). The wound was closed with a single 10-0 monofilament nylon suture. Transmission electron microscopy was used to examine the structure of the wall (**Figure 4**). Histologically, the iris simplex cyst is break off and partially lined by are present (**Figure 5**). It was therefore classed according to Shields [1] as a primary pupillary margin IPE cyst. After surgery, topical 0.1% fluorometholone was administered 4 times a day. One week later, BCVA was 20/20 in both distant and near vision. The intraocular

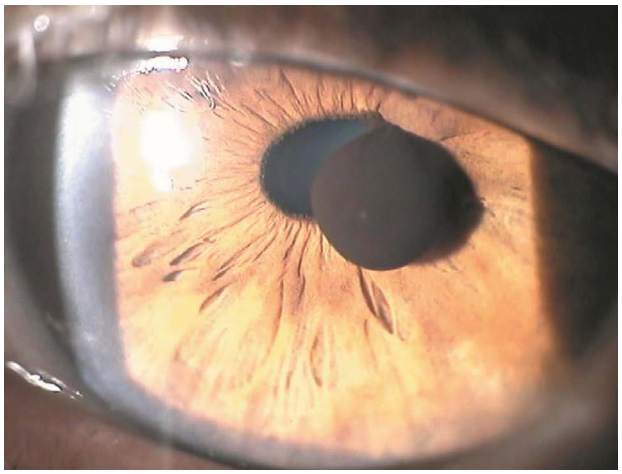

Figure 1. Solitary, dark-brown cyst in the supero-temporal aspect of the pupillary margin.

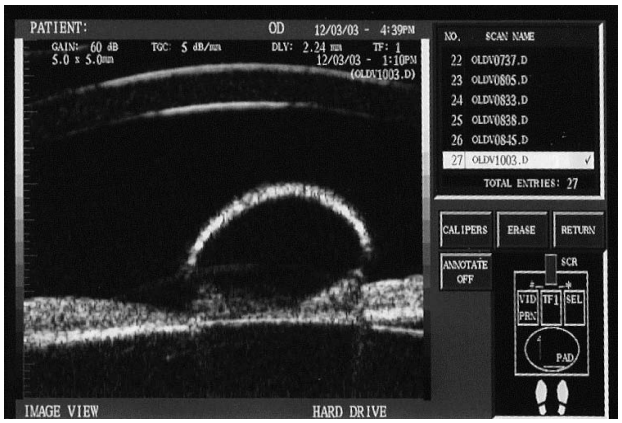

Figure 2. Ultrasound biomicroscopy of the left eye shows a pigmented cyst with a thin, medium reflective wall and acoustically clear contents, measuring 5.0 × 5.0 mm of diameter.

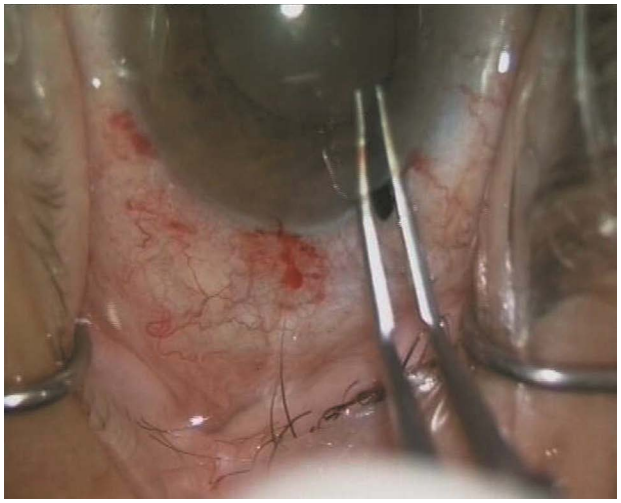

Figure 3. As the globe was rotated downwards, the cyst spontaneously ruptured, collapsed and was then extruded out through the limbal section.

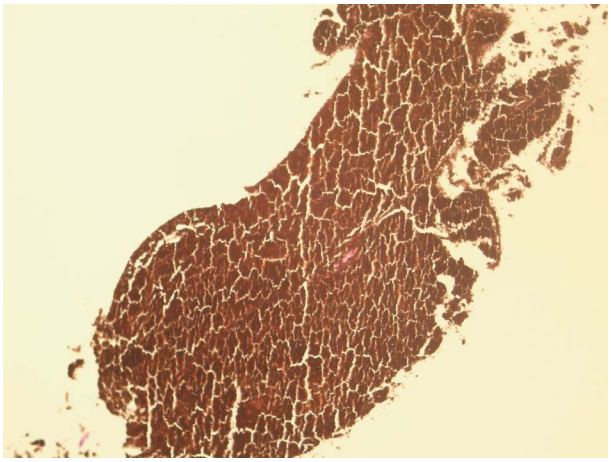

Figure 4. Microvillous processis are evident along the apical portions of the cells, connecting by occludens and adherens junctions. Small round or ovoid granules in early stages of melanization are seen (TEM; original magnification 11500x).

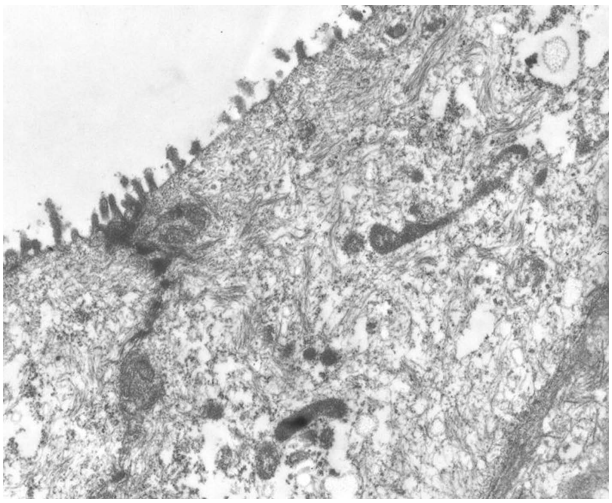

Figure 5. Histopathological examination of the tissue shows that it consists entirely of pigmented epithelium without connective tissue or vascular elements.

pressure, the shape of the anterior chamber and the reaction of the pupil to light were normal.

3. Discussion

Primary IPE cysts are found anywhere along the posterior surface of the iris from the pupillary margin to the anterior ciliary body. The great majority are stationary and small, and sometimes regress spontaneously. Their presence is occasionally responsible for angle closure glaucoma, plateau iris syndrome and secondary pigment dispersion syndrome. [6-9]. Central cysts account for approximately 3% of all primary IPE cysts [1,5]. They are usually dark brown, round or oval with a thin wall and sonolucent contents [1-5]. The high reflectivity of their wall is attributed to the epithelial cells, while their sonolucent core is compatible with a fluid content. The origin of primary central IPE cysts is not clear. Their appearance in early childhood in a few familial cases suggests that they may be hereditary with an autosomal-dominant pattern [7-10]. Lewis *et al.* [11] described an association with familial aortic dissection. Sallo and Hatvani [12] reported four cases of primary pupillary IPE cysts in a single family. These patients also suffered from transient vision disturbances. Our patient, however, had no history of associated systemic diseases and no other member of his family had a cyst. Primary IPE cysts have often been misdiagnosed as iris or ciliary body melanoma [13]. Cysts of the iris are more likely to be confused with melanoma than those of ciliary body because of their pigmentation and more anterior location. Diagnosis and monitoring of iris, ciliary body and posterior chamber cysts require the imaging of structures not always visible with a slit lamp and goniolens, even through a dilated pupil. UBM has thus become indispensable for evaluating anterior segment tumors and cysts and is used to confirm a clinical diagnosis of primary IPE cysts, differentiate them from cystic and solid tumors (iris-ciliary body melanoma and melanocytoma, adenoma of the IPE and medulloepithelioma), follow their course and promptly intervene if surgery is required. It also provides cross-sectional images illustrating the surface contour, internal reflectivity and borders, and reveals their size, extension and site. Management is determined by a cyst's site, size, local extent and growth pattern, and the presence of secondary complications. Most primary IPE cysts run a benign clinical course that seldom results in ocular complications. They are usually small and frequently regress spontaneously. Those that arise from the IPE layers are stationary, asymptomatic, and detected only incidentally. Shields believes that most primary IPE cysts are ophthalmic curiosities that need prolonged observation and no treatment [1]. A steadily growing cyst, however, may disturb vision by covering the visual axis and provoke an increase in intraocular pressure or even inflammation if it touches the corneal endothelium. Steps must then be taken to prevent or treat complications, such as pupillary obstruction, secondary glaucoma, iridocyclitis, corneal decompensation and loss of vision. Numerous forms of managements have been employed. Mitomycin injection into the cyst and needle aspiration with endodiathermy were proposed by Kawaguchi *et al.* [14] and Tsai *et al.* [15]. Kuchenbecker *et al.* [16], Öner *et al.* [17] and Baykara *et al.* [18] prefer Nd:YAG laser cystotomy as the least invasive procedure. Excision in our case, like in that reported by Verma *et al.* [19], was dictated by the fact that in strong daylight the cyst obscured the visual axis almost completely and caused a severe temporary decrease of visual acuity. There were no complications and no clinical or UBM evidence of recurrences or fresh lesions during a one-year follow-up.

In conclusion, in agreement with most authors we believe that stable and asymptomatic primary IPE cysts usually require no more than regular observation. Proper treatment depends on the overall clinical picture, which includes both the patient's general health and the size and location of the cyst. Lastly, the importance of early diagnosis should be stressed. UBM is a useful, readily available and noninvasive tool for this purpose, since it can be employed to both confirm the clinical diagnosis and follow the course of these cysts. We address future studies on anterior chamber OCT to "*in vivo*" analyze cyst morphology.

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