Clinical evaluation of primary peripheral cysts of the iris

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ABSTRACT

Background: Peripheral primary cyst of the iris is an uncommon benign condition that may masquerade as a tumor of the ciliary body. Careful examination utilizing the slit lamp and gonioscopy is necessary to successfully differentiate the two.

Conclusions: Given their uncommon occurrence and the importance of ruling out melanoma or other malignancies, this review is a guide in the evaluation and diagnosis of patients with this condition.

KEY WORDS: iris cyst, iris neoplasms, ciliary body neoplasms, gonioscopy

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Primary iris cysts are usually asymptomatic and are typically discovered on routine biomicroscopic examination as an ominous mound in the peripheral iris. Given their uncommon occurrence and the importance of ruling out melanoma or other malignancies, we have prepared this review as a guide in the evaluation and diagnosis of patients with this condition.

Iris cysts may be classified by etiology as being either primary or secondary. Primary cysts have no recognizable etiology, whereas secondary cysts can be attributed to a specific cause, such as surgical trauma, nonsurgical trauma, or miotic drugs. Primary iris cysts are almost always intraepithelial; the result of an incomplete closure or re-establishment of the space between the two epithelial layers of the iris.[1] Rarely, primary iris cysts may occur within the iris stroma. Secondary cysts vary widely in nature, but two common forms are epithelial downgrowths from the cornea that may occur following intraocular surgery, and cysts at the pupillary margin that may develop after prolonged miotic therapy.

Clinical characteristics of primary iris cysts

Primary iris cysts are of two types: cysts of the iris pigment epithelium and cysts of the iris stroma (Table 1).[1] Intraepithelial cysts may be classified based on their location within the eye: 1) cysts of the pupillary margin, 2) cysts in the midzone of the iris, 3) cysts in the irido-ciliary sulcus, 4) free-floating anterior chamber cysts, and 5) free-floating vitreous cysts. Iris stromal cysts may be classified as being either congenital or acquired.

Cysts of the pupillary margin

These congenital cysts may be unilateral or bilateral. Bilateral cysts tend to be multifocal and familial. An autosomal dominant mode of inheritance is suspected.[2,3] Unilateral cysts may be solitary or multiple, and appear to occur spontaneously. The lesions can be easily detected without pupillary dilation as densely pigmented globular protuberances at the pupillary margin. It is believed that the cysts represent a failure of closure of the anterior portion of the primitive optic cup (annular sinus of von Szily) during the development of the eye.[1] Secondary pupillary cysts are associated with the use of anticholinesterase miotics (echothiophate), although this can be prevented by the concomitant use of phenylephrine.[4] The cysts are asymptomatic and nonprogressive. Many cysts will rupture spontaneously or as a result of ocular trauma, leaving strands of pigment epithelium hanging from the pupillary margin (iris flocculi).

Midzonal iris cysts

Cysts that are located in the pigment epithelium between the pupil margin and the root of the iris are classified as midzonal.[1] Such cysts are relatively uncommon. They may be unilateral or bilateral, soli-

Table 1: Classification of iris cysts

- I. Primary cysts
  - A. Cysts of iris pigment epithelium
    1. Central (pupillary)
    2. Midzonal
    3. Peripheral
    4. Dislodged (free floating)
       a. Anterior chamber
       b. Vitreous chamber
  - B. Cysts of the iris stroma
    1. Congenital
    2. Acquired
- II. Secondary cysts
  - A. Epithelial
    1. Epithelial downgrowth cysts
    2. Pearl cysts
    3. Drug-induced cysts
  - B. Cysts secondary to intraocular tumors
    1. Medulloepithelioma
    2. Uveal melanoma
  - C. Parasitic cysts

tary or multiple. Midzonal cysts can be easily seen with pupil dilation. They appear as a rounded dark mass just posterior to the pupillary border. With a fully dilated pupil the cysts become stretched and everted so that they take on an elongated or fusiform shape resembling a ciliary body melanoma.[1] They do not transmit light because they arise from the densely pigmented epithelium of the iris, but occasional focal areas of pigment loss can be found on their surface. The cysts may demonstrate tremulous motion with eye movements.

Peripheral iris cysts

Cysts occurring in the irido-ciliary sulcus are by far the most common of all primary iris cysts, accounting for about three-fourths of all cases.[1] They are stable, nonprogressive lesions that rarely produce symptoms or complications of any sort. Multiple large cysts, however, are capable of producing secondary angle-closure glaucoma.[5,6] The lesions are characteristically unilateral and solitary. They are usually found in the infero-temporal quadrant; especially between 7:00 and 9:00 o'clock in the right eye and between 3:00 and 5:00 in the left eye. In one series of 46 eyes with solitary peripheral iris cysts, 39 (85 percent) of the lesions were located temporally, and of those 33 (85 percent) were located inferiorly.[1] The cysts are three times more common in women than men, and are most commonly diagnosed in the third and fourth decades of life.

The lesions are most often detected clinically as a localized anterior displacement of the iris stroma on routine slit lamp examination. The overlying iris stroma is usually normal. Peripheral iris cysts can be very difficult to visualize and may require maximal pupil dilation and use of a gonioscopy lens before the lesion can be seen. In contrast to pupillary and midzonal cysts, peripheral cysts transmit light readily. This is because they arise from nonpigmented ciliary epithelium.[1] The transparent walls of the cyst will often permit visualization of the ciliary processes through the walls of the lesion.

Free-floating cysts

Iris cysts that have become detached from the iris may move into either the anterior or, rarely, the vitreous chamber. Cysts in the anterior chamber will usually settle into the chamber angle and may give the appearance of a pigmented tumor of the peripheral iris or possibly an intraocular foreign body.[7] Differentiation is aided by recognizing that cysts have sharp abrupt borders unlike the gradual sessile borders of an iris melanoma.[8] The cyst may move about in response to changes in head posture. Rarely, this may interfere with vision and require surgical removal of the cyst.[9] Iris cysts that are displaced into the vitreous chamber become suspended in the vitreous gel and take on the characteristics of a large vitreous floater. A relationship between vitreous cysts and retinitis pigmentosa has been reported.[10]

Cysts of the iris stroma

Stromal cysts involve the iris tissue anterior to the pigment epithelium. They are rare and usually congenital.[11] Acquired stromal cysts appear spontaneously or, more commonly, result from trauma or inflammation. Stromal cysts have a nonpigmented anterior wall that may contain blood vessels. Congenital and acquired stromal cysts will often progressively enlarge over time and may eventually require surgical intervention.[11,12] In contrast, intraepithelial cysts tend to be stationary lesions that rarely progress and may even become smaller or disappear altogether.[1]

Clinical evaluation of peripheral iris cysts

Differential diagnosis

Ruling out the presence of an intraocular tumor is an important part of the diagnostic work-up of patients presenting with iris cysts. The differential diagnosis of primary iris cysts is summarized in Table 2.

Because of their location and characteristic appearance, cysts of the pupillary margin rarely present a diagnostic challenge. Midzonal and peripheral cysts, on the other hand, may resemble a melanoma or other tumor. In one series of 158 suspicious iris lesions (pseudomelanomas) primary iris cyst was the most common diagnosis, accounting for 61 (38.6 percent) of the lesions.[13]

Table 2: Differential Diagnosis of Primary Iris Cysts

I.	Central cysts
	Drug-induced cysts
II.	Midzonal and peripheral cysts
	Melanoma
	Leiomyoma
	Medulloepithelioma
	Medanocytoma
III.	Dislodged cyst in chamber
	Nevus
	Melanoma
	Foreign body
IV.	Dislodged cyst in vitreous chamber
	Cysticercosis
	Retinitis pigmentosa
	Persistent posterior hyaloid
V.	Stromal cysts
	Melanoma
	Metastatic carcinoma
	Secondary cysts

Iris tumors actually arise from the iris stroma and do not simply displace it anteriorly, as do iris cysts.[8] The stroma overlying an iris cyst is normal or only slightly thinned. In the case of an iris tumor, such as a melanoma, a distinct lesion involving the iris stroma can be detected on routine slit lamp examination.

Tumors of the ciliary body, however, present a diagnostic challenge since they often will present as mass lesions behind the iris. Careful examination, as described below, is necessary to differentiate tumors of the ciliary body from peripheral iris cysts.

Diagnostic techniques

The clinical evaluation of patients presenting with an elevation in the peripheral iris is primarily directed at differentiating iris cysts from tumors of the ciliary body. The clinical work-up described below is adapted from Shields.[14] It lists the recommended steps in making this differentiation.

External examination: Ciliary body melanomas will often produce engorgement of the overlying episcleral blood vessels. Presence of episcleral injection in the quadrant containing the lesion should lead one to suspect the presence of a tumor.

Pre-dilation slit lamp examination: Note whether the iris stroma in the region of the lesion is disorganized or abnormally pigmented. The iris stroma overlying an iris cyst is essentially normal, whereas all forms of iris neoplasia will cause stromal architecture or pigmentary changes to occur (Fig. 1). Also bear in mind that more than 70 percent of peripheral cysts occur in the infero-temporal quadrant. Lesions located elsewhere should be viewed with suspicion.

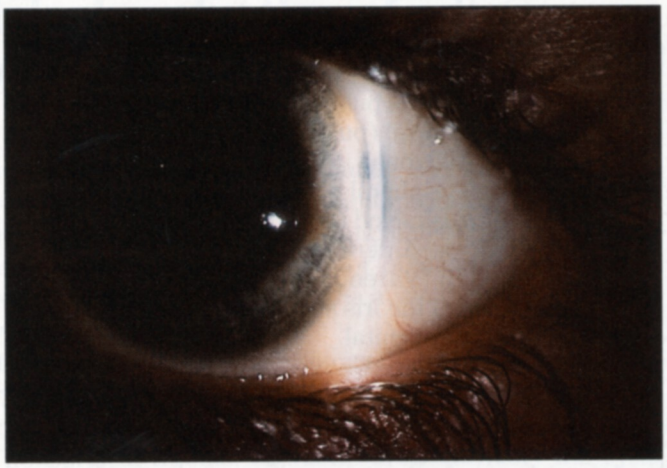

Figure 1: Appearance of peripheral iris cyst on routine slit lamp examination. Peripheral cysts are often discovered during routine anterior chamber depth assessments. Note the shallowing of the anterior chamber inferiorly.

Post-dilation slit lamp examination: With a widely dilated pupil the slit lamp is used to visualize the lesion in the posterior chamber. This requires that the pupil be so widely dilated that a sizable gap exists between the iris and lens through which one can examine the lesion. If the pupil is not sufficiently dilated, a cotton pledget saturated with tropicamide can be placed in the inferior fornix for 5 minutes to achieve maximal dilation.

The patient's gaze is then directed into the affected quadrant and the slit lamp arm and oculars are rotated to the opposite side. With this technique the cyst can be directly visualized in about 80 percent of cases (Fig 2a). The transparent walls of the cyst usually permit one to see the ciliary processes directly through the lesion. In some cases the walls of the cyst are so thin that the lesion becomes almost invisible.

To confirm the cystic nature of the lesion, an attempt should be made to retroilluminate it (Fig. 2b). To perform this maneuver shine the light beam off to the side of the lesion rather than directly on it. Arrange the lighting in such a way that the lesion is backlit by light reflecting off the ciliary body or fundus. If the lesion is a cyst it will glow. A solid mass will appear as a dark shadow. Ciliary body melanomas are solid, but certain tumors, such as medulloepitheliomas, can be quite cystic. In these cases, however, the fleshy ciliary body tumor associated with the cysts can usually be seen.

Gonioscopy: Once an attempt has been made to see the lesion with the slit lamp, a gonioscopy lens is placed on the eye. Generally speaking, the gonioscopy lens will afford a better view of the lesion (Fig. 3a and b). Again, the lesion should be directly seen and retroilluminated.

Gonioscopy also permits examination of the anteri-

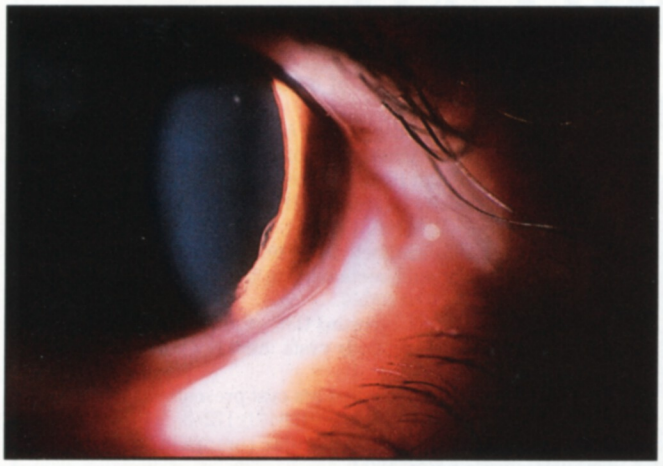

Figure 2a: Post-dilation view of a peripheral iris cyst. This large cyst can be easily seen.

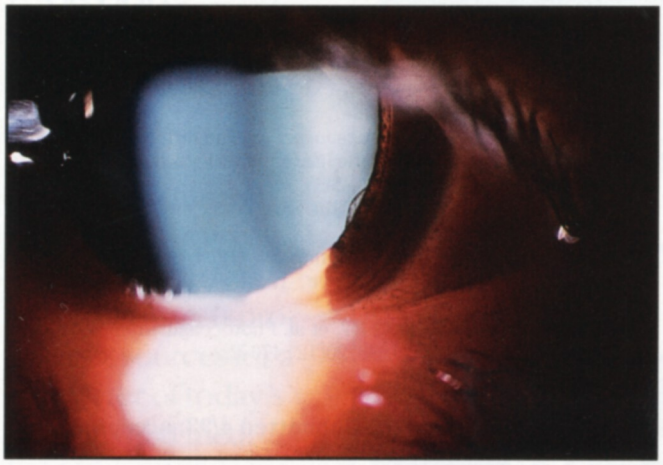

Figure 2b: Post-dilation view of a peripheral iris cyst. This large cyst can be easily retroilluminated without the aid of gonioscopy.

Figure 3a: Appearance of a peripheral iris cyst through the gonioscopy lens. Note the prominent anterior displacement of the iris.

Figure 3b: Appearance of a peripheral iris cyst through the gonioscopy lens. Note the visualization of the ciliary processes through the walls of the cyst.

or chamber angle where the root of the iris and face of the ciliary body are checked for signs of invasion by a ciliary body tumor.

Very rarely, a cyst cannot be seen and the diagnosis must remain presumptive. The typical anterior displacement of the iris stroma in the infero-temporal quadrant of a young adult woman without any evidence of ciliary body tumor, however, provides strong support for the diagnosis.[1]

Ancillary tests: If the above attempts to retroilluminate the lesion have failed, then an external transilluminator may be utilized. This involves viewing the lesion through the slit lamp (with or without a gonioscopy lens) and then placing a transilluminator on the limbus in the quadrant of the lesion. The light of the slit lamp is then turned off and the lesion is observed for transillumination.

Another technique is to transilluminate the globe by placing a fiber optic transilluminator in the fornix 180° from the lesion. The patient then looks in the direction of the transilluminator probe and the shadows of the ciliary body are observed in the quadrant of the lesion. If the pars plana transmits light normally then it is unlikely that a more posterior ciliary body tumor exists.

Indirect ophthalmoscopy should be performed in all cases to rule out the presence of a tumor invading the peripheral choroid and to detect other fundus disease.

Clinical management of primary iris cysts

Once the diagnosis of primary iris cyst has been made the patient is monitored on a periodic basis for any enlargement of the lesion. Because of the nonprogressive nature of intraepithelial cysts, treatment is rarely warranted. Stromal cysts are more prone to enlargement and, hence, more often require intervention. The indications for treatment, as proposed by Belcher et al.,[15] are as follows: 1) Secondary angle-closure glaucoma; 2) Obstruction of the visual axis or other visual symptoms; 3) Contact with the corneal endothelium causing corneal decompensation, an inflammatory reaction, or both.

Whenever treatment is indicated, the treatment of choice is laser photocoagulation with either the argon or neodymium-YAG laser.[15,16] The alternatives include needle aspiration with or without subsequent cautery, diathermy, or injection of sclerosing agents into the cyst; marsupialization; sector iridectomy; and cryotherapy. With the exception of cryotherapy, they are all invasive and therefore carry the risk of significant complications. Photocoagulation is a simple, effective technique that is remarkably free of complications.[15] However, because stromal cysts will often recur after conservative therapy such as photocoagulation or aspiration, surgical removal of stromal cysts is frequently necessary.[11]

Summary

Peripheral primary cyst of the iris is an uncommon benign condition that may masquerade as a tumor of the ciliary body. Careful examination utilizing the slit lamp and gonioscopy is necessary to successfully differentiate the two conditions. ■

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