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Major review

Iris cysts: A comprehensive review on diagnosis and treatment

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ABSTRACT

Iris cysts, both primary and secondary, are a diagnostic and treatment challenge. Primary cysts arise either from the iris pigment epithelium or the iris stroma. Posterior pigment epithelial cysts are subdivided according to their location as central, midzonal, and peripheral. Iris stromal cysts are classified either as congenital or acquired. Free-floating cysts are usually dislodged pigment epithelial cysts. Secondary cysts are classified according to the underlying cause as implantation cysts, drug-induced, uveitic, tumor-induced, parasitic, or as cysts associated with systemic disorders. Differential diagnosis is based on the clinical presentation and imaging. Ultrasound biomicroscopy is the gold standard for the imaging of iris cysts, combining excellent resolution with sufficient tissue penetration. Treatment of iris cysts depends largely on whether they become symptomatic or not. Symptoms include obstruction of the visual axis, corneal decompensation, secondary uveitis, and secondary glaucoma. Treatment options cover a range from simple observation to fine-needle aspiration (with or without intracystic injection of absolute alcohol or antimitotic agents), laser (argon, Nd:YAG), or surgical excision. In the past, the prevailing notion was that of a radical surgical intervention in the form of iridectomy or iridocyclectomy. Given the high rate of recurrence, a stepwise conservative approach is currently favored by most clinicians.

1. Introduction

The first report of an iris cyst dates back to 1830, when MacKenzie first described a posttraumatic cyst in the anterior chamber.[111] Although uncommon, iris cysts present as a diagnostic and treatment challenge. Iris cysts can be classified in many ways, but the main classification remains that of primary and secondary cysts proposed by Shields in 1981.[162,165] These major categories are further subcategorized based on the tissue of origin (Table 1).[162,168] Primary cysts are of neuroepithelial origin, whereas secondary cysts are the result of implantation, metastasis, long-term use of miotics, or parasites. Primary

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Table 1 – Shields classification of iris cysts*

Primary cysts		
Iris pigment epithelium cyst		Iris stroma cyst
i. Pupillary ii. Midzonal iii. Peripheral iv. Dislodged v. Free-floating 1. Anterior chamber 2. Vitreous chamber		i. Congenital ii. Acquired
Secondary cysts		
Epithelial	Cyst secondary to intraocular tumors	Parasitic cyst
i. Epithelial downgrowth cyst 1. Postsurgical 2. Posttraumatic ii. Pearl cyst iii. Drug-induced cyst	i. Medulloepithelioma ii. Uveal melanoma iii. Uveal nevus	

* Modified from Shields et al.[162]

cysts only seldom cause secondary complications in cases of enlargement. Secondary cysts have a tendency to cause complications such as decreased visual acuity, secondary glaucoma, corneal edema, or uveitis. Implantation cysts are caused by the deposition of surface epithelial cells from the surrounding tissues (cornea, conjunctiva, or even the eyelid) on the iris after penetrating or surgical trauma. Implantation can result in the formation of solid pearl lesions, epithelial fluid-filled cysts, or epithelial ingrowth.

Clinicians should also differentiate iris cysts from other entities such as iris or ciliary body melanoma. Clinical examination and ultrasound biomicroscopy (UBM) form the basis for the correct diagnosis.

Although pathogenic mechanisms have been largely elucidated, there is still controversy regarding the most appropriate treatment, with a shift toward a more conservative stepwise approach. Treatment options include observation of stationary cysts, fine-needle aspiration (FNA) and subsequent infusion of alcohol or antimitotic agents, the use of laser (either argon or Nd:YAG), and usually as a last resort, surgical excision.

In the following section, we review critically and in detail the available literature on iris cysts regarding both their diagnosis and treatment.

2. Brief overview of iris anatomy and embryology

The iris consists of 2 layers; the stroma anteriorly, behind which lies the pigment epithelium.

Iris stroma comprises highly vascular connective tissue that contains collagen fibers, fibroblasts, intracellular matrix, melanocytes, and occasional mast cells. The anterior stromal surface is bare of epithelium, covered by a densely arranged layer of fibroblasts, melanocytes, and collagen fibers called the anterior border layer. Entrapment of surface ectodermal cells into the iris stroma during embryogenesis gives rise to primary cysts that grow inside the iris tissue, the so called stromal cysts (congenital or acquired). Classification and pathogenesis are discussed in detail in Sections 3.1 and 3.3, respectively.

The posterior layer, which is the iris pigment epithelium, is composed of 2 layers of epithelial cells juxtaposed apex to apex. The anterior epithelial layer that lies in contact with the iris stroma contains a few melanin granules and is continuous with the outer pigmented layer of the ciliary body epithelium. The posterior epithelial layer cells are larger and contain numerous melanin granules. This layer is continuous with the inner nonpigmented layer of the ciliary body epithelium.[177] In the case of separation of the epithelial layers of the iris, a primary cyst lined by epithelium may form. Depending on the exact site of origin, these cysts present variable morphological characteristics. Classification and pathogenesis are discussed in detail in Sections 3.1 and 3.3, respectively.

Embryologically, the iris muscles (both the sphincter and dilator), as well as the iris pigment epithelium, are of neuroectodermal origin. All other iris tissues and cells arise from the mesectoderm.[113,145]

3. Primary cysts

3.1. *Classification*

Primary cysts classification follows the pattern introduced by Shields et al.[165] They may arise either by the iris pigment epithelium or the Iris stroma.

3.1.1. *Posterior pigment epithelial cysts*

Cysts of the iris pigment epithelium can be divided according to their exact location as central, midzonal, or peripheral.

1. Central: Central cysts are located from the pupillary margin to the iris root.
2. Midzonal: Midzonal cysts are located from the iris root to the ciliary body.
3. Peripheral: Peripheral cysts are located at the iridociliary sulcus.

3.1.2. *Cysts of iris stroma*

Cysts of the Iris stroma are located anteriorly to the iris pigment epithelium and as they develop inside the iris they usually cause iris deformation.

1. Congenital: Usually unilateral and solitary cysts that present before 10 years of age and often require treatment, they represent more than 50% of stromal cysts. A case of congenital stromal cyst is presented in Figure 1A.
2. Acquired: Usually present later in life and only seldom require treatment, in contrast to the congenital stromal cysts.

3.1.3. *Free-floating/dislodged cysts*

Free-floating cysts in the anterior chamber or vitreous are usually dislodged pigment epithelium cysts. A case of dislodged cyst into the vitreous cavity is presented in Figure 2.