

## **Implantation pearl cyst after uncomplicated clear cornea phacoemulsification mimicking an iris tumor**

---

Sara Vaz-Pereira, MD<sup>1,2</sup>, Gabriella De Salvo, MD<sup>3</sup>, Lamis Al Harby, MD<sup>4,5</sup>, Caroline  
Thaung, FRCOphth, FRCPath<sup>4,6</sup>, Mandeep S. Sagoo, MB, PhD, FRCS (Ed),  
FRCOphth<sup>4,5,6</sup>

<sup>1</sup>Department of Ophthalmology, Hospital de Santa Maria, Lisbon, Portugal

<sup>2</sup>Department of Ophthalmology, Faculty of Medicine, Universidade de Lisboa,  
Lisbon, Portugal

<sup>3</sup>Southampton Eye Unit, University Hospital Southampton Foundation Trust,  
Southampton, UK

<sup>4</sup>Moorfields Eye Hospital NHS Foundation Trust, London, UK

<sup>5</sup>Ocular Oncology Service, St. Bartholomew's Hospital, London, UK

<sup>6</sup>UCL Institute of Ophthalmology, London, United Kingdom

This work was carried out at the Oncology Clinic, Moorfields Eye Hospital NHS  
Foundation Trust, London, UK.

Presented as a poster at the 5th EUCORNEA Congress, 12/09/2014 - 13/09/2014,  
London, UK and as a case presentation on 18 June 2015, at the International  
Society of Ocular Oncology's 2015 Biennial Meeting in Paris, France.

The work described has not been published previously and is not under consideration for publication elsewhere.

**Correspondence and reprint requests:**

Mandeep S. Sagoo, Moorfields Eye Hospital NHS Foundation Trust, 162 City Road, London EC1V 2PD, United Kingdom. Telephone: (+44) 20 7253 3411. Fax: (+44) 20 7253 4696. Email: mandeep.sagoo@moorfields.nhs.uk

**Conflicts of interest and sources of funding:**

None declared. None of the authors has conflicts of interest with the submission. The authors have no proprietary interest in the subject or materials discussed in this manuscript. No funding was received.

**Authorship:**

All authors contributed and approved the final version of this publication.

SVP - (1) conception and design of the study, acquisition of data, analysis and interpretation of data, (2) drafting the article and revising it critically, (3) final approval.

GDS – (1) conception and design of the study, analysis and interpretation of data, (2) drafting the article and revising it critically, (3) final approval.

LAH – (1) acquisition of data, (2) critical revision of manuscript, (3) final approval.

CT – (1) acquisition of data, analysis and interpretation of data, (2) critical revision of manuscript, (3) final approval.

MSS – 1) conception and design, acquisition of data, analysis and interpretation of data, (2) drafting the article and revising it critically, (3) final approval.

**Short title:** Pearl cyst after phacoemulsification

**Key words:** anterior uveitis, clear cornea phacoemulsification, epithelial downgrowth, iris pearl tumor

**Word count: 997**

## Introduction

---

Epithelial downgrowth (ED) is a serious albeit rare complication of ocular surgery and trauma which can result in irreversible vision loss.<sup>1-6</sup> In the ocular surgery group, cataract surgery especially in the extracapsular era accounted for 86% of cases,<sup>1</sup> related to vitreous loss, persistent hypotony, capsule rupture, multiple surgeries, delayed wound healing, wound fistulas and iris incarceration. According to the literature, 82% of cases presented within the first year after intraocular surgery<sup>1,3</sup> with retrocorneal membrane (48%), painful glaucoma (32%), positive Seidel test (24%), corneal edema (22%), hypotony (17%), painless glaucoma (14%), iritis (11%) and iris cyst (8%).<sup>1</sup>

Epithelial cells that gain access to the anterior chamber can grow and present into 3 classical forms: 1) opaque sheet over the cornea, iris, and angle structures, 2) epithelial anterior chamber clear cyst or rarely 3) as a pearl white solid cyst, also referred as iris pearl tumor.<sup>2-4</sup> Histologically, they are arranged in a multilayer structure resembling corneal or conjunctival epithelium.<sup>1,2</sup> The sheet-like form is the commonest presentation and has the worse prognosis, followed by the serous and pearl cysts.<sup>2,4,5,7</sup> The serous cysts are transparent and often form in a wall of conjunctiva covered by epithelium, while pearl cysts are solid, opaque, and most commonly associated with cilia that enter the anterior chamber.<sup>7,8</sup> Because of their potential of growth and development of serious complications, most authors favor a surgical approach.<sup>8</sup>

We report a case of ED presenting as an iris pearl tumor two years after uncomplicated clear cornea phacoemulsification.

## Case report

---

A 71-year-old male presented with a three-month history of a red, painful, and photophobic left eye (OS). His ocular history was significant for bilateral uncomplicated clear corneal phacoemulsification with posterior chamber intraocular lens (PC-IOL) implantation two years before and his family and medical history were unremarkable. There was no history of trauma. Visual acuity (VA) was 6/9 in the right eye (OD) and 6/18 OS. OS biomicroscopy showed cilio-conjunctival hyperemia, +4 anterior chamber cells and a vascularized dome-shaped amelanotic iris mass at the 12 o'clock position, with a splashing of pigment over the surface with a slither hypopyon (Fig.1A-D). There was also a PC-IOL and there was no evidence of ED from the section site of his previous cataract surgery. No vitritis was identified and fundoscopy did not reveal any signs of posterior inflammation. Ultrasound B-scan detected an iris lesion at 12 o'clock with medium internal echogenicity and an elevation of 3.0 mm (Fig.2A) and anterior segment optical coherence tomography confirmed these measurements (Fig.2B). Treatment with topical steroids led to an improvement in inflammation, but not the mass dimensions. Uveitis workup was negative so an excisional biopsy by broad iridectomy was performed. Microbiology analysis of the aqueous aspirate was negative. Histopathology findings were of iris tissue partly covered by stratified squamous epithelium reminiscent of ocular surface epithelium.

The overall appearance was suggestive of ED (Fig.3). There was no evidence of neoplasia.

The eye settled well on topical medication and at 24 months follow-up, VA was 6/9 and there was no evidence of inflammation or recurrence (Fig.4A-C).

## Discussion

---

The incidence of ED after cataract surgery has been reported to range from 0.08%-0.12%<sup>1,2</sup> when intracapsular and extracapsular extraction techniques were used. With the advances in cataract surgery, sutureless phacoemulsification has become the procedure of choice for most surgeons. It allows for a smaller wound with better closure which results in better visual acuity with a lower rate of complications,<sup>2,9,10</sup> thus rendering ED very rare.<sup>2</sup> There are sparse reports in the literature on this subject over the past 20 years.<sup>2,4,6,10,11</sup> Of these, only Knauf et al<sup>10</sup> reported the cystic form, namely the serous cyst, presenting after 3 years of cataract surgery and managed successfully by surgical excision. To the best of our knowledge there have been no reports of the pearl cyst formation after uncomplicated clear cornea phacoemulsification. In general, the cystic forms have been considered more easily managed due to prompt detection and better surgical eradication.<sup>2,5</sup> Other reported successful treatment options for implantation anterior chamber cysts have included aspiration of cyst contents and endolaser photocoagulation of the residual cyst wall, aspiration and intralesional administration of mitomycin C or more invasive vitrectomy and local cryoablation of adjacent cyst wall.<sup>3,5,12</sup> The sheet-like form has been more often reported<sup>2,4,11</sup> and has been found to have the worst outcome as it is more difficult to diagnose and eradicate.<sup>2-4</sup> Extensive disease was also associated with a poorer outcome.<sup>4</sup>

The current case describes the uncommon pearl cyst presentation of ED, which occurred more frequently following trauma,<sup>7</sup> but uniquely now has been found after uncomplicated phacoemulsification. It presented as a solid iris lesion, as

documented by imaging, suggesting a neoplastic process. The natural sequence of events has been described following implantation of cilia into the iris after trauma: formation of the pearl cyst, followed by iridocyclitis and then enlargement of the cyst to cause secondary glaucoma.<sup>7</sup>

Other forms of ED can be associated with tumors. Stone et al<sup>4</sup> reported a case of pseudophakic chronic aggressive inflammation that later was associated with the development of a vascularized infiltrative iris mass, which was proved to be a metaplastic ED of a limbal squamous cell carcinoma with secondary intraocular inflammation. Therefore, malignant and other etiologies of iris cysts such as congenital cysts, gummas, tubercles, should be included in the differential diagnosis.<sup>7,8</sup>

Surgical treatment was the treatment of choice and during follow-up, no recurrence was identified. This is in accordance with previous reports that show that in patients with ED surgical treatment led to fewer enucleations than in those treated medically or not treated.<sup>1,2,4,7</sup>

In summary, ED is a very rare complication of modern cataract surgery. This case shows that, in this setting, in addition to the previously described serous cystic and sheet-like growth forms, epithelial growth can also present as an implantation pearl cyst. Clinicians should have a high index of suspicion for this condition in pseudophakic patients, when more frequent conditions have been excluded. Appropriate treatment includes early surgical excision as it may be associated with a more favorable prognosis.



## References

---

1. Weiner MJ, Trentacoste J, Pon DM, Albert DM. Epithelial downgrowth: a 30-year clinicopathological review. *Br J Ophthalmol*. 1989;73:6-11.
2. Vargas LG, Vroman DT, Soloman KD, et al. Epithelial downgrowth after clear cornea phacoemulsification: report of two cases and review of the literature. *Ophthalmology*. 2002;109:2331-2335.
3. Kim SK, Ibarra MS, Syed NA, Sulewski ME, Orlin SE. Development of epithelial downgrowth several decades after intraocular surgery. *Cornea*. 2005;24:108-9.
4. Srinivasan S, Jones DH, Jay JL, Roberts F. Epithelial downgrowth following clear cornea phacoemulsification in a buphthalmic eye. *Br J Ophthalmol*. 2004;88:152-3.
5. Haller JA, Stark WJ, Azab A, Thomsen RW, Gottsch JD. Surgical management of anterior chamber epithelial cysts. *Am J Ophthalmol*. 2003;135:309-13.
6. Stone DU, Char DH, Crawford JB, Margolis TP, Van Gelder RN, Strauss EC. Metaplastic squamous epithelial downgrowth after clear corneal cataract surgery. *Am J Ophthalmol*. 2006;142:695-7.
7. Sitchevska O, Payne BF. Pearl cysts of the iris. *Am J Ophthalmol*. 1951;34:833-40.
8. Song J, Bi H. Pearl cyst treatment by surgical excision. *Optom Vis Sci*. 2014;91:e294-7.
9. de Silva SR, Riaz Y, Evans JR. Phacoemulsification with posterior chamber intraocular lens versus extracapsular cataract extraction (ECCE) with

posterior chamber intraocular lens for age-related cataract. Cochrane Database Syst Rev. 2014;29;1:CD008812.

10. Knauf HP, Rowsey JJ, Margo CE. Cystic epithelial downgrowth following clear-corneal cataract extraction. Arch Ophthalmol. 1997;115:668-9.

11. Lee BL, Gatton DD, Weinreb RN. Epithelial downgrowth following phacoemulsification through a clear cornea. Arch Ophthalmol. 1999;117:283.

12. Yu CS, Chiu SI, Tse RK. Treatment of cystic epithelial downgrowth with intralésional administration of mitomycin C. Cornea. 2005;24:884-6.

## Figure legends

---

Figure 1 – External photograph of the left eye. (A) Image shows a vascularized dome-shaped amelanotic iris mass at the 12 o'clock position, with a splashing of pigment. (B) Magnified image of the lesion and cornea, where mild edema and keratic precipitates can also be identified. (C) Image shows cilio-conjunctival hyperemia with a discrete hypopyon. (D) Gonioscopic view of lesion showing an elevated lesion with no corneal contact. These features suggested a neoplastic process.

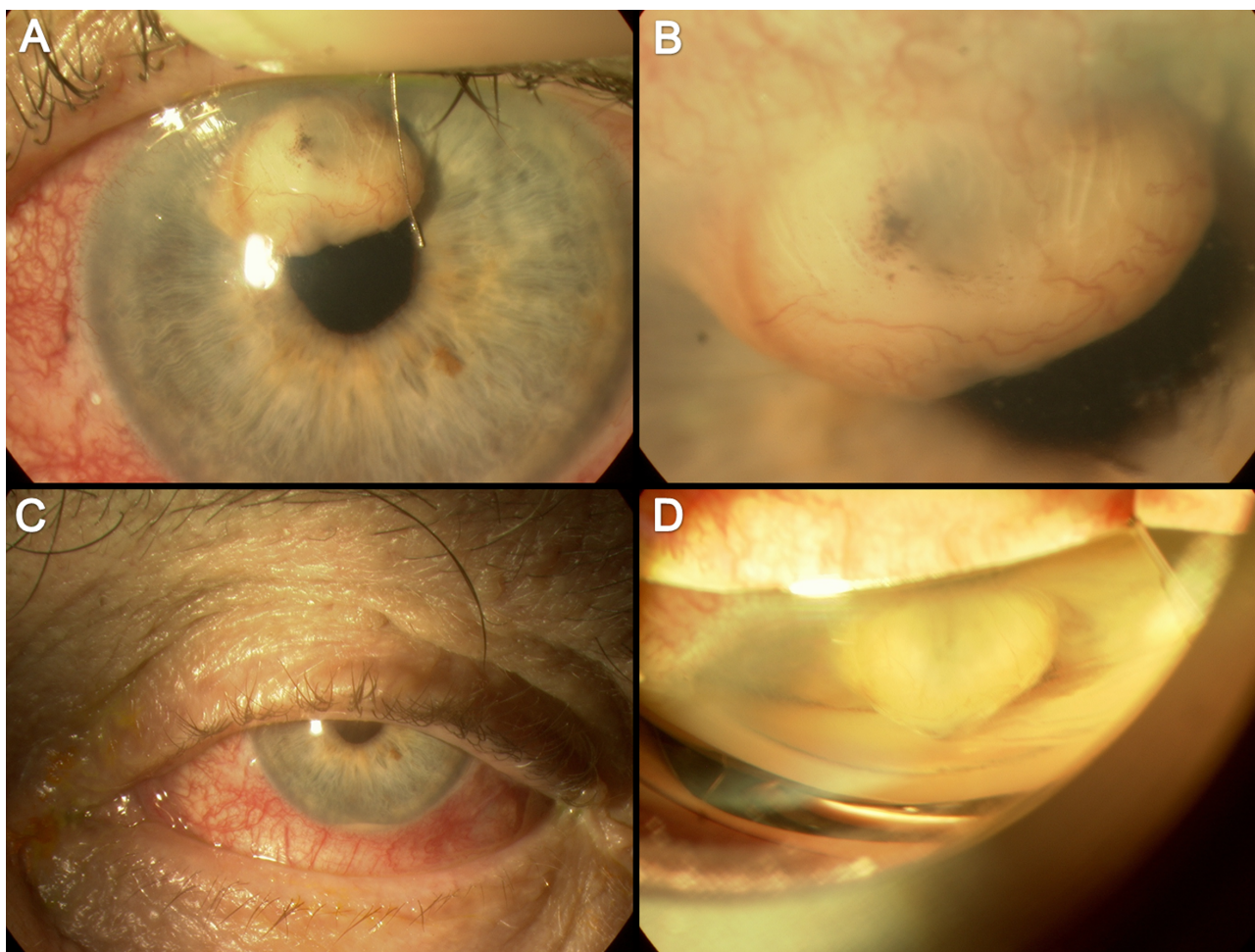


Figure 2 – (A) Ultrasound of left eye detected an elevated iris lesion at 12 o'clock with medium internal echogenicity, a transverse base of 4.8 mm, a longitudinal base of 3.2 mm and an elevation of 3.0 mm (B) Anterior segment optical coherence tomography confirmed ocular ultrasound measures.

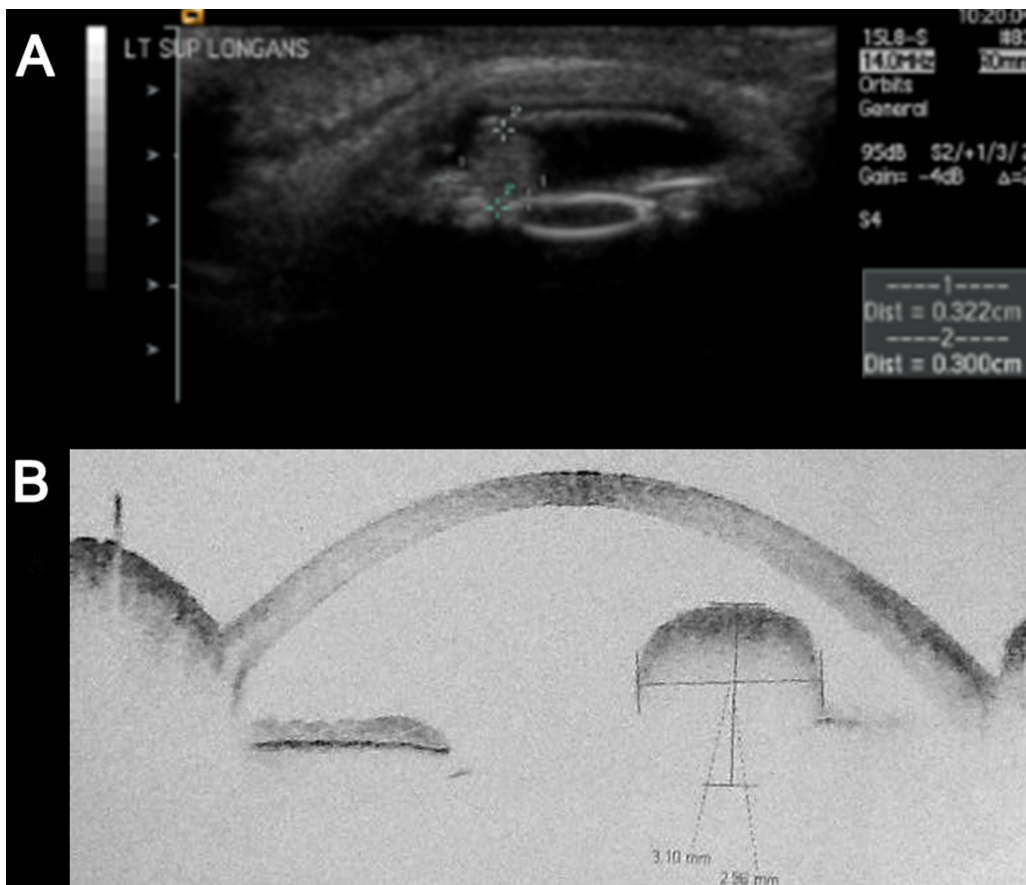


Figure 3 - Hematoxylin and eosin (x10 objective): cystic space containing keratin flakes and debris, lined by stratified squamous epithelium. The wall includes iris stroma and pigment epithelium.

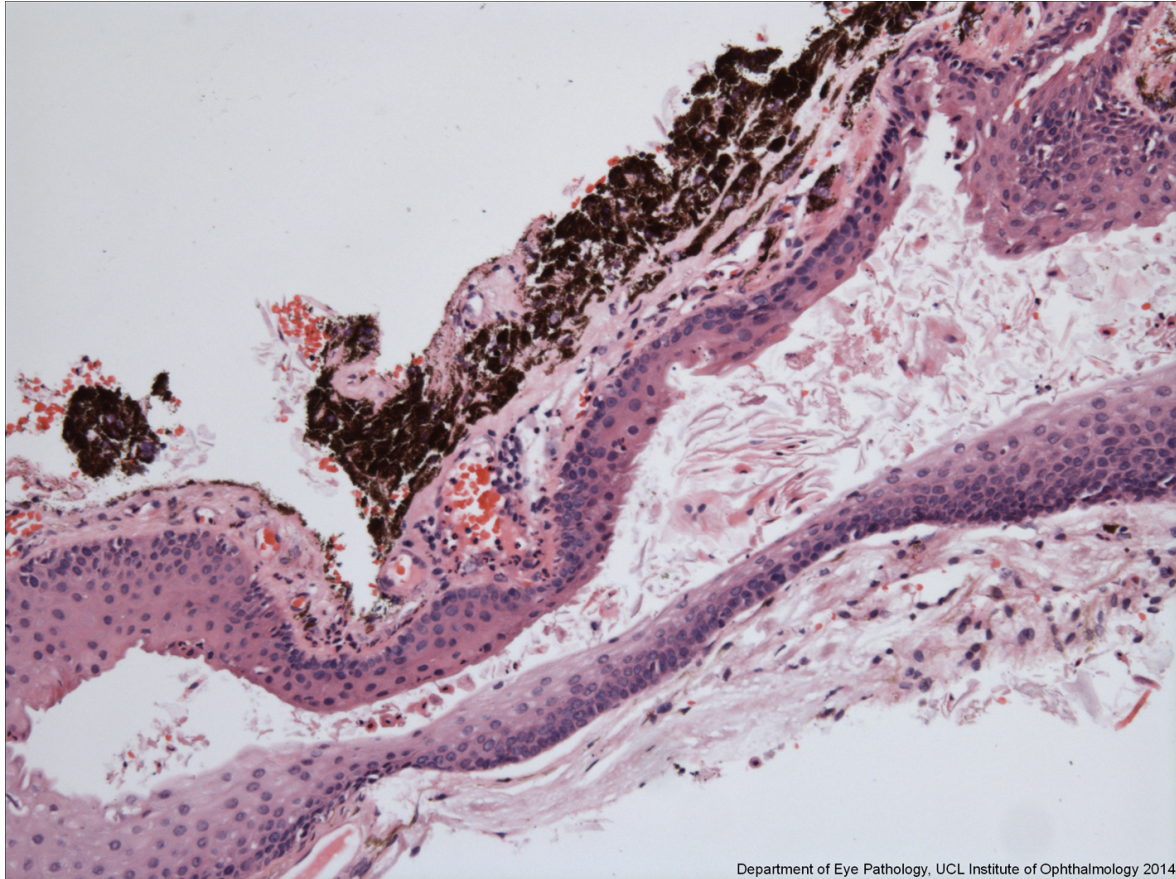


Figure 4 - Photographs of the left eye after broad iridectomy using different slit-lamp illumination techniques: (A) diffuse illumination (B) tangential illumination and (C) retro-illumination, which show a clear cornea, residual scar tissue at the capsulorhexis margin and no of recurrence of the lesion.

