



“Spontaneous” hyphema from a dislodged iris pigment epithelium (IPE) cyst

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INTRODUCTION

- Cystic lesions of the iris, although uncommon, have been well characterized and are classified according to the anatomic area of involvement.¹⁻⁵
- Spontaneous hyphema may occasionally be related to vascularized iris lesions.⁶
- Here, we present a unique case of a dislodged IPE cyst causing a hyphema.

CASE PRESENTATION

- A 78 year old man was doing yard work when he had an episode of painless white out of vision in his left eye. Over the ensuing hours, the eye became red and the vision returned, but was cloudy.
- There is no history of cancer, bleeding diathesis, ocular foreign body or aortic aneurysm.

OPHTHALMIC EXAM

	Right eye	Left eye
Visual acuity	20/25	20/40
IOP	12	24
Conj/Sclera	White and quiet	White and quiet
Cornea	Clear	Clear
Anterior Chamber	Deep and quiet	4+ RBC
Iris	Flat, round, prominent pupillary ruff/flocculae; prominent circumferential vessels	Flat, round, prominent pupillary ruff/iris flocculae; prominent bleeding vessel near margin
Lens	3+ NS, 2+ CS	3+ NS, 2+ CS
Gonioscopy OS only (Figure 2)	Inferior iris flocculus ~2mm in size w/ focal PAS and injected adjacent Schlemm's canal	

Ancillary testing

A stroke work-up was performed with the following findings:

- CT Head: unremarkable
- CT Orbits (w/ and w/o contrast): unremarkable
- CTA Head and Neck: <50% stenosis of bilateral carotid arteries

Several months earlier, the patient had a CTA of the thorax for chest pain, which was negative for an aortic aneurysm.

OPHTHALMIC IMAGING

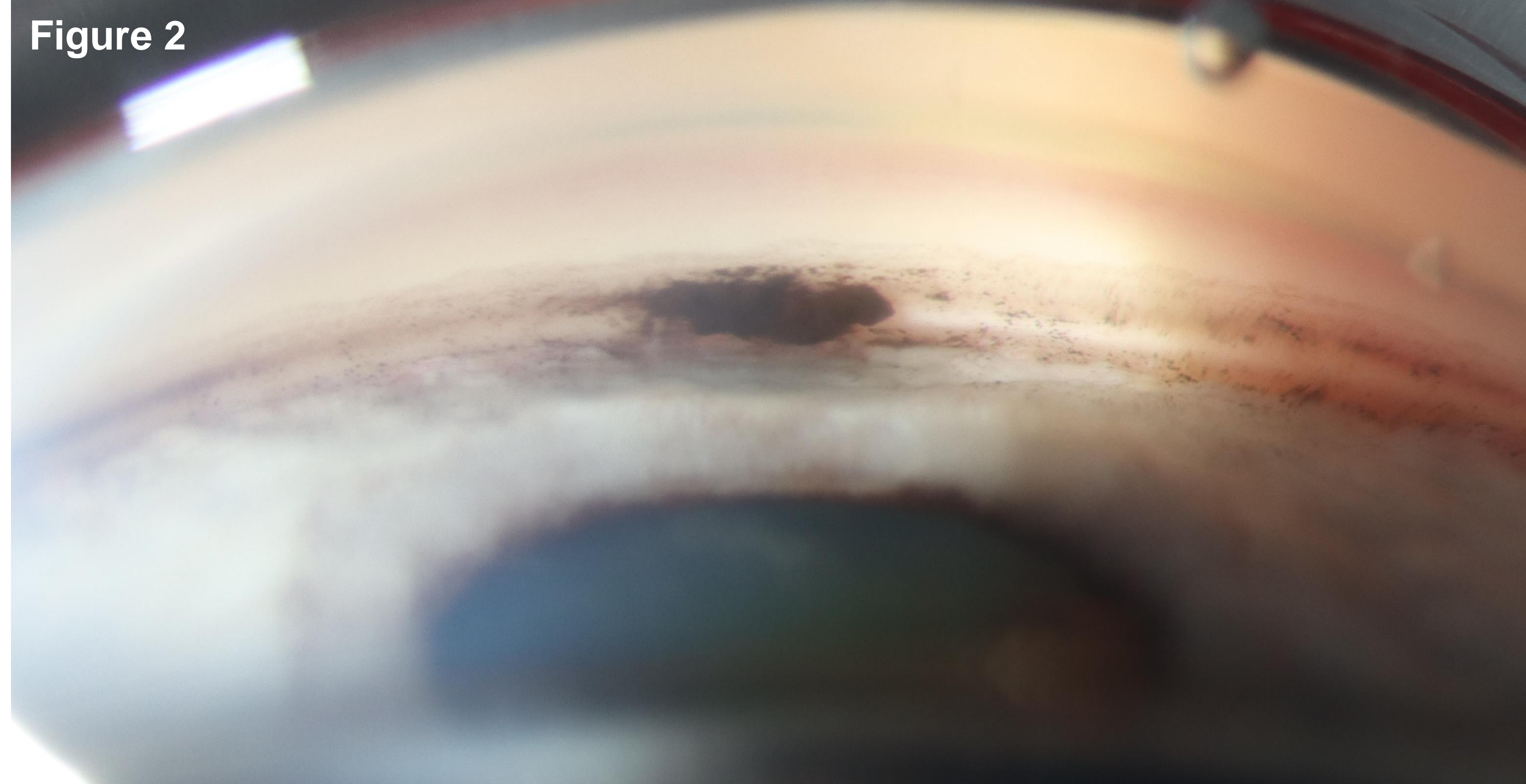
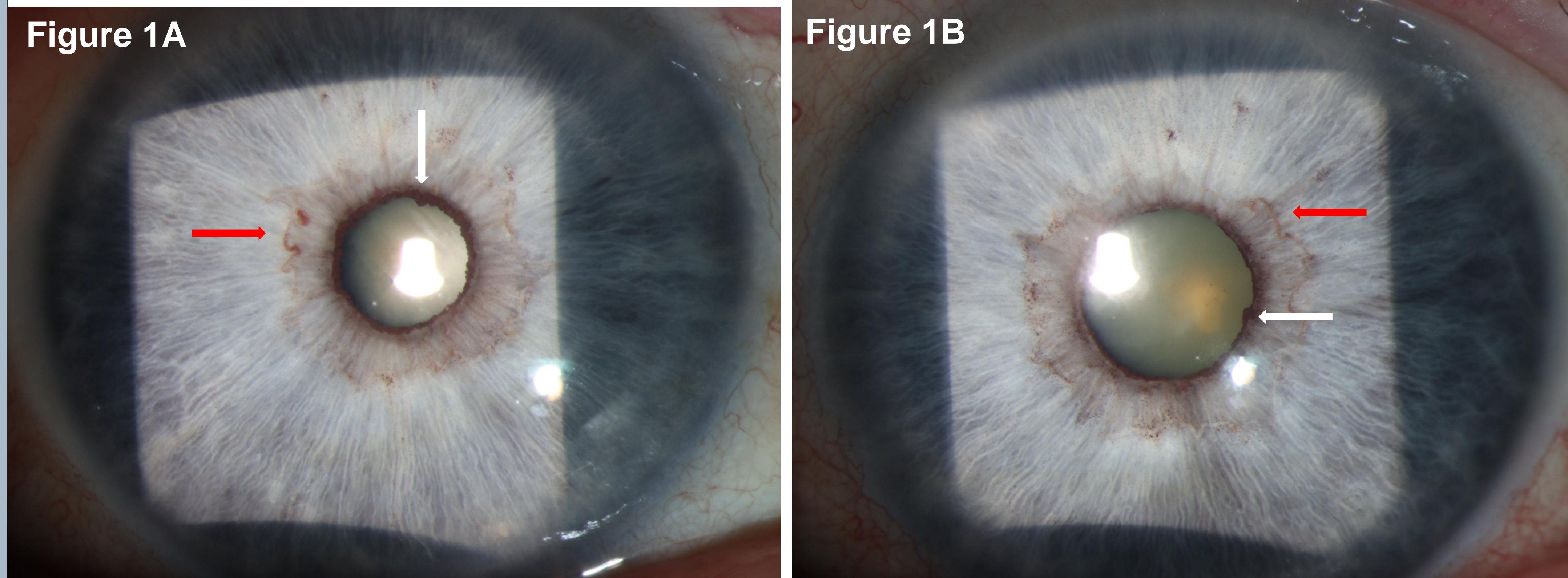
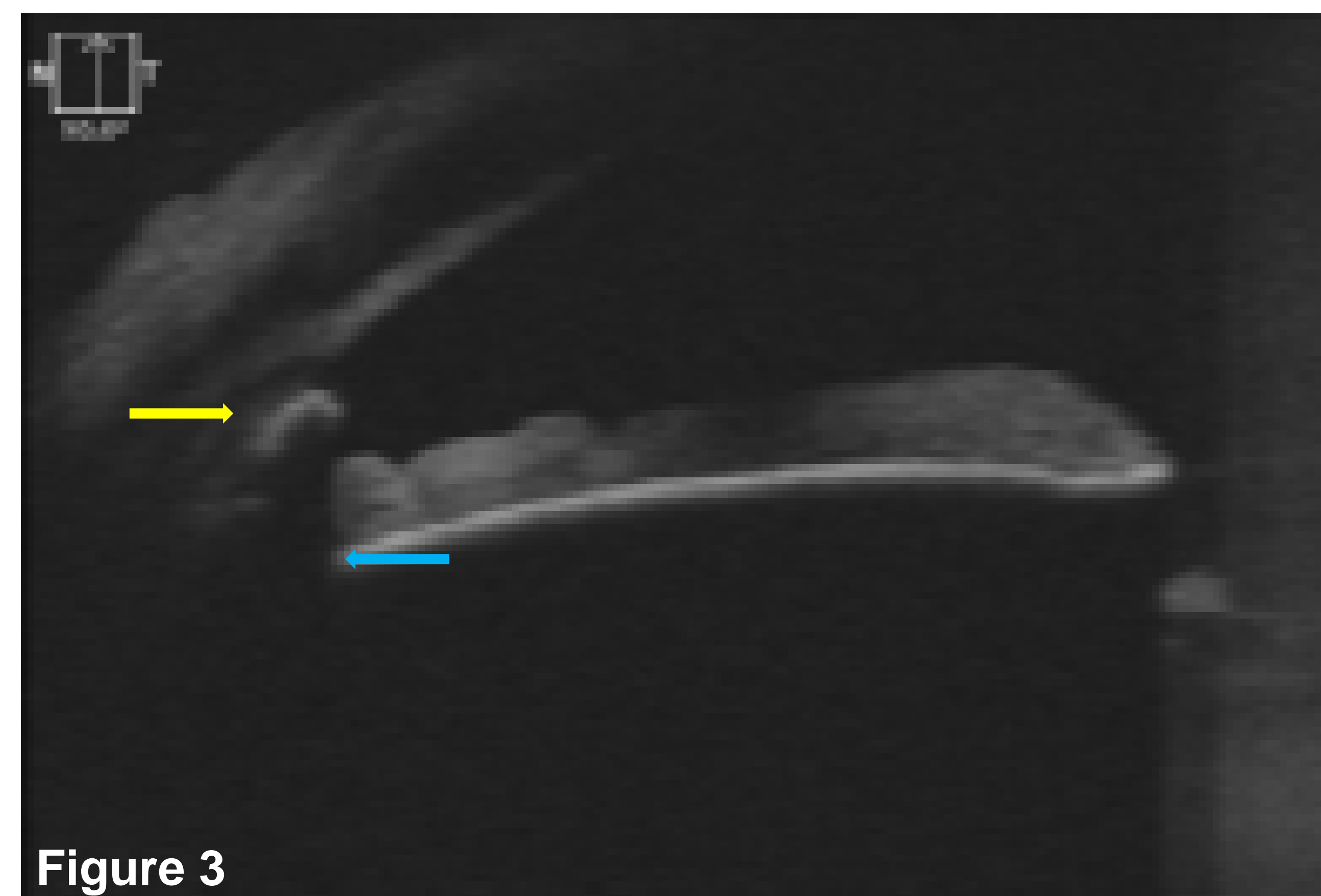


Figure 1. External photograph of the right (A) and left (B) eyes showing small iris floccule at the pupillary margin (white arrows) and prominent iris vasculature (red arrows).

Figure 2. Gonioscopic photograph of the inferior angle of the left eye demonstrating a deflated pigmented lesion with focal PAS.

Figure 3. Anterior segment OCT of the inferior angle of the left eye showing a hyper-reflective lesion (yellow arrow) with posterior shadowing (blue arrow).



DISCUSSION

- Iris lesions may be benign or malignant:⁵
 - Among malignant lesions, 21% are cystic.
 - Among cystic malignant lesions, 86% derive from IPE and 11% from iris stroma
- IPE cysts are benign and may be unilateral or bilateral.⁴
 - They are categorized anatomically as: pupillary, midzonal, peripheral or dislodged.
 - Anterior segment OCT shows a hyper-reflective anterior cyst wall with acoustic shadowing.
 - Complete extent of cyst best visualized on UBM.
- Dislodged IPE cysts account for 2-5% of all iris cysts^{1,4}
 - They may be free floating in the anterior chamber or vitreous or immovable in the inferior angle with varied visual symptoms.²
- Iris flocculi are a special form of pupillary IPE cyst that may wax and wane, thus appearing wrinkled.³
 - Related to mutations in ACTA-2 or MYH11
 - These are associated with familial thoracic aortic aneurysm and dissection, so surveillance is recommended
- Iris microhemangiomas may present with vascularized Cobb’s tufts at the pupil margin and spontaneously bleed.⁶
 - These lesions are evident angiographically and may be treated with argon laser.

CONCLUSION

- In cases of “spontaneous” hyphema, examine the anterior chamber angle and iris closely.
- Wrinkled or waxing/waning iris cysts (iris floccule) should have screening to rule out thoracic aortic aneurysm and dissection.

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