Essential iris atrophy mimicking iris neoplasm: an ultrasound biomicroscopic study

Hesham M. Lakosha, MD, FRCS(Edin); Charles J. Pavlin, MD, FRCSC; E. Rand Simpson, MD, FRCSC

Chandler and Grant[1] described three principle features of essential iris atrophy: distortion and atrophy of the iris, corneal edema caused by dystrophy of the corneal endothelium, and peripheral anterior synechiae (PAS) of the iris to the cornea at Schwalbe's line or anterior to Schwalbe's line.

We describe two cases of unilateral essential iris atrophy that were initially thought to be iris tumours. The diagnosis remained uncertain until the patients manifested the classic features of essential iris atrophy during follow-up. Ultrasound biomicroscopy (UBM) provided valuable information in establishing the diagnosis.

Case reports

Case 1

A 45-year-old white woman was referred to the Ocular Oncology Service for investigation of a possible iris tumour in her left eye. Four months previously she had noticed that the pupil in that eye was changing shape. Her medical history was noncontributory.

Examination showed an unaided visual acuity of 20/20 in either eye. The intraocular pressure (IOP) was 13 mm Hg in the right eye and 12 mm Hg in the left eye. The pupil in the left eye was peaked and drawn inferiorly, with stretching of the iris (Fig. 1, A). Gonioscopy showed tenting of the iris root, which covered the angle and contacted the cornea at 6 o'clock. The angle was otherwise widely open, and there was no evidence of guttate changes of Descemet's membrane. The fellow eye was normal. UBM showed an area of peripheral thickening of the iris with anterior synechiae to the peripheral cornea (Fig. 1, B).

Six months later the visual acuity in the patient's left eye was recorded as 20/40 correctable to 20/20 with 0.50 dioptres. Slit-lamp examination revealed increased stretching and thinning of iris stroma (Fig. 1, C). UBM showed an intrinsic iris cyst in the same quadrant as the PAS (Fig. 1, D).

Fifteen months after presentation a full-thickness stromal hole was noted in the same quadrant as the PAS (Fig. 1, E). Iris transillumination did not reveal any defects. UBM showed that the iris cyst had developed into a full-thickness stromal hole (Fig. 1, F). The iris pigment epithelium was intact. On examination 3 years later the pupil was more drawn inferiorly, with ectropion uveae of the inferior pupillary border. A second area of PAS with adjacent stromal atrophy was evident at 2 o'clock.

Ten months later the patient noticed a further change in the shape of the pupil. There was a third area of PAS at 9 o'clock. Inferior dragging of the pupil continued to the degree that the ciliary processes could be easily seen. One year later the visual acuity remained stable. The pupil was triangular in shape owing to the formation of PAS at three sites, causing ectropion uveae of the entire pupillary border and atrophy of adjacent iris stroma (Fig. 1, G). Transillumination showed a full-thickness iris hole at 9 o'clock. Gonioscopy revealed local angle closure by PAS. The IOP remained within normal limits.

Case 2

An 18-year-old white man was referred for assessment of a possible iris tumour in his right eye. He had

From the Ocular Oncology Service, Princess Margaret Hospital, Ontario Cancer Institute, Toronto, Ont.

Accepted for publication July 28, 2000

Reprint requests to: Dr. Charles J. Pavlin, Ocular Oncology Service, Princess Margaret Hospital, 610 University Ave., Toronto ON M5G 2M9; fax (416) 946-2189; CPavlin@home.com

Can J Ophthalmol 2000;35:390–3

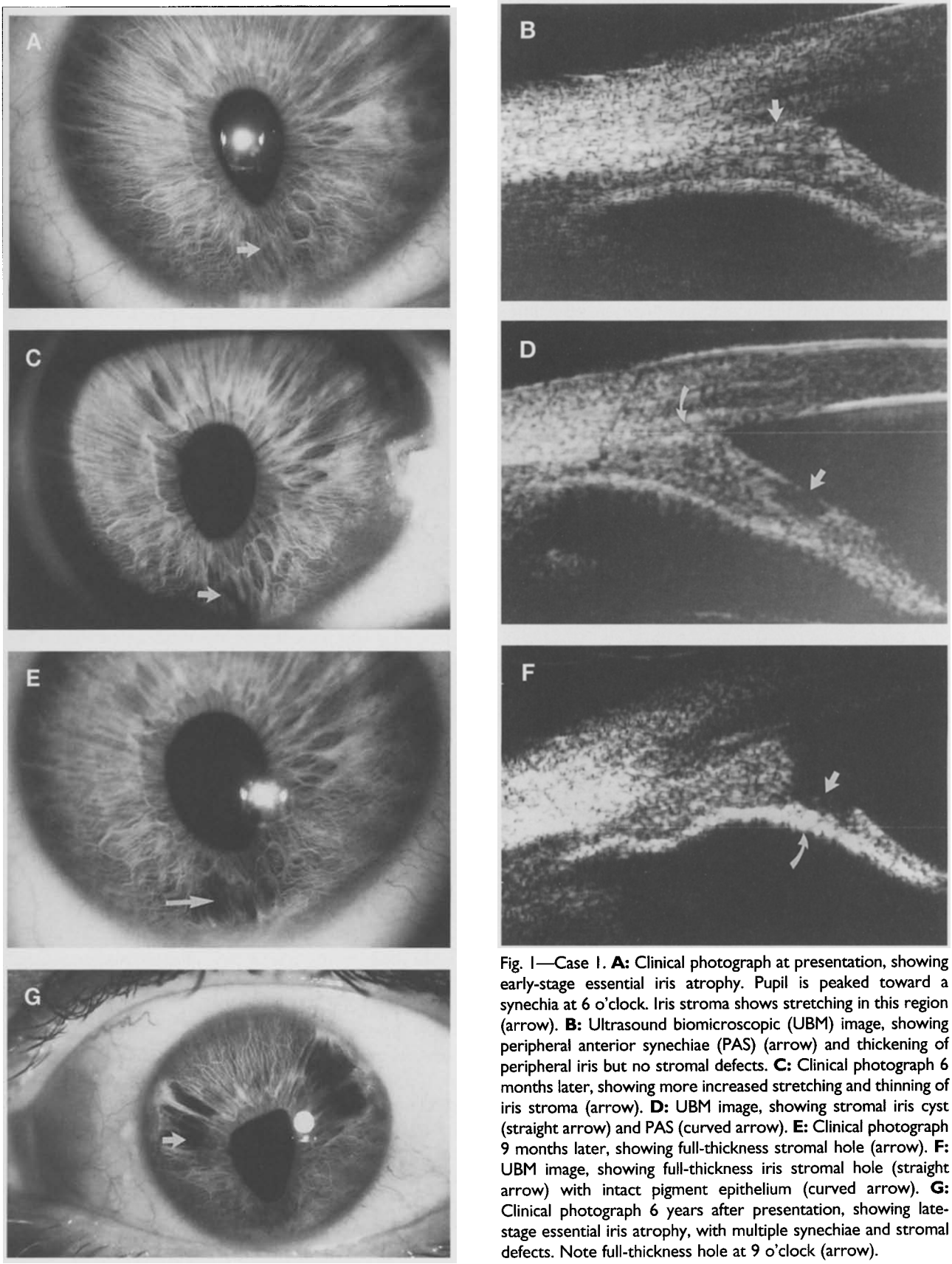

Fig. 1—Case 1. **A:** Clinical photograph at presentation, showing early-stage essential iris atrophy. Pupil is peaked toward a synechia at 6 o'clock. Iris stroma shows stretching in this region (arrow). **B:** Ultrasound biomicroscopic (UBM) image, showing peripheral anterior synechiae (PAS) (arrow) and thickening of peripheral iris but no stromal defects. **C:** Clinical photograph 6 months later, showing more increased stretching and thinning of iris stroma (arrow). **D:** UBM image, showing stromal iris cyst (straight arrow) and PAS (curved arrow). **E:** Clinical photograph 9 months later, showing full-thickness stromal hole (arrow). **F:** UBM image, showing full-thickness iris stromal hole (straight arrow) with intact pigment epithelium (curved arrow). **G:** Clinical photograph 6 years after presentation, showing late-stage essential iris atrophy, with multiple synechiae and stromal defects. Note full-thickness hole at 9 o'clock (arrow).

noticed a change in the shape of the pupil 2 months earlier. He was in good general health. Examination showed a visual acuity of 20/20 in either eye. The IOP was 14 mm Hg in the right eye and 15 mm Hg in the left. The pupil in the right eye was drawn toward 9 o'clock, where there was a raised yellow lesion. The iris adjacent to the lesion appeared thin and atrophic, with obvious ectropion uveae. Gonioscopy revealed the iris root being pulled up, and there was local angle closure. UBM showed a localized mass of the peripheral iris measuring 0.92 mm in thickness associated with PAS (Fig. 2, top). The ciliary body was clear, and the remainder of the ocular examination was unremarkable.

The clinical impression was that of an iris nevus, and the patient was followed every 6 months over 4 years, with no change in the size of the lesion or the appearance of the iris. Four years after presentation the pupil was noted to be more drawn toward the lesion, and there was a second area of PAS with adjacent iris stromal atrophy at 5 o'clock. UBM showed shortening of the iris and crowding at the angle. The iris in this region had contracted into the angle, with a change in shape and a broader synechial base (Fig. 2, bottom). The patient was last seen 2 years later. He reported glare in the right eye but no change in his vision. The pupil was dilated and distorted owing to the development of PAS at three locations (Fig. 3). Sectorial stromal atrophy and significant ectropion uveae were noted. Transillumination revealed a full-thickness iris defect at 4:30. Slit-lamp biomicroscopic examination of the cornea did not show guttate changes of the endothelium, and the IOP remained within normal limits.

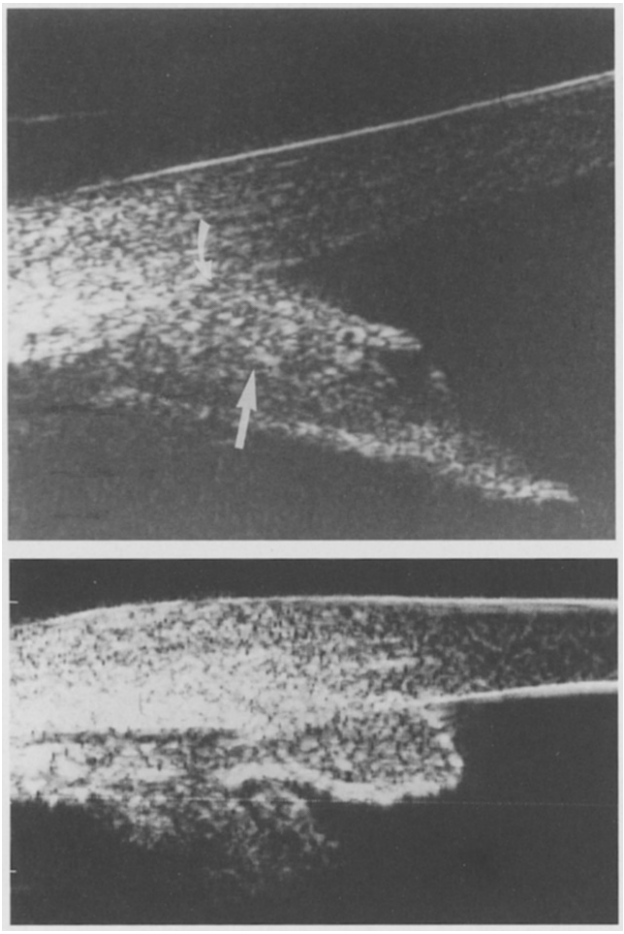

Fig. 2—Case 2. Top: UBM image at presentation, showing iris thickening (straight arrow) and PAS (curved arrow). Bottom: UBM image 4 years later, showing shortening of entire iris and crowding in angle. The area of anterior synechiae has increased.

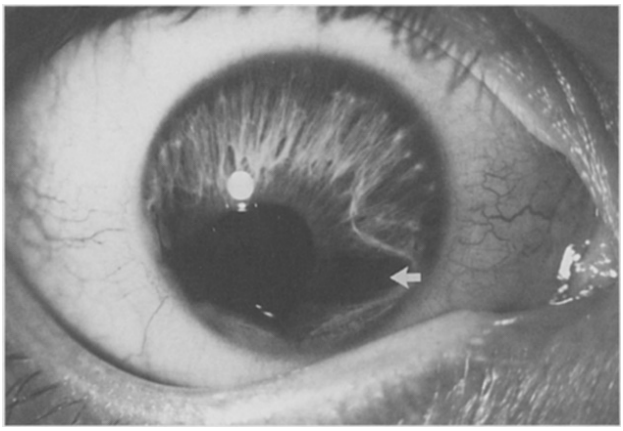

Fig. 3—Case 2. Clinical photograph 6 years after presentation, showing late-stage essential iris atrophy. The pupil has been enlarged and distorted by multiple PAS. Note full-thickness iris hole (arrow).

Comments

In their description of the pathogenesis of essential iris atrophy Campbell and colleagues[2] proposed that a membrane composed of a single layer of endothelial cells develops from proliferation of abnormal corneal endothelium and grows over the anterior chamber angle and the iris. Subsequent contraction of this membrane produces PAS and pupillary distortion with iris thinning.

Early features of essential iris atrophy, including iris distortion and atrophy and the formation of PAS, may be mistaken for an iris tumour. The location of these changes near the iris root and their clinical appearance may suggest a malignant neoplasm. Adding to the

diagnostic difficulty is the possible association of essential iris atrophy with iris nodules. Donaldson[3] was the first investigator to verify the clinical observation of iris nodules in essential iris atrophy. Shields and Campbell[4] reported that iris nodules might develop late in the course of the disease. Some of these nodules, although identified histologically as nevi, could result from stromal compaction by contraction of the endothelial membrane.[5]

In our first patient UBM showed thickening of the peripheral iris with PAS. The thickened iris did not resemble a tumour but, rather, a stretching of the iris stromal tissue due to synechiae. In his description of the natural course of the disease Becker implied that the iris changes precede the formation of PAS.[6] Conversely, Shields and associates[7] suggested that since PAS were present on the initial visit in the cases that they observed, it is likely that PAS formation precedes the iris changes. In our patient UBM showed PAS on the initial visit without evidence of stromal abnormality. A stromal cyst developed over the next 9 months; the iris stroma then looked stretched but did not show a hole. Later, a dehiscence of the iris stroma occurred with a stromal hole. This sequence implies that the synechiae occurred before iris hole formation. UBM can demonstrate cysts associated with iris tumours, but these are generally pigment epithelial cysts rather than stromal cysts,[8] as shown in our case.

In our second patient the initial presentation was that of a mass lesion of the peripheral iris. UBM revealed that the mass was solid and localized to the iris in the same area as the PAS. The mass did not involve the ciliary body and, most important, did not increase in size over time. UBM did show the region of iris thickening changing in shape as further contraction occurred. These UBM features helped rule out malignant neoplasm. Further synechiae and iris defects developed over time. Our patients did not show corneal endothelial changes. These changes may be absent on slit-lamp examination in this syndrome, and specular microscopy may be required to demonstrate the endothelial abnormalities.[9]

In summary, our two cases show how essential iris atrophy can be confused with an iris neoplasm. Features including iris stretching, distortion and PAS can also be seen with iris tumour. Our cases also show the utility of UBM in helping establish a diagnosis in this condition. UBM may be able to demonstrate the anterior synechiae and iris stromal changes before changes are noted in the slit-lamp appearance.

References

1. Chandler PA, Grant WM. *Lectures on glaucoma.* Philadelphia: Lea & Febiger; 1965. p. 276.
2. Campbell DG, Shields MB, Smith TR. The corneal endothelium in the spectrum of essential iris atrophy. *Am J Ophthalmol* 1978;86:317–24.
3. Donaldson DD. Anterior chamber, iris, and ciliary body. In: *Atlas of external diseases of the eye.* vol 4. St Louis: CV Mosby; 1973. p. 213–25.
4. Shields MB, Campbell DG. Iris nodules in essential iris atrophy. *Arch Ophthalmol* 1976;94:406–10.
5. Eagle RC, Font RL, Yanoff M, Fine BS. The iris naevus (Cogan–Reese) syndrome: light and electron microscopic observations. *Br J Ophthalmol* 1980;64:446–52.
6. Kolker AE, Hetherington J. *Becker-Shaffer's diagnosis and therapy of the glaucomas.* 4th ed. St Louis: CV Mosby; 1976. p. 217–8.
7. Shields MB, Campbell DG, Simmons RJ. The essential iris atrophies. *Am J Ophthalmol* 1978;85:749–59.
8. Fine N, Pavlin CJ. Primary cysts in the iridociliary sulcus: ultrasound biomicroscopic features of 210 cases. *Can J Ophthalmol* 1999;34:325–9.
9. Carpel EF. Iridocorneal endothelial syndrome. In: Krachmer JH, Mannus MJ, Holland EJ, editors. *Cornea.* St Louis: Mosby-Year Book; 1997. p. 1107–15.

Key words: ultrasound biomicroscopy, ultrasonography, imaging techniques, essential iris atrophy, iris neoplasm, intraocular tumours, glaucoma