European Journal of Ophthalmology / Vol. 16 no. 6, 2006 / pp. 873-875

SHORT COMMUNICATION

Iris melanocytoma mimicking the Cogan-Reese syndrome with monocular pigment dissemination

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PURPOSE. *To report a case of iris melanocytoma mimicking the Cogan-Reese syndrome.*
METHODS. *A 37-year-old woman presented with pigmentary glaucoma in her left eye. There was diffuse pigmentary dispersion in the anterior segment, pedunculated pigmented nodules on the anterior iris surface, mild iris atrophy, and ectropion iridis. Neither intrinsic vasculature nor a sector cataract was found. The angle was open with marked trabecular pigmentation and no anterior synechiae. The intraocular pressure was 30 mm Hg with maximum medical treatment and there was glaucomatous optic atrophy. The differential diagnosis included iris pigmented tumor and iridocorneal endothelial syndrome (Cogan-Reese syndrome). An iris biopsy was performed for diagnostic purposes.*
RESULTS. *Histologic diagnosis after evaluation of the specimen was iris melanocytoma.*
CONCLUSIONS. *This case presents signs considered quasi-pathognomonic of iridocorneal endothelial syndrome (Cogan-Reese syndrome): glaucoma, mild iris atrophy associated with pedunculated iris nodules, and ectropion iridis. Therefore, iris melanocytoma can present with features that mimic the Cogan-Reese syndrome. (Eur J Ophthalmol 2006; 16: 873-5)*

KEY WORDS. *Cogan-Reese syndrome, Iris melanocytoma, Pigmentary dispersion*

Accepted: July 4, 2006

INTRODUCTION

Melanocytoma is a variant of melanocytic nevus with distinctive clinical and pathologic features (1). Melanocytoma is usually located on or adjacent to the optic disk. Infrequently it can arise in the iris, ciliary body, choroid, or conjunctiva (2). Iris melanocytoma poses a diagnostic challenge. The differential diagnosis of such a lesion usually includes nevus or iris melanoma.

The iridocorneal endothelial (ICE) syndrome takes many clinical forms but usually includes some combination of iris atrophy, corneal edema, and secondary angle-closure glaucoma without pupillary block. This syndrome is caused by an abnormal corneal endothelium that forms a membrane over the anterior surface of the iris and the angle structures (3). The Cogan-Reese syndrome or iris nevus is a clinical form of the ICE syndrome and is differentiated from other forms by pigmented iris lesions. Some eyes have pedunculated iris nodules.

Case report

A 37-year-old woman presented with pigmentary glaucoma in her left eye. A previous left trabeculectomy had been performed at another hospital, the details of which were unavailable. On examination, the right eye was normal. The affected left eye had a best-corrected visual acuity of 20/400. Slit-lamp examination showed diffuse pigmentary dispersion in the anterior segment, pedunculated pigmented nodules on the anterior iris

Presented at the 5th International Glaucoma Symposium; Cape Town, South Africa; March 31 - April 2, 2005

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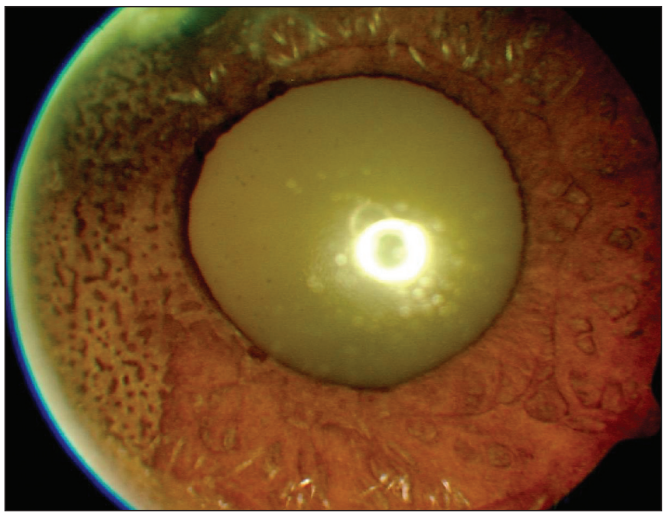


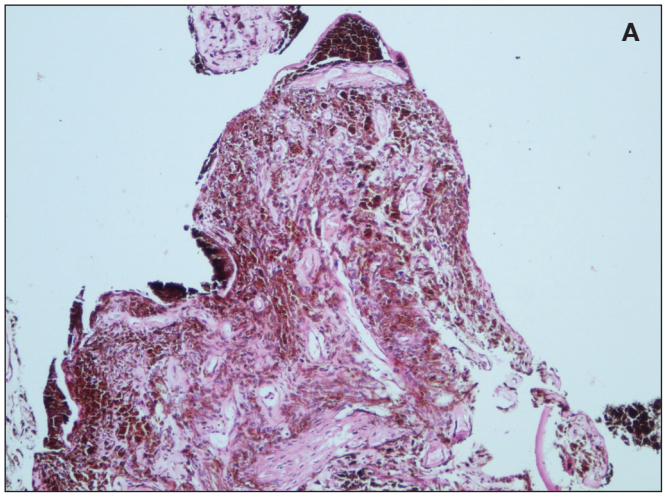
Fig. 1 - *Mild iris atrophy associated with pedunculated iris nodules and ectropion iridis.*

surface, mild iris atrophy, and ectropion iridis. The lesion was diffuse without a nodular component. There were no transillumination defects. Neither intrinsic vasculature nor a sector cataract was found (Fig. 1). The angle was open with marked trabecular pigmentation and no anterior synechiae. The intraocular pressure (IOP) was 30 mm Hg with maximum medical treatment, and the optic nerve showed advanced glaucomatous cupping. Ultrasound did not reveal any mass on the surface or in the ciliary body.

The diagnostic possibilities included iris pigmented tumor and the ICE syndrome (Cogan-Reese syndrome). An iris biopsy was performed for diagnostic purposes. We performed a superior scleral flap, opened the anterior chamber, and performed an iridectomy. Histologic examination of the iris fragments revealed copious pigment obscuring the nuclear details on routine hematoxylin-eosin sections (Fig. 2A). The tumor was comprised of plump, polyhedral nevus cells with large quantities of pigmented melanin that obscured the nuclear details. A bleached section disclosed bland nuclei and a low nuclear/cytoplasmic ratio (Fig. 2B). The histopathologic features were consistent with a melanocytoma.

DISCUSSION

Our case is one of a few well-documented cases of iris melanocytoma, which is rare and represents only 3% of all iris nevi. Related iris stromal and anterior chamber angle seeds are commonly seen. Secondary glaucoma appears in 11% of cases 5 years after diagnosis. However, ectropion iridis is found in only 6% of cases (4). Our case presented unusual signs because, to our knowledge, no study has previously described mild iris atrophy associated with pedunculated iris nodules and ectropion iridis in conjunction with iris melanocytoma glaucoma. These signs have been considered quasi-pathognomonic of the ICE syndrome (Cogan-Reese syndrome). In addition, iris melanocytoma is a discrete, localized le-



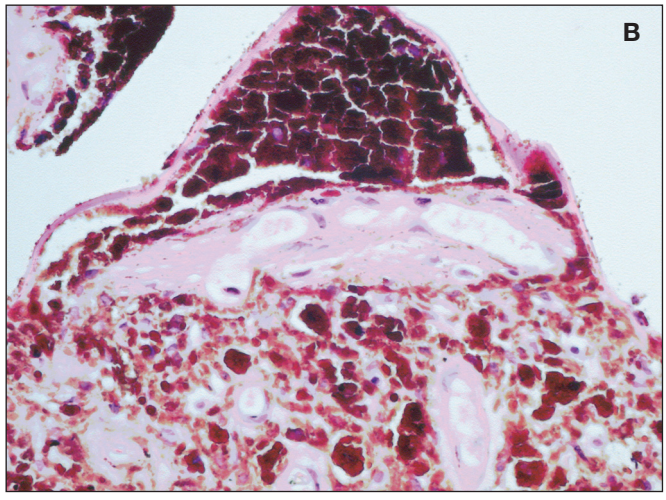


Fig. 2 - (A) *Histopathologic examination shows an intensely pigmented tumor comprised of plump polyhedral nevus cells with copious maximally pigmented cytoplasm that obscures the nuclear details (hematoxylin-eosin x 200).* **(B)** *A bleached section shows bland nuclei and a benign nuclear/cytoplasmic ratio (hematoxylin-eosin x 400).*

sion. However, we found a diffuse lesion.

The natural histories of both diseases differ. The Cogan-Reese syndrome is a clinical form of the ICE syndrome, a group of disorders characterized by abnormal corneal endothelium that causes variable degrees of iris atrophy, secondary angle-closure glaucoma, and corneal edema. Three clinical variants have been described: Chandler syndrome, essential progressive iris atrophy, and the Cogan-Reese syndrome. Various degrees of iris atrophy and corneal changes distinguish the specific clinical entities. The iris atrophy and corneal edema tend to be less severe in the Cogan-Reese syndrome. This condition is distinguished by tan pedunculated nodules or diffuse pigmented lesions on the anterior iris surface. Angle-closure glaucoma, which can be treated medically, occurs in about 50% of patients. When medical therapy fails, filtering surgery (trabeculectomy or tube-shunt procedures) can be effective. On the other hand, iris melanocytoma represents only 3% of all iris nevi (4). It usually is a discrete localized lesion that can sometimes undergo spontaneous necrosis with secondary pigment dispersion, elevated IOP, and iris heterochromia; however, other clinical presentations are possible as in our case. Frequent observation is usually recommended. Local resection is reserved for treatment of increased IOP or for diagnostic purposes.

In conclusion, iris melanocytoma can mimic the Cogan-Reese syndrome. It is important to differentiate between the diagnoses, because the treatments differ.

The authors have no proprietary interest in any aspect of this report.

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