Anterior subcapsular plaque cataract in hyperornithinaemia gyrate atrophy – a case report

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Abstract
Hyperornithinaemia gyrate atrophy (HOGA) is a rare autosomal recessive disorder in which chorioretinal degeneration occurs with cataracts, myopia, and hyperornithinaemia. We report the case of an 18-year-old female who presented with the typical features of HOGA, including posterior subcapsular cataracts and elevated plasma ornithine. She later developed distorted vision in both eyes owing to wrinkling of the anterior lens capsules. Histological examination following lens extraction showed the wrinkling was caused by focal distortion from capsular fibrosis (anterior subcapsular plaque cataract). This specific lens change has not been linked previously with HOGA.

Hyperornithinaemia gyrate atrophy (HOGA) is a rare autosomal recessive disorder in which chorioretinal degeneration occurs with cataracts, myopia, and hyperornithinaemia. The ocular changes may be caused by either hyperornithinaemia or possibly hypoprolinaemia. Two clinical subgroups have been described based on vitamin B6 responsiveness with vitamin B6 responsive patients having a less severe and more slowly progressive disease. We report a case which presented with posterior lens opacities typical of HOGA, but later developed anterior subcapsular plaque cataracts bilaterally; a specific lens change which has not been previously linked with HOGA.

Case report
An 18-year-old female presented with reduced visual acuities of 6/12 right and left. A presumptive diagnosis of HOGA was made on the basis of posterior subcapsular cataracts, myopia, confluent well circumscribed areas of peripheral chorioretinal degeneration, and a subnormal electroretinogram.

Ten years later she developed distorted vision in the right eye. Examination revealed visual acuities of 6/60 (6/18 with a pinhole) right, 6/18 left and a wrinkling of the right anterior lens capsule (Fig 1). The fundus changes in both eyes were typical of HOGA (Fig 2). Her own plasma ornithine was markedly elevated at 776 μmol/l, while her parents, brother, and daughter all had normal 24 hour urine and plasma ornithine levels.

Three months later the previously normal left anterior capsule became wrinkled. Histological examination following lens extractions showed extensive degenerative changes with formation of globular aggregates in the posterior suture region and posterior migration of the lens epithelium to form a uniformly thickened 'double layer' posterior capsule. The anterior capsule wrinkling was due to focal distortion from capsular fibrosis (anterior subcapsular plaque cataract). Her visual acuities have now improved to 6/12 in each eye.

Discussion
This patient illustrates all the typical features of HOGA, including the specific light and electron microscopic appearance reported in HOGA cataracts. She had, in addition focal proliferation of epithelial cells to form anterior subcapsular cataracts. The only previous report of this type of cataract in HOGA concerned a postmortem study on an eye from an 89-year-old female with previous unspecified ocular trauma with the lens also being subluxated and hypermature.
Wrinkling of the anterior capsule occurs in hypermature cataracts, and with capsule fibrosis initiated by anterior segment inflammation caused by trauma, surgery, uveitis, or atopy. The lenses in our study developed anterior capsule wrinkling in the absence of any predisposing factors suggesting anterior subcapsular plaque may be another specific cataractous change associated with HOGA.

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