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**Granular Corneal Dystrophy, type 2**

* In the late 1880 a German neurologist, Wilhelm Heinrich Erb, coined the term “muscular dystrophy” and characterized this group of diseases as progressive, degenerative primary muscular abnormalities
* Dystrophy derives from the Greek “dys” meaning wrong, difficult, and from “trophe”, meaning nourishment.
* In 1890, the German Ophthalmologist Arthur Groenouw, working in Breslau, then Germany (currently Wroclaw, Poland), published the first clinical observations about degenerative corneal conditions which he called “Knötchenförmige Hornhauttrübungen”, which were translated into English as “Noduli Corneae”.
* His description is rather detained: numerous scattered corneal deposits with round to ragged edges which are stable in time, populate predominantly the central cornea, and do not elicit an accompanying inflammatory reaction. Some larger deposits elevate the overlying epithelium
* Subsequently the term corneal dystrophy began being used
* Early in the twentieth century ophthalmologist observed several types of corneal dystrophies and early descriptions were published of what used to be called Groenouw type I and II, Fleischer familial corneal dystrophy type I and II, or Bücklers corneal dystrophy type I and II.
* Max Bücklers, German Ophthalmologist considered granular dystrophy as the most frequent of the corneal dystrophies (1938)
* J. Reimer Wolter and William M. Cutler reported granular corneal dystrophy in the American Medical Literature in 1958.
* 1961 Sam T. Jones and Lorenz E. Zimmerman published a detailed histopathologic description of granular, macular, and lattice corneal dystrophies
* 1988 the first report of a corneal condition with characteristics of granular and lattice dystrophies was published by Robert Folberg. His study included Italian families in Pennsylvania, USA, which had their origins traced back to the Avellino region in Italy.
* The term Avellino Dystrophy was largely used, although in some reports the term combined granular-lattice corneal dystrophy was favored
* Avellino dystrophy is found to have its genetic defect mapped to the long arm of chromosome 5. Independently granular dystrophy and lattice dystrophy were also linked to the same genetic marker on 5q.
* 1997 granular dystrophy Groenouw type I (CDGG1), Reis-Bücklers (CDRB), lattice type I (CDL1) and Avellino (ACD) were all four found to be 5q31-linked human autosomal dominant corneal dystrophies, all implying changes in transforming growth beta-induced gene (TGFBI).
* Avellino corneal dystrophy was found to not be restricted to Avellino region in Italy
* The nomenclature of these corneal dystrophies was unified. The TGFBI corneal dystrophies include:
  + Granular corneal dystrophy type 1 was linked to mutation R555W in exon 12
  + Granular corneal dystrophy type 2 was linked to mutation R124H in exon 4
  + Granular corneal dystrophy type 3 (Reis-Bücklers dystrophy)
  + Thiel-Behnke corneal dystrophy was linked to mutation R555Q in exon 12
  + Lattice corneal dystrophy type 1 was linked to mutation R124C in exon 4
* Lattice corneal dystrophy type 2 (gelsolin type) has been reclassified as systemic amyloidosis and is no longer considered a corneal dystrophy in IC3D classification.

Granular corneal dystrophy type 2 (GCD 2) shows variable clinical expression and onset (as early as age 3 in homozygotes and as early as age 8 in heterozygotes, but most heterozygotes are diagnosed in their second decade or even early adulthood).

Because of this late onset of the disease, some patients undergo corneal surgery (LASIK, LASEK, PTK, or PRK) without being properly diagnosed. Marked exacerbation of the clinical course was observed in GCD 2 following such corneal surgical procedures.

I am presenting here the case of a now 49 year old female who had LASIK performed age 31 while considered to have normal corneas. Age 42 she had both LASIK flaps removed because of massive interface accumulations of granular hyaline deposits. Later on, age 48, she had full PK performed on one side due to further exacerbation of her granular deposits.

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