Lattice Corneal Dystrophy
Type 3: A Case Report

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Introduction
Corneal dystrophies are a group of heterogenous bilateral, genetically determined non-inflammatory diseases that are restricted to the cornea. Corneal dystrophies may have an autosomal dominant or recessive, or X-linked recessive Mendelian mode of inheritance. Clinically, the corneal dystrophies can be divided into 3 groups. Some affect primarily the corneal epithelium and its basement membrane or Bowman’s layer and the superficial corneal stroma (Anterior corneal dystrophies), the corneal stroma (Stromal corneal dystrophies) or Descemet’s membrane and corneal endothelium (Posterior corneal dystrophies). Here is a rare case of a middle aged lady presenting with unilateral corneal stromal (lattice) dystrophy.

Case Report
A 57 year old lady presented to our out patient department in November 2010 with complaints of defective vision in her left eye since 6 months. She did not have any pain or redness. She had undergone cataract surgery in both eyes in 2002. Post operative period was uneventful for both eyes. On examination she had a BCVA of 6/9 in her right eye and 6/60 in her left eye. Slit-lamp examination of right eye showed clear cornea with PCIOL in situ (Figure 1). In the left eye the cornea had thick, ropy lattice lines involving epithelium and anterior stroma (Figure 2 and 3). There was no evidence of corneal erosions. The PCIOL was in situ and there was no evidence of PCO. Fundus examination was within normal limits. Specular microscopy was normal for both eyes. There were no systemic features suggestive of amyloidosis. There was no history of any corneal disease in the family. A diagnosis of Lattice (stromal) corneal dystrophy type 3 was made in view of late presentation, unilateral occurrence and absence of recurrent erosions. The patient is currently under follow-up.

Discussion
Corneal stromal dystrophies are caused by progressive accumulation of deposits within the stroma. These may or may not affect vision and may or may not be symmetrical. They usually present in the 2nd or 3rd decade of life. The different types of stromal dystrophies are Lattice corneal dystrophy, Granular dystrophy, Avellino dystrophy, Macular dystrophy, Gelatinous drop-like dystrophy, Schnyder crystalline dystrophy, Francois-Neetens fleck dystrophy and Congenital hereditary stromal dystrophy.

Lattice corneal dystrophy (LCD) is the most common of the corneal stromal dystrophies. The disease has been linked to a mutation in the TGF β1 gene, also known as BIGH3 gene. This gene codes for keratoepithelin, a protein secreted by corneal epithelium. LCD is of 3 types mainly:

LCD Type 1 (Biber-Haab-Dimmer): It is an AD disorder with gene locus on chr 5q31 and presents at the end of 1st decade with recurrent erosions which precede typical stromal changes. Slit lamp examination reveals anterior stromal dots which, with time coalesce into fine spidery branching lattice lines. Eventually a generalized haze settles in, which impairs vision and obscures lattice lines. Histopathology shows amyloid which stains with Congo red.

LCD Type 2 (Finnish type or Meretoja syndrome): Inherited as AD with gene locus on chr 9q34. It presents in middle-age with progressive facial palsy and corneal involvement. Recurrent erosions are less common. Cornea shows randomly scattered, short fine lattice lines which are sparse, more delicate and more radially oriented than in type 1. Systemic features of amyloidosis involving central and peripheral nervous systems (Familial amyloid polyneuropathy type IV), skin, kidney and heart may be seen. Histology shows amyloid deposits in corneal stroma and other involved sites.

LCD Type 3 and 3A: Inheritance of type 3 is presumed to be AR and that of type 3A is AD with gene locus on chr 5q31 in both. It presents between 4th and 6th decades with visual impairment. Recurrent erosions are uncommon. Cornea shows thick ropy lines extending from limbus to limbus with minimal intervening haze. There may be gross asymmetry or the lesion may be unilateral for a time. Progression is rapid with even trivial trauma. Type 3A differs from type 3 in that it may be associated with corneal erosions, is more common in Whites and has an AD inheritance.

LCD Type 4 has been described as a late-onset dystrophy occurring in deep stroma, also associated with BIGH3 gene. Many patients with LCD will require surgical intervention for treating recurrent erosions and decreased vision. If the disease is located anteriorly in the stroma, patients can often be successfully treated with phototherapeutic keratectomy. Some require penetrating or lamellar keratoplasty. The disease can recur in corneal grafts.

Summary
Corneal dystrophies are a rare group of disorders seen mostly in younger population. But these entities should be suspected in older patients presenting with unexplained
vision deterioration and a normal posterior segment. A thorough slit lamp examination is therefore mandatory in such patients so that appropriate therapy can be instituted. This case has been reported for its rare unilateral presentation in an older patient.

References
2. Gordon K Klintworth- Corneal dystrophies- Orphanet journal of rare diseases 2009. 4:7,
Pentacam

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Introduction

Pentacam is a device that does an anterior segment tomography as opposed to a topography in most other topographers. It is as different as a CT scan and a regular X ray.

It is an imaging technique where a series of cross-sectional images are merged to allow for a computer-generated three-dimensional reconstruction. The advantages of anterior segment tomography compared to corneal topography are similar to the advantages of CT scanning compared to standard X ray. Anterior segment tomography allows not only the visualization of the anterior corneal surface, but the posterior corneal surface, the anterior chamber, the corneal thickness, portions of the angle of the eye, and the lens. The computer-generated three-dimensional reconstruction allows for a much expanded range of applications. Measurements such as anterior chamber depth, corneal thickness, and lens density are samples of measurements available with anterior segment tomography that were not available with older devices that just measured the anterior corneal surface.

Principle

The Scheimpflug principle was a technique used to capture images when the image plane is not parallel to the lens plane (figure-2). This is named after an Austrian captain who used this technique to correct distortions seen in aerial photographs.

The Pentacam is a combined device consisting of a slit illumination system and a Scheimpflug camera which rotates around the eye.

A thin layer within the eye is illuminated through the slit. Being not entirely transparent the cells scatter the slit light. In doing so they create a sectional image which is then photographed in side view by a camera. This camera is oriented according to the Scheimpflug principle, thus creating an image of the illuminated plane which appears completely sharp from the anterior surface of the cornea right up to the posterior surface of the crystalline lens. Swiveling around the eye, the slit-camera device generates a series of radially oriented images of the anterior eye chamber. In the subsequent analysis of the sectional images, tissue boundaries are detected and point clouds are assigned to the various tissue layers (anterior and posterior corneal surfaces, iris, crystalline lens).

The sectional images are saved, corrected in relation to a common reference point and then put together to create a three-dimensional model of the entire anterior eye chamber. This makes it possible to generate reproducible tomographic images of the anterior eye chamber in any desired plane.

After correction for Scheimpflug distortion and light refraction at tissue interfaces the exact location of image edge points in the eye is determined by means of ray tracing. Eye movements during image acquisition are captured by a second camera (pupil camera) and also taken into account in the mathematical evaluation. This produces a set of three-dimensional measurement data which gives a precise geometric description of the anterior eye segment. This data

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in turn can be used to generate data on elevation, curvature, pachymetry, depth of the anterior eye chamber etc. in the well-known form of colour maps.

Differences in brightness between individual layers provide an indication of any opacity, since transparent layers result in less slight scattering. This information can be used to assess cataracts.

This contact-free, hygienic measurement process takes less than two seconds.

This is the time it takes to generate 50 sectional images yielding in turn 138,000 true elevation values.

**Uses of the pentacam**

1. LASIK

The most important use of the pentacam is in refractive surgery. The pentacam gives a quad map giving the anterior and posterior elevation of the cornea, a limbus to limbus corneal pachymetry and the anterior curvature map. A patient can have a normal anterior curvature and still have an ectatic posterior surface which we may miss with a regular topography. Changes on the posterior surface and/or changes in pachymetric distribution or progression will typically predate any changes of the anterior corneal surface.

![Figure 3. The Quad Map](image)

Comparing the posterior corneal surface pre-operatively to the postoperative map is the most sensitive method to identify early ectatic change.

The Pentacam displays the pachymetric distribution as graphs of the corneal thickness spatial profile (CTSP) and percentage thickness increase (PTI). It records the thinnest point as the starting value, then it displays the average of the thinnest values on imaginary concentric rings accordingly to the diameter in the CTSP. The PTI display shows the same data as the CTSP, but it considers the percentage of increase of the average at each ring from the thinnest value. Both the CTSP and PTI graphs are displayed in conjunction with the mean and two standard deviations from normality. The rationale for the CTSP and PTI graphs is that normal corneas are thinner in the center and gradually thicken toward the periphery. Ectasia is defined as a progressive thinning disease. The pachymetric distribution in ectasia has a more abrupt thickening in the periphery, which is consistent with thinning, the hallmark of ectasia.

The Belin/Ambrosio Enhanced Ectasia Display (BAD) is a display used to check the suitability of the eye for LASIK.

The goal of the BAD was to combine elevation based and pachymetric corneal evaluation in one comprehensive display to give the clinician a global view of the tomographic structure of the cornea. Deviation of normality values were implemented for the front and back enhanced elevations, pachymetric distribution and vertical displacement of the thinnest in relation to the apex. The final D was developed for having lower than 5% of false positives and false negatives. The relevance of the BAD was determined in a study of eyes with highly asymmetric keratoconus, in which it was confirmed the superior sensitivity of the display over front surface curvature and central thickness evaluation. The combination of the new elevation based approach and the pachymetric distribution has increased sensitivity to more than 90% in corneas with normal curvature maps from patients previously diagnosed with “unilateral keratoconus”.

By looking at the five previous mentioned parameters (change in anterior elevation, change in posterior elevation, corneal thickness at the thinnest point, thinnest point displacement, and pachymetric progression) and performing a regression analysis against a standard data base of normal and keratoconic corneas. It reports five new terms (D values for standard deviation from the mean) representing the front surface (Df), back surface (Db), pachymetric progression (Dp), thinnest point (Dt), and thinnest point displacement (Dy).

A sixth term (D) is the final overall map reading taking each of the five parameters into account. Each individual parameter D and the final D number have been normalised to their mean value and are reported as standard deviations from the mean. The individual parameters are also colour coded based on their variation from the norm. The parameter is indicated in yellow (suspicious). The major advance is that while an individual parameter(s) may fall outside the norm the final overall comprehensive reading may still be viewed as normal. Conversely, multiple yellow or suspicious parameters may be significant enough for the final reading D to be RED or abnormal.

The Belin/Ambrosio Enhanced Ectasia Display is the first comprehensive refractive surgical screening tool to be fully elevation based and to incorporate data from the posterior corneal surface and corneal thickness map. The
Fredy T. Simon - Pentacam

newest release (version II) takes the analysis one step further by normalizing each parameter (allowing for an easier interpretation of relative risk) and provides a final overview reading ("D" value) of the entire map. It is hoped that this additional information will simplify the interpretation of the maps and provide greater specificity and sensitivity for detecting early ectatic disease.

Figure 4. A normal Belin/Ambrosio Enhanced ectasia map

Figure 5. An abnormal BAD map

2. Cataract Evaluation

Pentacam gives the cataract surgeon an objective evaluation of the cataract density and volume. This can help the surgeon with the pre op planning of the phacoemulsification parameters to be used so as to increase the efficacy and safety of the surgery.

The Pentacam’s Scheimpflug camera offers 3D lens densitometry via the PNS software (Pentacam nuclear grading system). The PNS sampling technology allows surgeons to evaluate the individual components of the cataract in three dimensions and determine the relative density of the nucleus and epinucleus. They may then modify and set their preferred phaco settings linked to the cataract’s grade. Ultimately, surgeons may be able to establish default phaco settings that they initiate at the start of surgery based on the PNS grade. The PNS is user-friendly, reproducible, transferable, and it is based on the Pentacam’s objective calibration system. After scanning an individual’s cataract, the surgeon selects the Scheimpflug overlay and then picks the most centered cross-section. Then, he or she clicks on the PNS nuclear grading tab, and a graph with a gradation number from zero to five will appear. This number is important, because it provides a comparison between what is visible at the slit lamp and the PNS’ grading of the nucleus. The physician may then mark this number on the chart and use it in the OR for setting the phaco parameters. The result is a user-friendly nuclear grading system.

Donald Dixon calculated the power used in the different nuclear grades based on pentacam and found that the power used correlated with the density of the nucleus.

Figure 6. Cataract on the pentacam

3. IOL Power Calculation Post LASIK

In cases with previous refractive surgery the keratometric readings based only in anterior corneal curvature, such as those obtained with a Placido-based topography or a keratometer, will give an erroneous value, because they are unable to measure the posterior corneal radius. These instruments also assume that the ratio between the posterior and anterior corneal radii is 82%, this ratio is changed following all types of ablative procedures. In addition, topographers and keratometers are blind to the exact center of the cornea and must extrapolate this information. Since the pentacam calculates the posterior curvature and gives a true reading of the centre of the cornea the reading are more accurate.

The Holladay Report, a module developed by Oculus with Dr. Jack T. Holladay. It gives the physician a realistic measure of...
the optical power of the cornea at different zones, as well as an estimation of the preoperative simulated K readings. The ophthalmologist has to be aware that, although these values are more precise than those obtained with other instruments, they can only be used to calculate the IOL power in formulas that consider the origin of the data, like the BESSs or Holladay 2 formulae. Calculation methodologies such as the Aramberri Double-K (2) method or the Haigis-L formula were developed using K readings obtained from keratometers and could give incorrect results a central corneal power from the Pentacam is used.

4. Other Uses

a. Glaucoma

Pentacam can be used to get various parameters of the anterior chamber such as the chamber depth, chamber volume, chamber angle etc. This is especially useful in angle closure glaucoma. Any anterior chamber volume less than 100 mm³ is considered abnormal so also any angle less than 250.

b. Pentacam is also used to plan the depth and position in inserting intracorneal rings for keratoconus. It can accurately calculate the depth of implantation and the centre of the cone.

c. Pentacam also has software for contact lens fitting, keratoconus grading etc.

Conclusion

The Pentacam is an anterior segment tomographer with various applications for the anterior segment surgeon. It is most valuable in LASIK. There is still a lot of varied opinions on the criteria to reject patients for LASIK. With the BAD map we can have definite criteria for accepting or rejecting a patient for LASIK.

References