Unilateral Lattice Corneal Dystrophy: A Case Report

Abstract: There are few reported cases of unilateral lattice corneal dystrophy (LCD). This case presents a patient with unilateral LCD with family history of bilateral, type 1 LCD.

I. Case History
   A. Patient demographics: 38-year-old African American male
   B. Chief complaint: Presented as a new patient for an emergency visit complaining of severe eye pain
      1. Location: Right eye
      2. Duration: 21 days
      3. Timing: constant throughout the day, initial symptoms began in the morning
      4. Severity: 10/10
      5. Other symptoms: severe redness and decreased vision that had not improved over the prior 3 weeks
      6. Modifying factors: eye patch and towel to cover the eye, but had not received any medical care or other treatment
   C. Ocular, medical history
      1. Further questioning revealed that the patient had been suffering recurrent episodes of a painful, red eye for the previous 20 years. The episodes always involved only the right eye and occurred once or twice each year. Each episode lasted 2 weeks to a month, and involved redness, pain, and decreased vision. The patient reported that each previous episode had resolved with his vision returning completely. The patient reported that he only sought treatment for these episodes while he was serving time in prison 3 years prior to his current presentation. He reported being told he had “herpes in his eye” and that he needed a corneal transplant. The patient was given an unknown eye drop to use “almost every 10 minutes” for an unknown duration. All other episodes resolved without intervention.
      2. All other ocular history was negative
      3. Medical history was unremarkable
   D. Medications: None
   E. Other salient information: Family ocular history
      1. Positive for mother with corneal transplant on one eye and scheduled to have surgery on fellow eye
      2. He was unsure of the reason for the corneal transplant at the initial presentation.

II. Pertinent findings
   A. Clinical
      1. Visual acuity: Hand motion at 4 feet OD, 20/20 OS with no improvement on pinhole OD
2. Pupils: Equal, round, and reactive to light with no afferent pupillary defect present OD, OS
3. All other entrance testing was normal OD, OS
4. Anterior segment evaluation revealed diffuse, 4+ injection of the bulbar conjunctiva and 2+ injection of the inferior and superior palpebral conjunctiva OD. There was no foreign body present upon lid eversion, and no papillae or follicles in either eye. There was an epithelial defect measuring 6.5 mm horizontal by 3.5 mm vertical located centrally OD. Sodium fluorescein stained throughout the defect, but rose bengal did not stain anywhere on the cornea. The central stroma surrounding the defect showed 2+ edema. Underlying the epithelial defect and extending nasally and temporally to about 3 mm from the limbus and superiorly and inferiorly to 3.5 mm from the limbus were multiple linear, branching refractile lines in the anterior stroma OD. There was stromal haze between the linear opacities, and the opacities were most dense centrally. The branching lines were best viewed in retroillumination. There was no corneal neovascularization present in either eye. The anterior chamber was difficult to view OD, but appeared quiet with no cells or flare. All anterior segment findings were normal in the left eye. The left cornea was clear throughout, with no linear stromal opacities and no epithelial staining OS.
5. Posterior segment: Unremarkable in both eyes

B. Other findings: Family clinical history
   1. At his 1-day follow up visit, he reported that his mother has “some corneal dystrophy”.
   2. Further record review in subsequent days revealed that the patient’s mother was diagnosed with lattice corneal dystrophy OU.

III. Differential diagnosis
   A. Primary/leading differential for epithelial defect: Recurrent corneal erosion OD
   B. Other differentials for epithelial defect
      1. Active geographic Herpes Simplex Virus epithelial keratitis
      2. Corneal abrasion
   C. Primary/leading differential for linear, branching stromal lines: Lattice corneal dystrophy
   D. Other differentials for linear, branching stromal lines
      1. Polymorphic amyloid degeneration
      2. Avellino dystrophy
      3. Prominent corneal nerves
      4. Prominent corneal ghost vessels

IV. Diagnosis and discussion
   A. Primary diagnosis: Recurrent corneal erosion secondary to lattice corneal dystrophy OD only
      1. Despite the unilateral presentation, the refractile, linear, branching opacities in the central anterior stroma, the history of recurrent corneal
erosions, and the family history of corneal transplant all suggested lattice corneal dystrophy.

2. Active HSV keratitis with previous stromal keratitis and ghost vessels was ruled out as the epithelial lesion did not stain with rose bengal, there was no reduction in corneal sensitivity OD, the stromal opacities did not extend from the limbus as in neovascularization, and there was no stromal scarring or any other stromal opacities in any configuration other than linear branches as evidence of past episodes.

B. Elaborate on the condition: Lattice corneal dystrophy (LCD) is the most common of the stromal corneal dystrophies. There are multiple types of LCD all of which are characterized by amyloid accumulation in the anterior stroma appearing as a linear branching pattern that affects the central cornea and can increase over time. All types are also characterized as generally bilateral.

1. Type 1 is the most common form of LCD and is autosomal dominant in inheritance with onset of symptoms (erosions and blurred vision) at the end of the 1st decade. Recurrent corneal erosions are common and can even precede the characteristic stromal changes. Substantial visual impairment often requiring penetrating or deep lamellar keratoplasty occurs late in the 5th decade. Corneal signs of Type 1 LCD begin with anterior stroma refractile dots that coalesce over time to the characteristic fine, branching lattice lines that start at the center and superficial stroma. The lattice lines spread deeper and more peripherally and are accompanied by progressive stromal haze that causes visual loss.

2. Type II LCD, also known as Meretoja syndrome, also has autosomal dominant inheritance and is associated with systemic amyloidosis. Onset is typically in the 2nd decade and the lattice lines are generally shorter, finer, and more randomly scattered than those in type I and erosions are rare.

3. Type III and IIIA LCD present much later in life with an age of onset of type III in the 4th to 6th decades and type IIIA at the 8th decade. Type III is autosomal recessive in inheritance and type IIIA is autosomal dominant. LCD types III and IIIA are characterized by thicker, rope-like opacities.

C. Expound on unique features: There have been very limited and sporadic reports of unilateral lattice corneal dystrophy. In this case report, the presentation matches that of the classic type I LCD in all aspects except the unilateral presentation.

1. One case series describes 5 cases of unilateral LCD and one case of bilateral LCD in one family. The authors concluded that unilateral LCD differs from the classic type I LCD in that unilateral LCD has late onset, minimal symptoms, and preservation of good vision.1

2. A second, more recent paper presents the case of a 63-year-old man with unilateral LCD presenting with decreased vision, but no history of erosions. They reported that this case represented a unilateral, late-onset variant of LCD related to a mutation of the TGFBI gene.2

3. Another paper presents a different picture than those listed above through three case reports of unilateral LCD that are all indistinguishable
from type 1 LCD including one patient that developed lattice lines in the fellow eye 13 years after he was first examined.³

V. Treatment, management
A. Treatment and response to treatment
1. Bandage contact lens inserted OD, Moxeza qid OD and ibuprofen 400 mg q4-6 hours for pain
2. One drop of homatropine 5% instilled in clinic OD
3. The patient continued with the above treatment and daily office visits for three days until the epithelial defect resolved. Best corrected visual acuity stabilized at 20/50+ OD after resolution of the epithelial defect.
4. The lattice lines and stromal haze between them remained after the resolution of the erosion.
5. The patient was then instructed to use Muro 128 5% ung qhs OD and artificial tears qid to no more than six times a day OD. This patient was seen at a community health facility for patients without insurance, limiting treatment options. The lack of availability of preservative free artificial tears precluded the patient from more frequent dosing of lubrication.

B. Long-term treatment and response to treatment
1. Over the following 10 months, the patient presented on three separate occasions with recurrent corneal erosions.
2. After healing of the latest erosion, treatment was altered due to present research
   a. Doxycycline 50 mg po bid, fluorometholone 0.1% tid, Muro 128 5% ung qhs OD, FreshKote qid OD
   b. There have been no new erosions since his last presentation.
3. Supporting research: Treatment of recurrent corneal erosion with oral doxycycline and topical steroid has been shown to be an effective treatment option that may help patients that have failed other forms of non-surgical treatment. Both doxycycline and topical steroids (methylprednisolone specifically in this study) produce a statistically significant decrease in the number and activity of matrix metalloproteinase-9 in human epithelial cultures.⁴

VI. Conclusion
Unilateral lattice corneal dystrophy has been rarely documented and with variable conclusions as to its place among the classic types of LCD. This case demonstrates a unilateral variant of LCD that is consistent in its presentation to type 1 LCD in its early age of onset, presence of recurrent erosions, and decreased best corrected vision. This case report, along with a literature review of previous reports, suggests that unilateral LCD can either be similar to type I in all aspects except the unilateral nature, or can be a variant that is late-onset without erosions or decreased vision. More genetic research and reports of unilateral LCD are necessary.
Acknowledgements: Sylvia Elizabeth Sparrow, OD, FAAO

Bibliography