Case report

Unilateral lattice corneal dystrophy in a young female: A unique case report

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Abstract

Background: Unilateral lattice corneal dystrophy is a rare entity. **Objective:** To highlight the evidence of unilateral lattice corneal dystrophy in a young female. **Case**: A young 28 years old female presented to the outpatient department of Ophthalmology with slowly progressive diminution of vision in left eye for one month. On ophthalmological examination best corrected visual acuity (BCVA) was 20/20 and 20/40 with refractive error of plano and -0.75D Cyl @30 for right and left eye respectively. Ocular examination of right eye was unremarkable. On slit lamp examination, left eye showed multiple radial lattice lines in branching spider like pattern in the temporal cornea with pupillary margin involvement. The lattice pattern was confined to anterior to midstroma of the cornea with intact epithelium and unremarkable endothelium. The lesions did not involve the limbus. These lattice lines were prominent on retroillumination. In temporal quadrant near pupillary margin a small radial nebulomacular corneal opacity was seen without any corneal vascularisation or edema. The anterior chamber was deep and quiet. Corneal sensations were markedly reduced. Intraocular pressure was 10 and 12mmHg for right and left eye respectively with noncontact tonometry. Fundus examination was unremarkable. Family history and systemic history was negative. Optical coherence tomography(OCT) showed hyperreflective material in midstoma confirmed the diagnosis of unilateral lattice corneal dystrophy(LCD) with an apparently healthy fellow eye.

Key words: Lattice dystrophy, Optical coherence tomography,Autosomal dominant

Introduction

Lattice corneal dystrophy(LCD) is usually bilateral corneal amyloidosis and heredity disorder transmitted in autosomal dominant pattern. It is characterised by refractile lattice lines with a double contour in the corneal stroma. (Manniset al, 1997)LCD is classified into four clinical subtypes. (Stewart et al,2000) LCD type I also known as Biber-Haab-Dimmer is bilaterally symmetrical, autosomal dominant corneal disorder that is characterised by numerous translucent fine lattice lines associated with haze in the superficial and middle layers of the central stroma. The symptoms appear during the first or second decades of life. Progressive diminution of vision can be seen in most of patients which may require penetrating keratoplasty(PK) by age of 40 years. LCD type II is associated with systemic amyloidosis also known as Meretoja's syndrome).The

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lattice lines peripheral in location and are more thicker than those of LCD type I. They extend from the limbus involving mid-stroma towards superficial stroma centrally (Purcell et al 1983). The visual acuity is good until progression of age. LCD type III is a late in onset,commonly asymmetrical and present with diminution of vision in the fifth to seventh decades of life. Lattice lines are thicker and extend from limbus to limbus and are easily visible on direct illumination than that of LCD type I(Hida et al, 1987). LCD type III A differs from the type III by its occurrence in whites, autosomal dominant inheritance pattern and the presence of recurrent erosions.

Unilateral and asymmetrical cases of LCD type I are reported in literature despite generally being considered bilateral and symmetrical (Raab et al, 1974).We report a case of Unilateral case of LCD which appeared clinically like LCD I in a younger female.

Case report

A 28 years old female presented to the us with diminution of vision of her left eye since last one month. It was slowly progressive and painful in nature. On ophthalmological examination best corrected visual acuity (BCVA) was 20/20 and 20/40with refractive error of plano and -0.75D Cyl 30 for right and left eye respectively. Ocular examination of right eye was unremarkable. Extraocular movements were full free and painless for both eyes. On slit lamp examination of her left eye, showed multiple radial lattice lines distributed as network of branching spider like pattern in the temporal part of cornea involving the centre of pupil (Figure 1). The lattice lines seen in the cornea involved the anterior to midstroma of the cornea with clear stroma in between. Endothelium was clear without any deposits or any descemet's striae. The lesions spared the peripheral cornea and did not involve the limbus. These lattice lines were prominent on retroillumination (Figure 2). On fluorescein staining epithelium was intact. A small radial nebulomacular cornea opacity was seen near the pupillary margin on temporal aspect of cornea. The anterior chamber was deep and quiet. Corneal sensations were markedly reduced. No corneal vascularisation or edema was seen. Fundus examination was within normal limits. Intraocular pressure was 10 and 12mmHg for right and left eye respectively with noncontact tonometry.

There was no history of any systemic illness. Family history was negative with parents and siblings.Pachymetry showed Central corneal thickness of 528µm and 490µm for right and left eye respectively. On specular bimicroscoppy endothelial cell density was 1755mm^2 and 1419 mm^2 for right and left eye for respectively. No evidence of polymegathism, pleomorphism or guttae was seen. Keratometry was 45/44@90 and 44/43.25@90 for right andand left eye respectively . On optical coherence tomography there were hyperreflective material seen in midstoma (Figure 3).On the basis of detailed examination diagnosis of unilateral LCD1 dystrophy was made.

Differential diagnosis were prominent corneal nerves in leprosy, keratoconus, syphilis and ghost vessels but there was no continuation seen from the peripheral cornea or limbus.

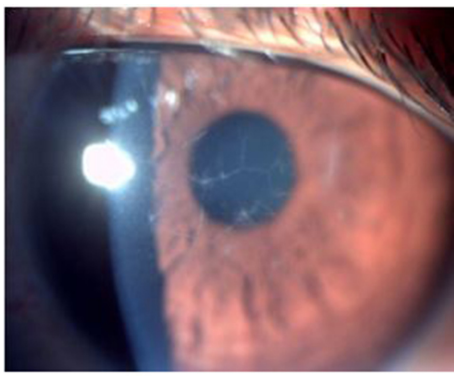

Figure 1: Showing multiple, radially oriented, branching lattice lines

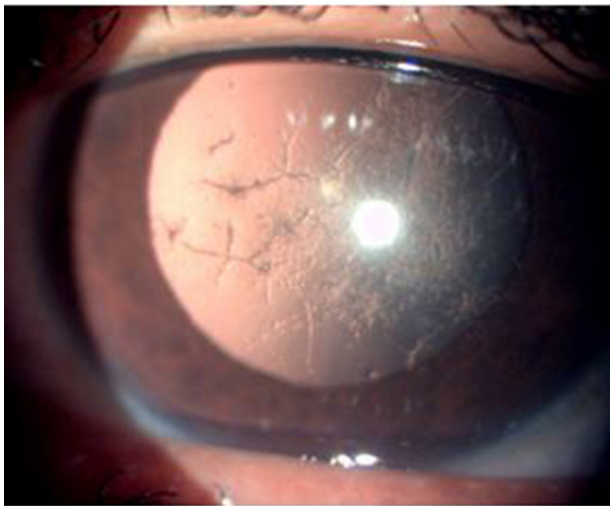

Figure 2: Refractile interlacing lattice lines on retroillumination

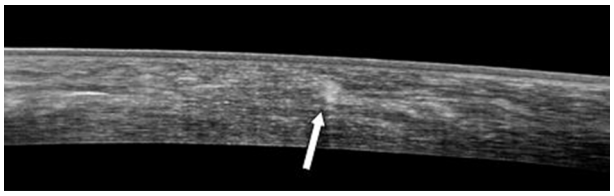

Figure 3: Showing hyperreflecitve material in the midstroma on optical coherence tomography

Discussion

According to Reshmi et al(1971) LCD is classically a bilateral condition. Isolated case reports of unilateral corneal dystrophy have been reported.Raab et al(1974)reported a series of six cases of unilateral LCD classifying them as a sub-group of LCD. All cases involved men between 31–80 years with an average age of 59 years. Most of the patients were asymptomatic with visual acuity of 20/80 or better in the affected eye. Unilaterality, late onset, minimum symptomatology and relatively good visual acuity distinguish these patients from those with classical LCD.

Sridhar et al(2001)reported three cases of unilateral LCD with one patient presented in early third decade of life and two cases in the early fourth decade. Two patients were males and one was female. Clinically the lattice lesions were similar to classic LCD type I. Two of the patients required PK because of poor vision. In one patient, the lattice changes developed in the other eye after 15 years which was sixth case of the unilateral lattice series of Raab et al(1974). The authors concluded that lattice changes can be rarely unilateral and may cause significant visual impairment warranting PK. Lattice lesions may develop many years later in the other eye which should be explained to all patients with apparent unilateral LCD.

Hirano et al(2001) reported two Japanese patients who were clinically diagnosed with late onset sporadic LCD.In one patient, the changes were unilateral with thick lattice lines extending from limbus to limbus in the mid-stromal layer with grey colored nodular deposits in the central cornea were observed. These two cases were caused by the Leu 527 Arg mutation of TGFBI gene.

Sridhar et al (2002)reported a case of unilateral LCD type 1 which presented in the ninth decade.

Sridhar et al(2001) concluded that unilateral LCD with lesions resembling LCD type I can occur in two clinical types. First type presents in late life predominantly in males with minimal symptoms and good visual acuity. The second type occurs in third and fourth decade involving other eye later in life and the visual acuity is impaired warranting PK.

Conclusion

LCD type I is rarely unilateral and occur in males predominantly, presentation is in late life and may cause significant vision loss. In the present case LCD type I was reported in a younger female without any significant visual loss which is quite rare.

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