A CARE-compliant article: ipsilateral progressive idiopathic lipid keratopathy

A case report

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Abstract

Rationale: Lipid deposition on the cornea without previous infection, inflammation, vascularization, or trauma is idiopathic lipid keratopathy. In vivo laser confocal microscopy (IVCM) and anterior segment optical coherence tomography (AS-OCT) are 2 noninvasive methods that can help identify the structures and morphological characteristics of the focus.

Patient concerns: A 63-year-old woman with ipsilateral corneal lipid deposits developing from a small white spot into a yellow-white superotemporal elliptic shape within a year. AS-OCT showed peripheral deep stromal deposits. IVCM showed hyper-reflective material with typical crystalline-like or needle-like structures in the superotemporal area.

Diagnosis: Idiopathic lipid degeneration.

Interventions: Topical steroids eye drops 3 times a day for a month and further consultation every 3 months.

Outcomes: This patient of idiopathic lipid keratopathy was observed every 3 months and till now we have reviewed this patient twice. Topical steroids eye drops were only used during the first month. No further development was observed about the lesion and the patient’s visual acuity remained good.

Conclusion: IVCM and AS-OCT can help identify the characteristic crystalline-like or needle-like hyper-reflective material that could help diagnosis of idiopathic lipid degeneration.

Abbreviations: AS-OCT = anterior segment optical coherence tomography, IVCM = in vivo laser confocal microscopy, KP = keratic precipitate, LDL = low-density lipoprotein.

Keywords: anterior segment OCT, idiopathic lipid degeneration, in vivo confocal microscopy

1. Introduction

Lipid deposition may result in the opacification of cornea and subsequent visual acuity loss. Lipid keratopathy can be idiopathic without any evidence of previous corneal diseases, or also can be secondary to trauma, other corneal diseases, or systemic diseases.\cite{1,2}

There have been few reports about idiopathic lipid keratopathy in previous literature and the majority of cases reported bilateral involvement. Here we present a case of a 63-year-old woman with progressive ipsilateral idiopathic lipid keratopathy, as well as the results from the anterior segment optical coherence tomography (AS-OCT) and in vivo laser confocal microscopy (IVCM).

2. Case report

A 63-year-old Asian woman noticed a white spot on her left eye 12 months ago in the superotemporal periphery, with no discomfort. Since then, the focus had progressively enlarged. She had no previous eye trauma, corneal inflammation, family history of the same symptom, no history of hyperlipidemia or hypertension or other systemic diseases. Other metabolism disorders were not apparent.

Best-corrected visual acuity was 20/20 for the right eye and 20/20 for the left eye. Slit lamp examination showed a yellow-white elliptic focus in the superotemporal periphery cornea with an arcuate pattern focus around it. The central area of the cornea was clear, and the opacification did not cover the pupil under the normal daylight (Fig. 1). No KP was observed and the anterior chamber was clear with normal depth. No other ocular abnormalities were found. AS-OCT revealed a hyper-reflective area in the medium and deep stroma associated with limbus (Fig. 2). In vivo laser confocal microscopy on the edge of lesion (Fig. 3A) showed unclear tissues and cell structures and a few hyper-reflective needle-like structures in medium stroma of left eye. In the central area (Fig. 3B), plenty of hyper-reflective needle-like structures can be observed with no cell structures. No
Figure 1. Photographs of yellow-white elliptic and arcuate pattern of lipid deposition in the superotemporal peripheral cornea with neovascularization in stroma from the limbus.

Figure 2. A, a hyper-reflective area in the medium and deep stroma associated with limbus could be seen. B, At the central area, the cornea got thicker and was hyper-reflective in all layers without clear cornea structures.

Figure 3. A and B, characteristic cholesterol crystals were found at the focus area by in vivo laser confocal microscopy, with hyper-reflective crystalline-like structures in medium and deep stroma, and less in anterior stroma. C, the lack of dendritic cells in the central cornea sub-epithelial layer showed no evidence of previous inflammation of the cornea.
evidence of the inflammation of the cornea was found as there was no dendritic cell in the central cornea sub-epithelial layer (Fig. 3C). No other ocular abnormalities were observed. Systemic evaluation had nothing remarkable.

Diagnosis of idiopathic lipid keratopathy for this patient is based on the lipid deposition on the cornea, and the history of no previous infection, inflammation, vascularization, or trauma. [3,4] The patient was given topical steroids eye drops 3 times a day and asked for further consultation 1 month later. Topical steroids eye drops were only used during the first month. This patient was observed every 3 months and till now we have been reviewed this patient twice. No further development was observed about the lesion and the patient’s visual acuity remained good.

3. Discussion

Most of the previously reported cases of idiopathic lipid keratopathy were bilateral keratopathy, and we reported an ipsilateral one with no history of infection, trauma, inflammation, or vascularization. We also reported results from IVCM and AS-OCT, which were rarely reported in the previous cases.

In vivo laser confocal microscopy showed unclear tissue structure and a few needle-like structures on the edge of lesion and in the central area more characteristic hyper-refractive materials were observed. Figure 3A shows the edge of lesion, and Figure 3 -B shows the central area. Figure 3 -C shows the central cornea sub-epithelial layer, and there’s no evidence of the inflammation of the cornea. These findings supported the histopathological description that the opacity of the cornea was caused by cholesterol deposition in the corneal stroma. [5] and the cholesterol structure fitted the hyper-refractive material with typical crystalline-like or needle-like structures in IVCM, which was important for the diagnosis.

AS-OCT showed that at the central of the lipid deposition, the cornea was thicker and hyper-reflective in all layers and the structure of normal cornea was unclear. At the peripheral area, the focus was located in the medium and deep stroma. The edge was sharp and clear, and the limbus was not affected. In the lesion there was also needle-like structure. These findings clearly described the location of the lipid deposition and could better show the size of focus. From this point of view, AS-OCT can be a good follow-up measurement of the disease.

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If we say IVCM is a microcosmic way to see this disease, then AS-OCT is focus on the macroscopic way relatively. AS-OCT can show the size and scope of the focus, also can reveal that which layer of the cornea has been affected. Therefore it is a good measurement of follow-up to see if the lesion is getting thinner or smaller.

In previous literature, treatments of lipid keratopathy can be various, such as steroids therapy, fluorescein-potentiated argon laser therapy, photodynamic therapy (PDT), fine-needle diathermy (FND), and penetrating keratoplasty (PKP). PDT can eliminate the supplying vessels as well by intravenous injection of a light-sensitive substance and irradiation of the focus with low-power laser. [8] FND is a much cheaper way to occlude the established supplying vessels and corneal micro perforation is a potentially serious adverse event. [9] Finally, treatment for the severe cases with opacification of the visual axis and low visual acuity is penetrating keratoplasty (PKP). But PKP can also cause graft rejection diseases, thus converting this treatment to a high-risk procedure. [10] For this patient, considered of her own financial problem, we first used typical steroids eye drops, and we will close follow-up the development of the focus and to see if further treatment is needed.

The nature of idiopathic lipid keratopathy is not clear yet. Some researchers suggested that there might be some low-grade inflammation at the corneal-scleral limbus or ocular surface sometimes not noticed by patients. Functional defect of the vascular endothelial cells with leakage of cholesterol into adjacent corneal tissues have been hypothesized as pathogenic mechanisms in idiopathic lipid keratopathy. [1,3,7] Rabbit models revealed that the lipid accumulation occurred probably due to the uptake of plasma low-density lipoprotein (LDL) by keratocytes in excess of the cellular capacity to metabolize or excrete the lipids, and then the lipids were retained in the keratocytes as non-membrane-limited droplets of cholesterol esters. [11] In Eduardo Alfonso case, histochemical analysis of the corneal lipid focus revealed the presence of neutral fats, free fatty acids, cholesterol, and a small amount of phospholipids. [12] More cases are needed to clarify these pathophysiological mechanisms.

Author contributions

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