An Unusual Presentation of Chandler Syndrome: Report of A Case and Brief Discussion

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Abstract

***Purpose*:** To report an unusual presentation of chandler syndrome

***Case report*:** A 22-year-old female came to our clinic with complaint of progressive visual loss in her left eye since three days ago. She had a history of trauma with finger-nail to her left eye eight days ago and had been managed with topical antibiotic. In her examination, the left eye had visual acuity (VA) of counting finger at three meters, with intraocular pressure (IOP) of 14 mmHg, and there was a 3+ corneal stromal edema without epithelial defect. After medical management when edema decreased, specular biomicroscopy revealed decreased endothelial cell count and increased average cell size, pleomorphism and polymegathism, and epithelial transformation in the left eye and normal indices in the right eye. After one month, edema was completely resolved and VA improved to $^{20}/_{25}$ but specular biomicroscopy remained abnormal. Chandler variant of iridocorneal endothelial syndrome (ICE) was diagnosed.

***Conclusion*:** Although corneal edema after blunt trauma is a usual manifestation, in long lasting edema after trauma, we should consider other possible endothelial causes of stromal edema including toxic insult, inflammatory processes, and endothelial dystrophies.

***Keywords*:** Trauma, Iridocorneal Endothelial Syndrome, Chandler's Syndrome

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Introduction

Iridocorneal endothelial syndrome (ICE), which is an epithelial transformation of corneal endothelium, almost always is a unilateral disorder. Confocal microscopy shows some epithelial-like multilayered endothelial cells with nuclear hyperreflectivity. It has a probable association with HSV and presents as three different syndromes depending on the extent of its ocular involvement.

Chandler's subtype affects young to middle aged females. It is confined to corneal inner surface and can involve angle structures by migration of this abnormal epithelial-like layer. These patients may show corneal edema due to endothelium dysfunction, but their important feature is glaucoma due to outflow obstruction. Here we present an unusual and early presentation of this syndrome in a young girl, with severe corneal edema after blunt trauma.

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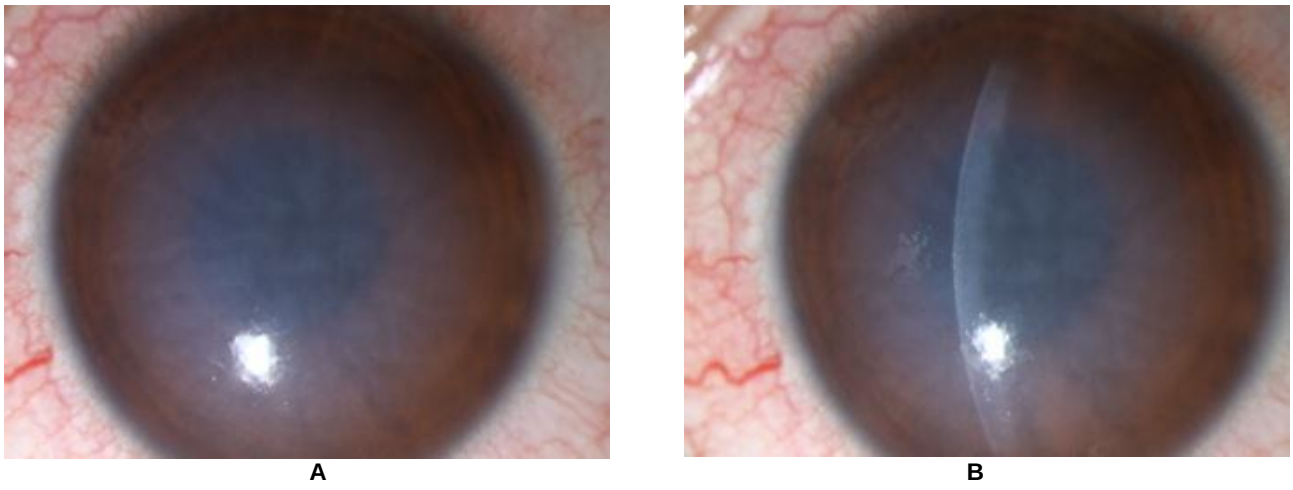
Case report

A 22-year-old female came to our clinic with complaint of progressive visual loss in her left eye since three days ago. She had a history of trauma with finger-nail to her left eye eight days ago and had been managed with topical antibiotic (Ciplex) in another center. She declared she had no previous ocular surgery.

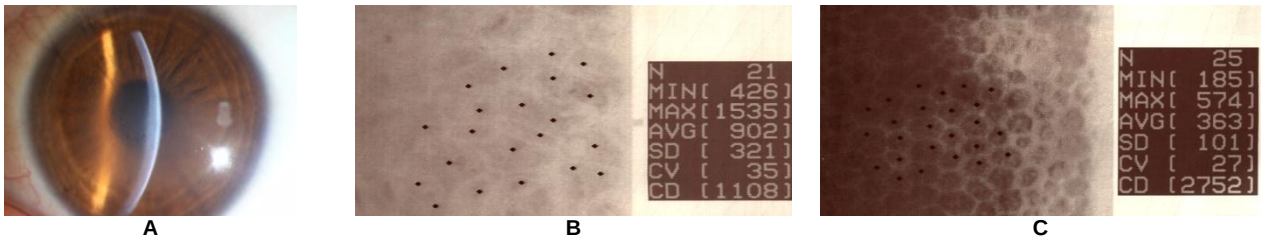
In her examination, the left eye had visual acuity (VA) of counting finger at three meters, with intraocular pressure (IOP) of 14 mmHg, and there was a 3+ corneal stromal edema with no epithelial defect. Anterior segment showed no obvious sign of iridocyclitis and posterior segments was poorly visible (Figures 1A, 1B). Reverse afferent pupilary activity was normal. Right eye had VA of $^{20}/_{20}$, IOP of 16 mmHg and all anterior and posterior findings were normal.

Topical Ciplex was discontinued, topical and systemic steroid was started in addition to topical Timolol and Nacl 5% drops. After four other days, edema decreased and VA improved to $^{20}/_{50}$. Posterior segment examination was performed and showed no abnormal finding. Specular biomicroscopy revealed decreased endothelial cell count and increased average cell size, pleomorphism and polymegathism in the left eye and normal indices in the right eye (Figures 2A, 2B, 2C). After one month, edema was completely resolved and VA improved to $^{20}/_{25}$ but specular biomicroscopy remained abnormal (Figure 3). Confocal microscopy was performed and showed epithelial like endothelium in the left cornea (Figure 4A, 4B). On retrograde detailed examination of the angle, no abnormality was observed. As it was a unilateral condition, although the patient had no angle abnormality in gonioscopic exam yet, an early stage Chandler's syndrome was considered as the diagnosis. The patient is under regular observations for any progression, angle involvement or glaucoma formation.



A B

Figure 1. Diffuse (A) and Slit photograph (B) of the patients left eye showing stromal edema



A B C

Figure 2. Slit photograph of the patients left eye showing decreased corneal edema after four days of medical treatment (A) Specular microscopy at midperiphery of the same eye at the same time, showing reduced cell density (B), and increased average cell size and its coefficient of variation while having normal parameters in the right eye (C)

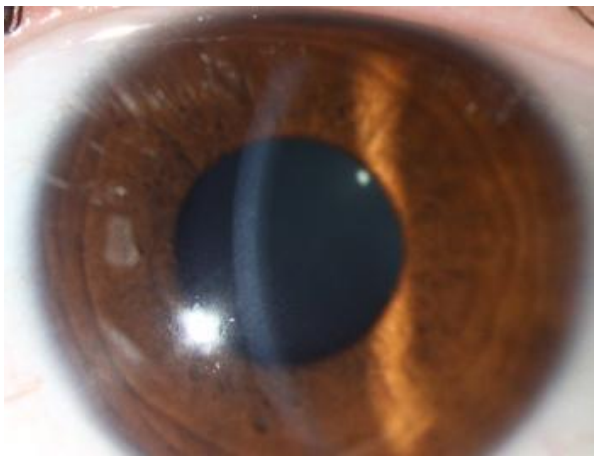
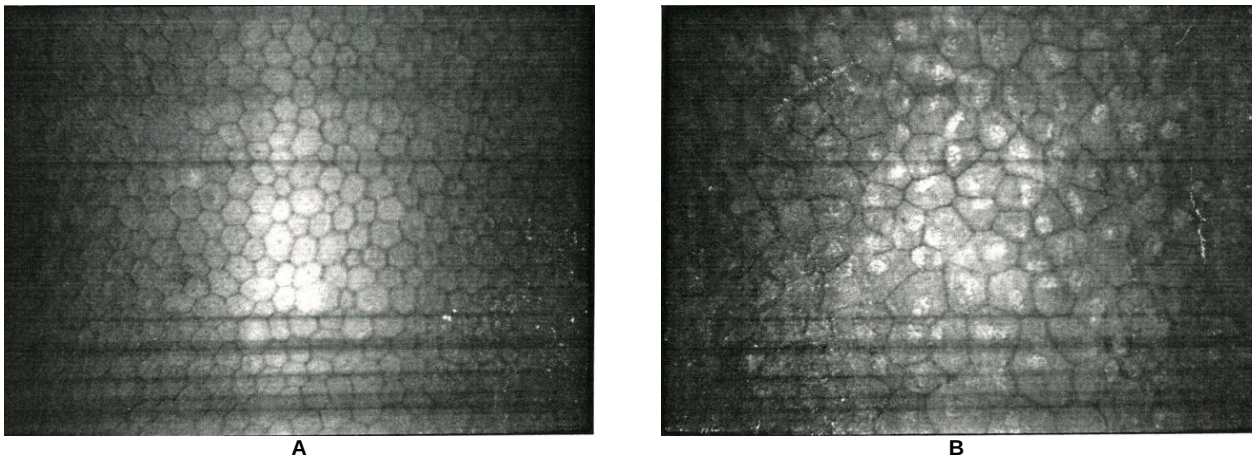


Figure 3. Slit photograph of the same eye showing completely resolved stromal edema after one month of follow up.

On confocal scan of right cornea, all layers from epithelium up to endothelium look unremarkable and endothelial cell density is 3003.8 cell/mm^2 with mean cell area of 332.9 µm^2. On the left side epithelioid metaplasia of endothelial cells with large polygonal morphology and apparent nucleoli is noted with cell density of 1120.3 cell/mm^2 and mean cell area of 892.6 µm^2. Also included is a diffuse scattered inflammatory cell infiltration. No typical acanthamoeba or fungal elements are seen.
Given the confocal scan features, "iridocorneal endothelial (ICE) syndrome" involving left eye with superimposed non-specific keratitis is suggested.



A B

Figure 4. Confocal scan of the patient's right eye after one month of follow-up, showing normal appearance of endothelial cells (A) Abnormal large polygonal cells were found in the left eye with epithelial metaplasia at the same time (B).

Discussion

Although corneal edema after blunt trauma is a usual presentation, in persisting edema after trauma with an intact epithelium, we should also consider other possible endothelial causes of stromal edema. These include toxic insult, inflammatory processes, and endothelial dystrophies.

Ocular blunt trauma

It has been reported that the traumatic eye, compared with the unaffected fellow eye, can have a decrease of 1.2% in endothelial cell density (ECD) in patients without angle recession, and up to 21.2% decrease in patients with more than 180° angle recession.[1] Our patient had a more pronounced decrease in ECD and no angle recession in gonioscopic exam.

Toxic insult

There are many known toxic substances as preservatives in surgical solutions which can

cause damage to endothelial cells when used intraocular.[2,3] Some systemic medications such as Amantadine have also been reported to cause corneal edema.[4] Long-term topical epinephrine hydrochloride has been associated with decreased ECD when used in patients with ocular hypertension.[5] Our patient had no medical history except a short-term (7 days) usage of topical Ciplex.

Inflammatory processes

Endothelitis can cause endothelial dysfunction and corneal edema. It is mostly associated with herpes virus and has its own typical features. Our patient had no symptom or sign suggestive of possible keratouveitis.

Endothelial dystrophies

This group of disorders can lead to corneal edema, in their course or after an external precipitating factor such as trauma or raised IOP, due to endothelial pump dysfunction. These can be differentiated by their pathognomonic features in biomicroscopic imaging like confocal scan. Fuchs' endothelial dystrophy (FED) is slightly more common in women, usually begins in 4-5th decade of life and affects both eyes. In pathology, Descemet's membrane is grossly thickened with accumulation of abnormal wide-spaced collagen and numerous guttae which can be visible with retro-illumination. Endothelial cells are progressively reduced in number and attenuated in end stage leading to corneal edema; this process may be aggravated or accelerated by intraocular trauma or surgery. On Confocal microscopy, gutta can be seen as elevations on endothelial cells. Posterior polymorphous corneal dystrophy (PPCD) is also a bilateral -but asymmetric- condition affecting younger patients. In Specular microscopy Descemet membrane has characteristic vesicular and band-like changes, endothelial cell count is mildly diminished with varying amounts of polymegathism and pleomorphism; they show characteristics of epithelial cells with prominent round nuclei and numerous projections (microvilli) and sometimes consist of more than one cell layer (transformed into stratified squamous epithelium, in 60%). These patients usually present with decreased vision due to corneal edema. Patients with Chandler's subtype of ICE-syndrome may also show corneal edema due to endothelium dysfunction. As our patient had a unilateral condition, an early stage of Chandler's syndrome was considered as the diagnosis. Trauma has been a stressor and so a precipitating factor to decompensate endothelium causing corneal edema. There have been few reports on traumatic corneal edema being first presentation of some dystrophies like FED. But, this is the first report accounting trauma as a possible precipitating factor for cornel edema in Chandler syndrome. Once again, we highlight the important role of confocal scan in helping to make the diagnosis.

Conclusion

Although corneal edema is a usual manifestation after blunt trauma, in long lasting edema with intact epithelium we should consider other possible endothelial causes of stromal edema including toxic insult, inflammatory processes, and endothelial dystrophies. With the help of careful history taking and diagnostic cellular imaging we can come to the right diagnosis.

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