**CASE REPORT**

**Keratoconus with Corneal ‘Pips’**

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**ABSTRACT**

Keratoconus is a common ectatic corneal disorder. There are several causes of corneal opacity in keratoconic patients. This case illustrates two different causes, in the same patient, in either eye, and diagnostic imaging characteristics on Fourier-domain optical coherence tomography (FD-OCT) that aided surgical management.

**Keywords:** Contact lens intolerance, Fourier-domain optical coherence tomography, Keratoconus, Proud nebula

**CLINICAL PRESENTATION**

A 26-year-old Pakistani woman, a known case of keratoconus, presented with poor vision in both eyes. The patient had no history of eye surgery, trauma or systemic disease and was intolerant of contact lenses. At presentation, her best corrected visual acuity (BCVA) was 6/45+1 OD (right eye) and 6/15 OS (left eye).

Slit-lamp biomicroscopy revealed bilateral steepening of the cornea inferiorly, with opacities near the apex of both keratoconic cones. The corneal opacities appeared dense, but in the left eye, they were smaller in size and more superficial in location (Fig. 1).

**FD-OCT FINDINGS**

Fourier-domain optical coherence tomography (FD-OCT, RTVue; Optovue, Inc., Fremont, CA) sections of the opacity in the left cornea revealed the lesion to be above the level of Bowman’s membrane (corneal ‘pip’ or proud nebula). A similar nebula-like lesion was also seen on the right eye but additionally included underlying areas of scarring, extending beyond Bowman’s membrane (Fig. 1) (see overleaf).

The pachymetry map of FD-OCT showed that both corneas were thin (353 μm OD, 424 μm OS) (Fig. 2) (see overleaf). In the central 5 mm region of each eye, the asymmetry in thickness was more negative than −45 μm between pairs of I-S octants (the average thickness of the inferior [I] octant minus the average thickness of the superior [S] octant), and likewise for the IT-SN octants (the average thickness of the inferotemporal [IT] octant minus the average thickness of the superonasal [SN] octant). Furthermore, the minimum-maximum difference was more negative than −100 μm in either eye. These findings fulfill all the OCT diagnostic parameters for keratoconus1, 2.

**DIAGNOSIS**

Bilateral keratoconus, complicated by corneal pip in the left eye, and corneal pip and stromal scarring in the right eye.
TREATMENT
The patient underwent a surgical superficial keratectomy to the left eye. Under topical anaesthesia, the corneal opacity was successfully cleared after excision of overlying corneal epithelium and careful dissection of the plane above Bowman’s membrane. Postoperative BCVA of the left eye was 6/7.5+1.

The right eye underwent deep anterior lamellar keratoplasty (DALK) resulting in a BCVA of 6/18^2 one week postoperatively.

DISCUSSION
We described a case of bilateral keratoconus, complicated by contact lens intolerance and corneal opacity from a corneal ‘pip’ or proud nebula, imaging characteristics on FD-OCT and clearance via surgical superficial keratectomy.

Keratoconus is a bilateral, usually asymmetrical^1 corneal dystrophy affecting corneal shape, characterised by progressive corneal steepening, most typically inferior-temporal to the centre of the cornea^4, with corneal thinning, induced myopia, astigmatism (regular and irregular), and in more advanced cases, may present with hydrops, or scarring from previous hydrops^5.

Proud nebulae or corneal ‘pips’ are usually observed in the context of contact lens use in keratoconic patients, and are rarely seen in normal subjects. It is a localised area of raised corneal tissue, usually sited at or near the apex of the keratoconic cone.
Thought to have a histologic resemblance to Salzmann degeneration, the raised profile of the proud nebula can cause difficulties in contact lens fitting, rendering its use intolerable, or rarely on its own, obscure vision\(^6\). In this circumstance, visual rehabilitation is complicated by both contact lens intolerance and corneal haze.

There are a few published reports on the histopathologic characteristics of proud nebulae/nodules and to our knowledge, there are no published materials on the FD-OCT appearance of a proud nebula in keratoconus.

Histologic examination of these fibrotic nodules revealed randomly arranged fibrillar collagen interspersed with few fibrocytes. The overlying epithelium is described as fragmented with discontinuous, and multilaminar basement membrane and diminished hemidesmosomes\(^7\).

The FD-OCT characteristics of high signal-to-noise ratio coupled with a high depth of resolution, and high speed, allowed for the precise identification and differentiation of key corneal anatomical landmarks. Here, the FD-OCT (RTVue; Optovue, Inc., Fremont, CA) offers a distinct advantage over Time-Domain OCT systems. The RTVue used in this case is 13 times quicker and features a depth resolution that is over three times higher (5 μm) than the fastest commercial TD-OCT system, the Visante anterior segment OCT (Carl Zeiss Meditec, Inc., Dublin, CA)\(^2,8\).

The principle of management of proud nebulae entails the removal of these sub-epithelial lesions to improve contact lens tolerance. For more superficial scars, DALK and penetrating keratoplasty are not indicated and several treatment alternatives have been reported. They include surgical superficial keratectomy and eximer laser phototherapeutic keratectomy (PTK)\(^7,9\).

CONCLUSION
In instances whereby a corneal pip or proud nebula impedes visual rehabilitation both through contact lens intolerance and the presence of significant corneal scarring, and when surgery or laser is contemplated, the FD-OCT can be an invaluable aid to the surgeon in choosing the most appropriate modality of treatment.

REFERENCES