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PHOTO ESSAY

Deep blue dot corneal degeneration: confocal characteristics

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Abstract
Purpose To discuss the clinical features, differential diagnosis and the novel confocal microscopic findings noted in the rare 'deep blue dot corneal degeneration'.
Methods Observational case report.
Results Slit-lamp biomicroscopic examination revealed bilateral, numerous, circular to oval discrete blue opacities at the level of deep stroma and fine grey linear opacities at the level of mid to deep stroma. Confocal microscopy demonstrated two types of corresponding hyper-reflective extracellular lesions: oval deposit-like, most concentrated at a depth of 430–480 µ and needle-like at the depth 330–370 µ.
Conclusions Deep blue dot corneal degeneration is a rare entity where blue deposits of amyloid are seen in the deep corneal stroma. It should be considered as a differential diagnosis when an old-aged person presents with good vision and the above mentioned findings.

Keywords Deep blue dot corneal degeneration · Confocal microscopy · Hyper-reflective lesions · Needle-like · Oval dense deposits · Extracellular deposits

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Slit-lamp bio-microscopic examination of a 60-year old male with best corrected visual acuity of 6/12 OU revealed bilateral, numerous, circular to oval discrete medium-sized blue opacities at the level of deep stroma, most concentrated at the centre. Fine grey linear opacities were also noticeable at the level of mid- to deep stroma (Fig. 1). The differential diagnosis included deep blue dot corneal degeneration, cornea farinata, fleck corneal dystrophy, pre-Descemet dystrophy and Cogan microcystic corneal dystrophy [1–5]. Confocal microscopy characterised the lesions as extracellular deposits. Needle-like hyper-reflective opacities were visible roughly at the level of 180 µm, which increased in number up to a depth of 330–370 µm and then gradually disappeared (Fig. 2). Oval dense deposits which became noticeable at around a depth of 250 µm, gradually increased in number with depth, attaining a peak at around 430–480 µm (Fig. 3). The endothelium was normal.

Comment

The age of presentation and the clinical features, being predominantly bluish, discrete, medium–large-sized and central distribution, best support the diagnosis of 'deep blue dot corneal degeneration' [1]. In contrast to the above, cornea farinata shows fine dust-like opacities [2]. The pre-Descemet dystrophy exhibits focal

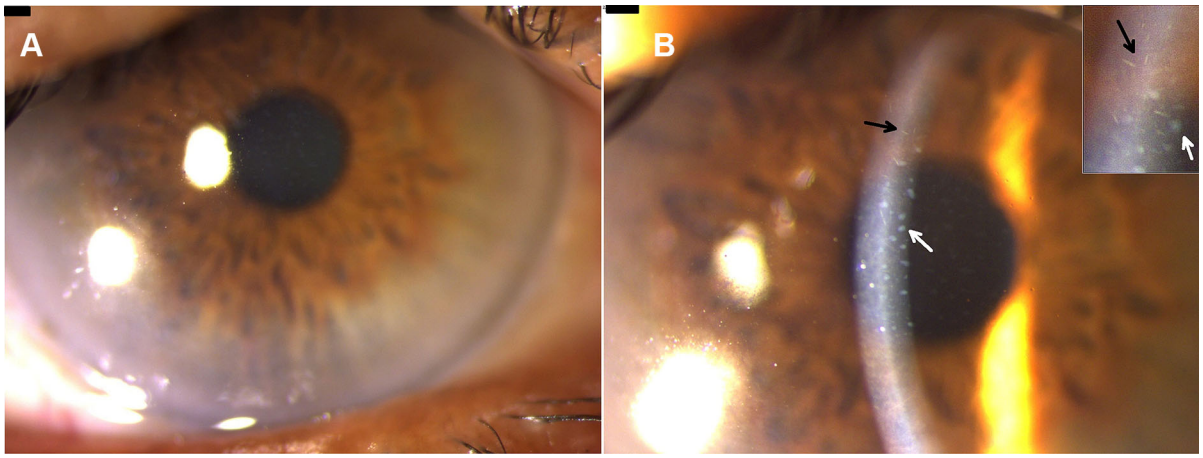

Fig. 1 Slit lamp bio-microscopic photography. **a** Diffuse illumination shows faintly visible blue-coloured scattered opacities on the central cornea, **b** direct focal illumination shows grey coloured needle-like lesion in the mid-stroma (black arrow) and blue-coloured discrete oval lesions at deep stroma (white arrow). Inset shows a magnified picture of the same

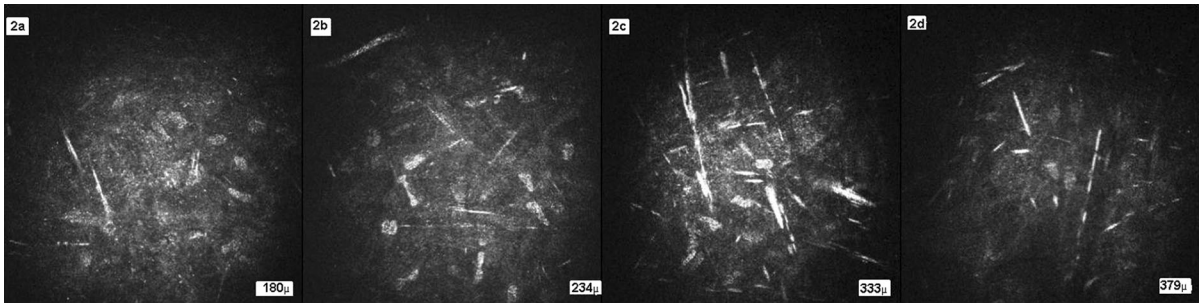

Fig. 2 Corneal confocal microscopy sections showing needle-like hyper-reflective lesions at various levels. **a** At 180 µm, scarce lesions visible, **b** at 234 µm, gradual increase in number of lesions perceptible, **c** at 333 µm, high density of lesions clearly seen, **d** at 379 µm, the number of lesions appears to start reducing gradually

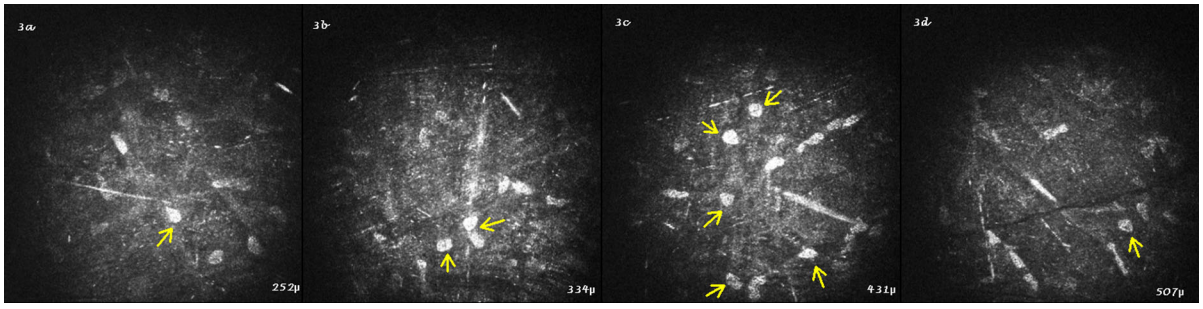

Fig. 3 Corneal confocal microscopy sections showing hyper-reflective oval dense deposits at various levels. Lesions have been marked by yellow arrow to aid differentiation from keratocyte nuclei. **a** At 252 µm, the lesions first appear, **b** at 334 µm, gradual increase in number is noticeable, **c** at 431 µm, high density of oval lesions is observed, **d** at 507 µm, a decrease in number of lesions is seen

polymorphic grey opacities, while the fleck corneal dystrophy shows translucent grey-white flour-like opacities over the entire stroma [3, 5]. Cogan microcystic corneal dystrophy can exhibit occasional blue flecks; however, they lie on the anterior cornea [4].

The confocal microscopic characteristics of this case are unique as compared to the expected features of its clinical differential diagnoses. Cornea farinata and fleck corneal dystrophy are known to exhibit intracellular reflective particles, the former in the deep stroma [2] and the latter scattered throughout the stroma [3]. Pre-Descemet dystrophy typically has intra- and extracellular deposits immediately above the Descemet membrane [3]. The blue flecks of Cogan microcystic dystrophy have been proven to be subepithelial [4]. Strikingly different from the above, our case displayed two types of hyper-reflective extracellular lesions, needle-like at the level of mid-stroma and oval deposit-like at the level of deep stroma.

This typical clinical picture and unique confocal microscopy findings characterise the 'deep blue dot corneal degeneration', which have been histologically proven to consist of amyloid [1].

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Research involving human participants The procedures performed in this report involving human subject were in accordance with the ethical standards of the institutional research committee and with the 1964 Declaration of Helsinki and its later amendments.

Patient consent The patient has consented for the submission of the case report to the journal.

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