In vivo confocal microscopy in different types of posterior polymorphous dystrophy

Kalpana Babu, *MRCOphth;* ***K R Murthy,*** *FRCS*

Posterior polymorphous dystrophy is a rare corneal dystrophy, usually detected by chance. This case series describes the morphologic features in the three different types of posterior polymorphous dystrophy using confocal microscopy.

Key words: Confocal microscopy, posterior polymorphous dystrophy.

Indian J Ophthalmol 2007;55:376-8

Posterior polymorphous dystrophy is a rare, often innocuous, autosomal dominant and usually asymptomatic corneal dystrophy which may frequently be diagnosed by chance in later life.[1] It is asymmetrical and consists of three types: vesicular, band-like and geographic (placoid) patterns. The principle of confocal microscopy allows us to study the morphology of the cornea *in vivo*. This series describes the morphologic features in different types of posterior polymorphous dystrophy using confocal microscopy using the Rostock corneal module of the Heidelberg retinal tomograph II (Heidelberg Engineering, Germany). A Tomey pachymeter (Tomey Corporation, Japan) was used to obtain the central corneal thickness.

Case Reports

Case 1

A 28-year-old asymptomatic male was diagnosed on routine examination to have a vesicular type of posterior polymorphous dystrophy in both eyes by slit-lamp examination. The best-corrected visual acuity was 20/20 (Snellens) in both eyes. White flecks with guttae were seen involving the Descemet's membrane and endothelium [Fig. 1A]. Central corneal thickness measurements were 570 μm and 566 μm in the right and left eyes respectively. Confocal microscopy findings included vesicular abnormalities ranging from 10 to 160 μm. Endothelial cells were visible within some vesicular lesions [Fig. 1B]. Patchy hyperreflective lesions were noted at the level of the Descemet's membrane and around the vesicular lesions [Fig. 1C]. The endothelial densities were 1757 ± 40 cells/mm^2 and 1497 ± 51 cells/mm^2 (normal: 2218 to 4434 cells/mm^2)[1] in the right and left eyes respectively.

Case 2

A 72-year-old asymptomatic male was diagnosed to have a band type of posterior polymorphous dystrophy in the left eye on slit-lamp examination [Fig. 2A]. The best-corrected visual acuity was 20/30 (Snellens) in both eyes. Clinically, his right eye appeared normal. Central corneal thickness measurements were 526 μm and 611 μm in the right and left eyes respectively. Confocal microscopy findings in both eyes showed endothelial

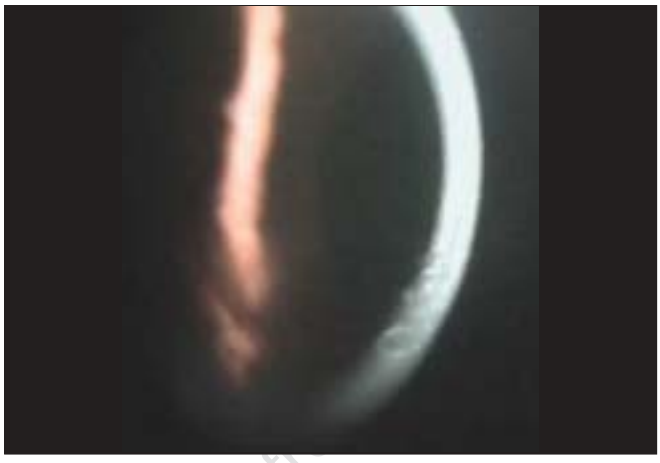

Figure 1A: Slit-lamp photograph showing vesicular type of posterior polymorphous dystrophy

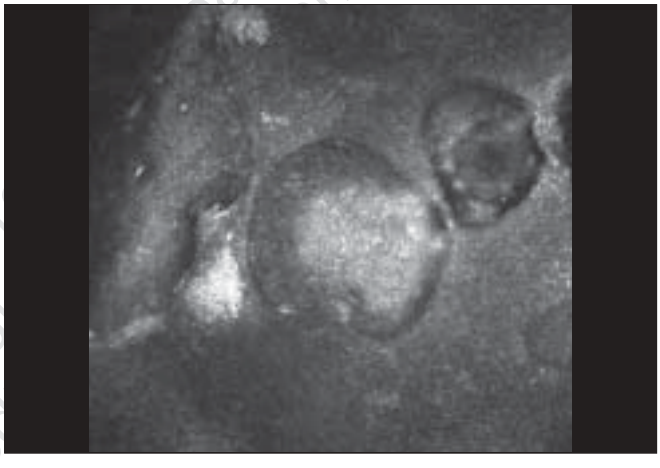

Figure 1B: Confocal microscopy showing vesicles with abnormal endothelial cells within them

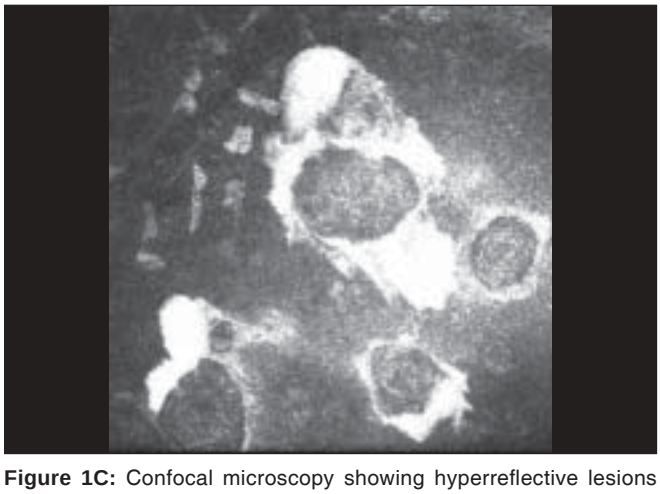

Figure 1C: Confocal microscopy showing hyperreflective lesions around the vesicles

Prabha Eye Clinic and Research Center, Bangalore and Vittala Institute of Ophthalmology, Bangalore, India

Correspondence to Dr. Kalpana Babu, Prabha Eye Clinic and Research Center, 504, 40th cross, Jayanagar 8th block, Bangalore - 560 070, India. E-mail: kb_7375@yahoo.co.in

Manuscript received: 03.03.06; Revision accepted: 03.10.06

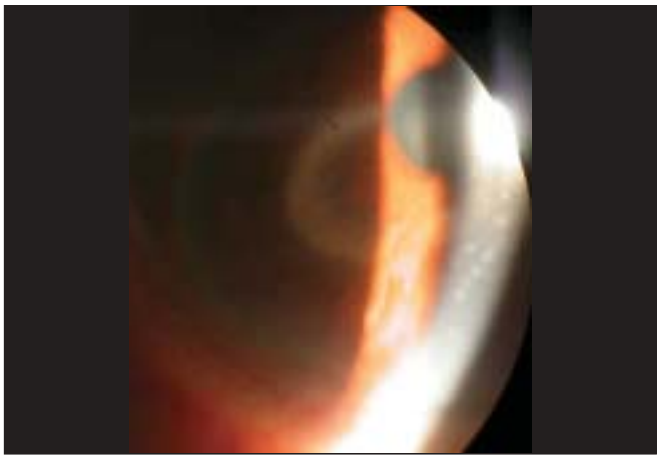

Figure 2A: Slit-lamp photograph showing band type of posterior polymorphous dystrophy

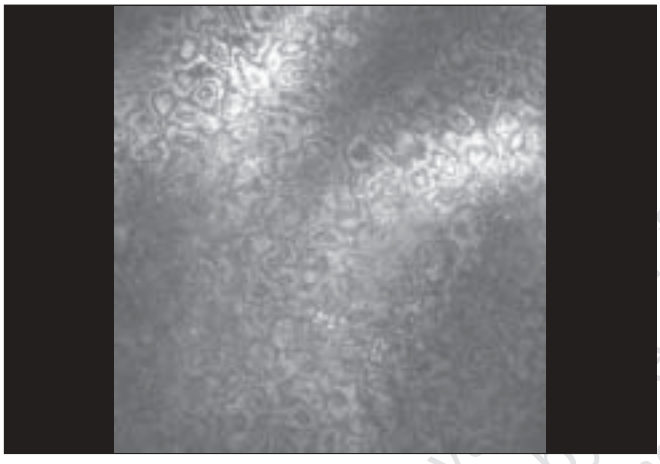

Figure 2B: Confocal microscopy showing endothelial polymorphism. Occasional bright endothelial nuclei also seen

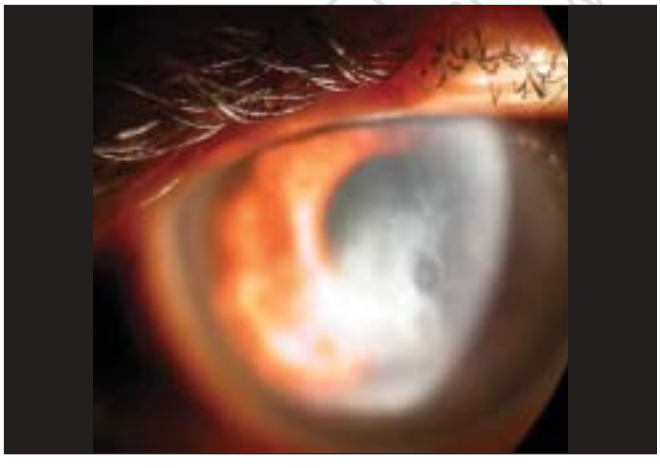

Figure 3A: Slit-lamp photograph showing geographic pattern of posterior polymorphous dystrophy

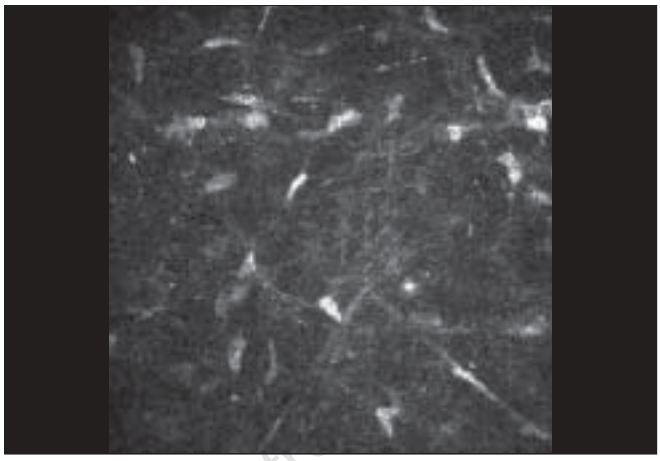

Figure 3B: Confocal microscopy showing needle like lesions in the stroma with few keratocytes suggestive of scarring

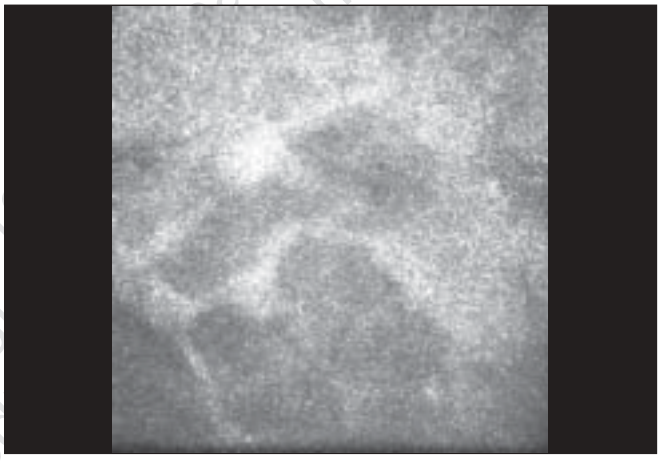

Figure 3C: Confocal microscopy showing large lacunae with surrounding indistinct endothelial cell borders

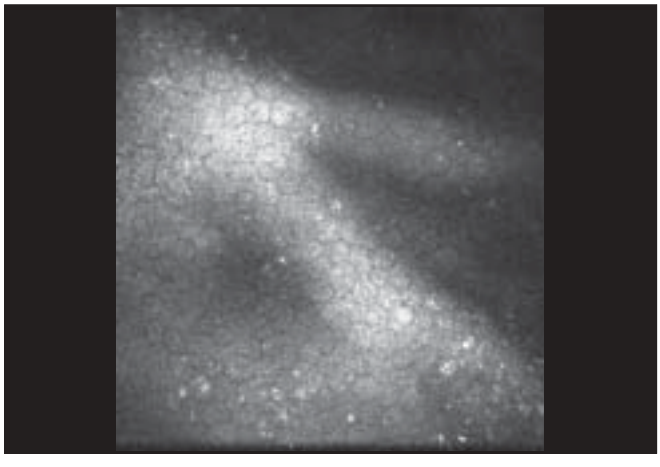

Figure 3D: Confocal microscopy showing dot-like hyperreflective lesions in the endothelium involving the other eye

polymorphism and polymegathism with folding and scalloping of the endothelial cell borders [Fig. 2B]. Occasionally, bright endothelial nuclei were noted. The endothelial densities were 2056 ± 66 cells/mm^2 and 1609 ± 40 cells/mm^2 (normal: 2218 to 4434 cells/mm^2)[1] in the right and left eyes respectively.

Case 3

A 64-year-old symptomatic woman with decrease in vision was diagnosed to have a geographic pattern of posterior polymorphous dystrophy in the right eye on slit-lamp examination [Fig. 3A]. Her best-corrected visual acuity was

counting fingers three meters and 20/60 (Snellens) in the right and left eyes respectively. Clinically, her left eye was normal. Central corneal thickness measurements were 668 μm and 559 μm in the right and left eyes respectively. Confocal microscopy findings in the right eye showed needle-shaped lesions suggestive of scarring in the stroma [Fig. 3B]. Large vesicles were noted in the endothelium. The borders of the endothelial cells appeared indistinct [Fig. 3C]. The left eye showed plenty of hyperreflective dot-like lesions in the endothelium and polymegathism [Fig. 3D]. The endothelial cells could not be counted in the right eye due to indistinct cell margins while the endothelial cell density in the left eye was 1541 ± 45 cells/mm^2 (normal: 2218 to 4434 cells/mm^2).[1]

Discussion

In vivo confocal microscopy allows observations of the living human eye at a cellular level. It is based on the principle that the illumination (condenser) and observation (objective) systems are focused on a single point (have common focal points).[2] This eliminates out of focus information and brings a lateral resolution of 1-2 μm and an axial resolution of 5-10 μm increasing the magnification up to 600 times. Findings such as cracks, vesicles, craters have been described in general in posterior polymorphous dystrophy.[3,4] This case series delineates the morphological features seen on confocal microscopy in each type of posterior polymorphous dystrophy.

In case 1, both eyes showed similar findings on slit-lamp and confocal microscopy examinations. The presence of vesicles and patchy hyperreflective lesions at the level of the Descemet's membrane and around the vesicles corresponded to the guttae and white flecks seen clinically.

In case 2, although band-shaped areas were seen clinically only in the left eye, the right eye also showed similar changes on confocal microscopy. The confocal microscopy findings of the folding and scalloping of the endothelial cell borders represent edges of individual vesicles that might have fused to give the above appearance.[5]

In case 3, clinically evident geographic pattern was noted in the right eye. The confocal microscopy findings of needle-shaped lesions in the stroma with large vesicles in the endothelium and indistinct cellular margins indicate the corneal edema with scarring seen in these eyes. However, certain changes like dot-like hyperreflective lesions were also seen in the left eye indicating abnormal changes in the endothelium.

Confocal microscopy thus enhances the clinicopathological diagnosis and aids in the follow-up of corneal dystrophies with subtle clinical presentations.

References

1. Vanathi M, Tandon R, Sharma N, Titiyal JS, Pandey RM, Vajpayee RB. *In vivo* slit scanning confocal microscopy of normal corneas in Indian eyes. *Indian J Ophthalmol* 2003;51:225-30.
2. Guthoff RF, Stave J. *In vivo* micro morphology of the cornea: Confocal microscopy principles and clinical application. *In*: Reinhard T, Larkin F, editors. Cornea and External eye disease. 16th ed. Springer; 2006. p. 174-81.
3. Cheng LL, Young AL, Wong AK, Law RW, Lam DS. Confocal microscopy of posterior polymorphous endothelial dystrophy. *Cornea* 2005;24:599-602.
4. Patel DV, Grupcheva CN, McGhee CN. *In vivo* confocal microscopy of posterior polymorphous dystrophy. *Cornea* 2005;24:550-4.
5. Laganowski HC, Sherrard ES, Kerr Muir MG. The posterior corneal surface in posterior polymorphous dystrophy: A specular microscopical study. *Cornea* 1991;10:224-32.