

History

- A five-month-old baby was seen in clinic with bilateral congenital corneal opacities. The right cornea is shown in Figure 1.
- Examination revealed corneal oedema and opacities of Descemet's membrane and endothelium with a few vesicular lesions.
- Corneal transplantation was performed and specimen sent for ophthalmic histopathological and ultrastructural assessment.



Figure 1

Questions

- What are the clinical differential diagnoses for congenital diffuse corneal haze?
- Figure 2 represents PAS and H&E stained sections of the posterior aspect of the cornea. What are they showing?
- In Figure 3 the top image is an immunohistochemistry section for AE1/AE3. What is it demonstrating and which alternative markers could also be used?
- Figure 4 represents EM sections of the posterior corneal surface. What are the most relevant features?
- Considering the morphological features, what is the most likely diagnosis?

SECTION EDITOR



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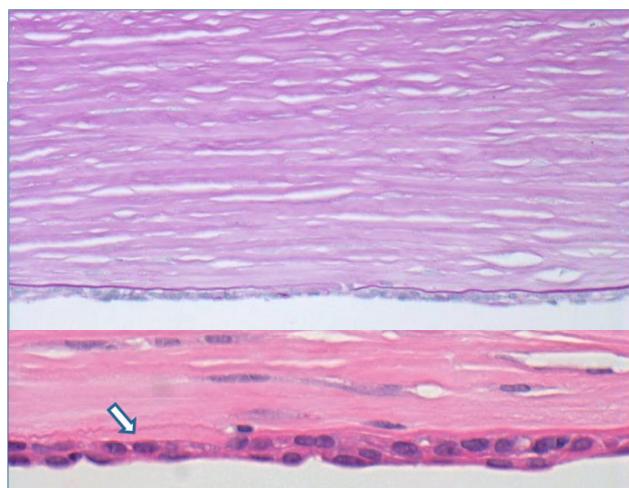


Figure 2

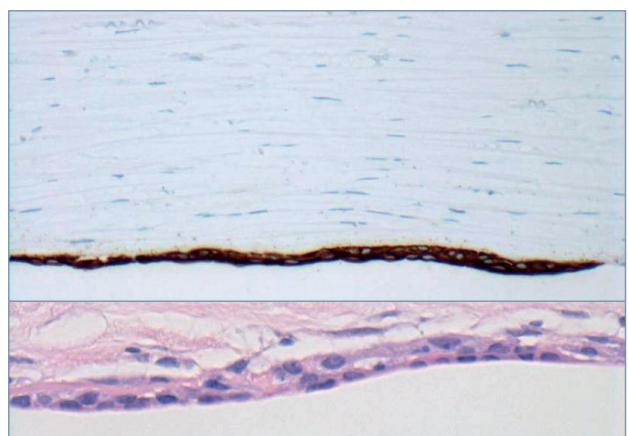


Figure 3

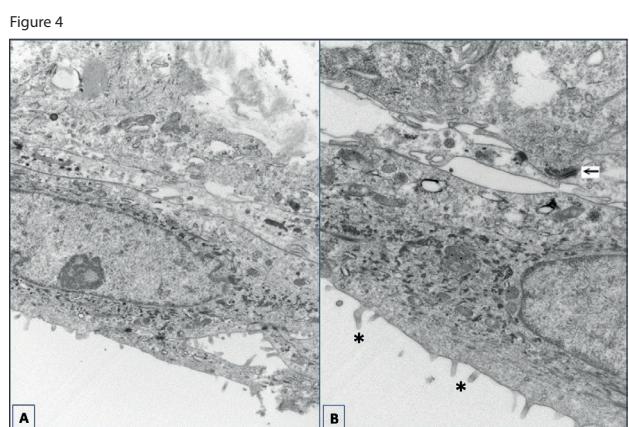


Figure 4

- Posterior polymorphous corneal dystrophy.
- These are epithelial characteristics in the abnormal cornea. Arrow is pointing to a desmosome and the stars indicate microvilli.
- Figure 4A demonstrates multilayered endothelium. In Figure 4B the another area of the multilayered endothelium.
- Figure 4B demonstrates multilayered endothelium. In Figure 4B the arrow is pointing to the abnormal endothelium.
- Posterior polymorphous corneal dystrophy.

- CK8, CK18, and CK19. The H&E stained section at the bottom shows markers. Other antibodies also expected to be positive are anti-CK7.
- as usual. Normal corneal endothelial cells do not express epithelial markers. Other antibodies also expected to be positive are anti-CK7.
- marker by the arrow. The arrow points to the abnormal endothelium. Corneal stroma is negative.
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ANSWERS