

Posterior Crocodile Shagreen

Shinji Makino MD, PhD^{1*}

¹Inoda Eye Clinic, Nasushiobara, Tochigi 329-3156, Japan

DOI: [10.36347/sjmcr.2024.v12i06.027](https://doi.org/10.36347/sjmcr.2024.v12i06.027)

| Received: 04.05.2024 | Accepted: 09.06.2024 | Published: 11.06.2024

*Corresponding author: Shinji Makino

Inoda Eye Clinic, Nasushiobara, Tochigi 329-3156, Japan

Abstract

Case Report

A 62-year-old Japanese man presented for refractive evaluation. His best corrected visual acuity was 20/20 in both eyes. Slit-lamp evaluation of the cornea revealed bilateral deep stromal haze centrally in a mosaic pattern with intervening cracklike clear zones. The patient was diagnosed with posterior crocodile shagreen. This case highlights the importance for clinicians to be aware of posterior crocodile shagreen as central posterior corneal cloudiness.

Keywords: Corneal opacity, corneal dystrophy.

Copyright © 2024 The Author(s): This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY-NC 4.0) which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

INTRODUCTION

In the revised third edition of the IC3D classification of corneal dystrophies, corneal stromal dystrophies include macular corneal dystrophy, Schnyder corneal dystrophy, congenital stromal corneal dystrophy, fleck corneal dystrophy, posterior amorphous corneal dystrophy, central cloudy dystrophy of François, pre-Descemet corneal dystrophies, punctiform and polychromatic pre-Descemet corneal dystrophy.

Posterior crocodile shagreen is characterized by a mosaic pattern of corneal clouding affecting the deep

corneal stroma but sparing the corneal epithelium and endothelium.

Herein, we report a case of posterior crocodile shagreen.

CASE REPORT

A 62-year-old Japanese man presented for refractive evaluation. His best corrected visual acuity was 20/20 in both eyes. Slit-lamp evaluation of the cornea revealed bilateral deep stromal haze centrally in a mosaic pattern with intervening cracklike clear zones (Figure 1). From his clinical appearance, the patient was diagnosed with posterior crocodile shagreen.

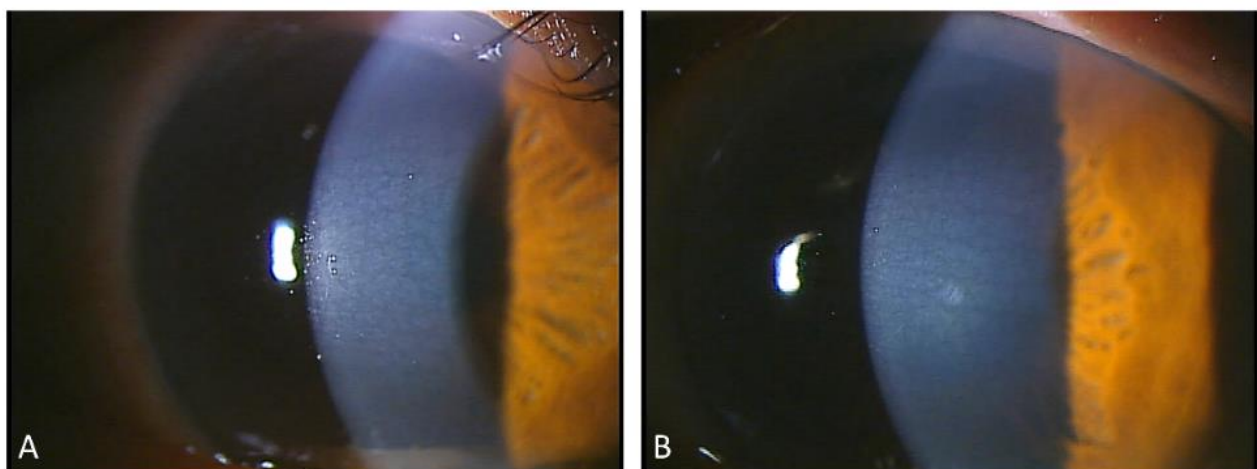


Figure 1. Slit-lamp photographs of the right (A) and left (B) eyes

Note polygonal gray–white stromal opacities separated by linear areas of clear cornea typical of posterior crocodile shagreen.

DISCUSSION

Posterior crocodile shagreen is characterized by a mosaic pattern of corneal clouding affecting the deep corneal stroma but sparing the corneal epithelium and endothelium [1]. The name posterior crocodile shagreen reflects the location of the opacities and the resemblance of their pattern to crocodile skin. The feature in this patient is indistinguishable from central cloudy dystrophy of François with an autosomal dominant inheritance [1, 2]. Both posterior crocodile shagreen and central cloudy dystrophy of François are asymptomatic, often found incidentally, and are nonprogressive diseases [1, 2]. Neither condition is believed to cause significant visual loss.

There are few reports describing the histopathological findings in central cloudy dystrophy of François or posterior crocodile shagreen [1]. On transmission electron microscopy in posterior crocodile shagreen, extracellular vacuoles, some containing fibrillogranular material, electron-dense deposits and a sawtooth lamellar pattern have been reported [1, 2]. On confocal biomicroscopy in posterior crocodile shagreen, small highly refractile granules and deposits in the anterior stroma were reported [1, 3]. The pathogenesis of posterior crocodile shagreen is thought to be involved a

primary degenerative process which leads to reproducible changes in the posterior stroma, possibly related to local changes in the arrangement of the collagenous lamellae [1]. In familial cases (central cloudy dystrophy of François), this process may be accelerated leading to the clinical manifestations at an earlier age.

CONCLUSION

This case highlights the importance for clinicians to be aware of posterior crocodile shagreen as central posterior corneal cloudiness.

Disclosure: The author declares no conflict of interest.

REFERENCES

1. Weiss, J. S., Rapuano, C. J., Seitz, B., Busin, M., Kivelä, T. T., Bouheraoua, N., ... & Lisch, W. (2022). IC3D Classification of Corneal Dystrophies—Edition 3. *Cornea*, 10-1097.
2. Belliveau, M. J., Brownstein, S., Agapitos, P., & Font, R. L. (2009). Ultrastructural features of posterior crocodile shagreen of the cornea. *Survey of ophthalmology*, 54(5), 569-575.
3. Woodward, M., Randleman, J. B., & Larson, P. M. (2007). In vivo confocal microscopy of polymorphic amyloid degeneration and posterior crocodile shagreen. *Cornea*, 26(1), 98-101.