CFEH facebook Cases

CFEH Facebook Case #17

A 63 year old Caucasian female presented at CFEH. She was a low myope and generally asymptomatic although did mention she sometimes bumps into things. BCVA 6/6 in each eye. She has previously undergone a tracheotomy and radiotherapy for a vascular malformation near her larynx and currently takes Crestor, Atacand and Nexium. The Optomap image, fundus autofluorescence and OCT line scans are included for the right eye only, although both eyes had a similar appearance.

What is your diagnosis and management?

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Optometry
NEW SOUTH WALES
AUSTRALIAN CAPITAL TERRITORY

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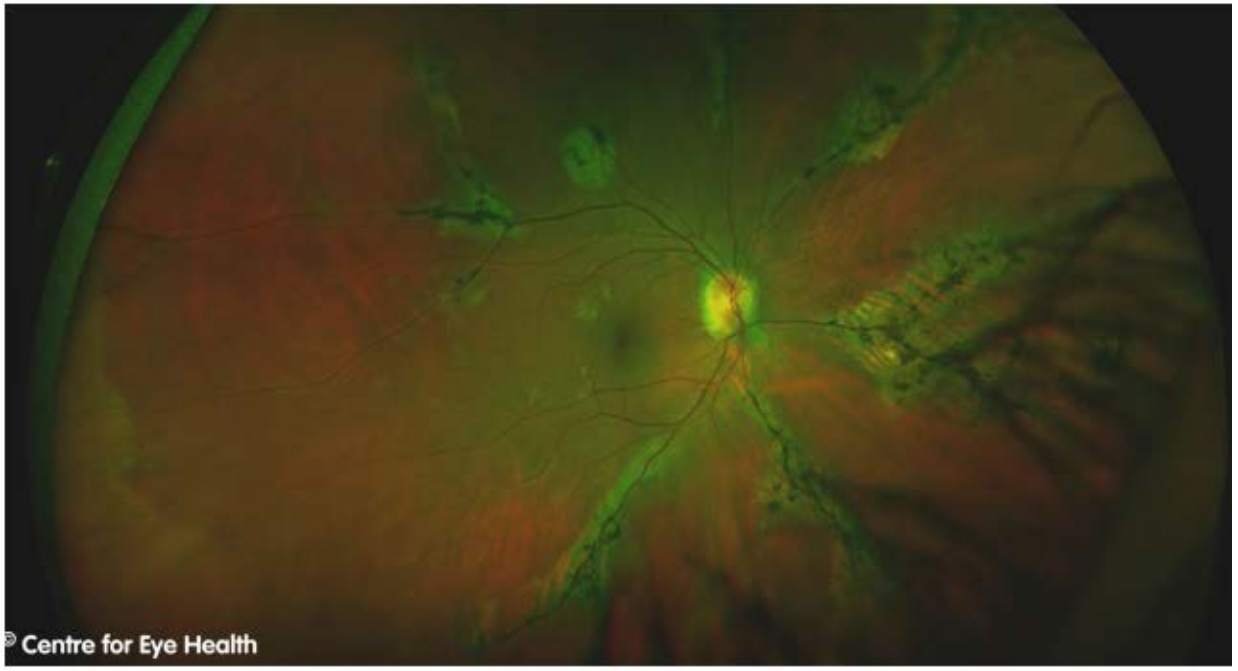
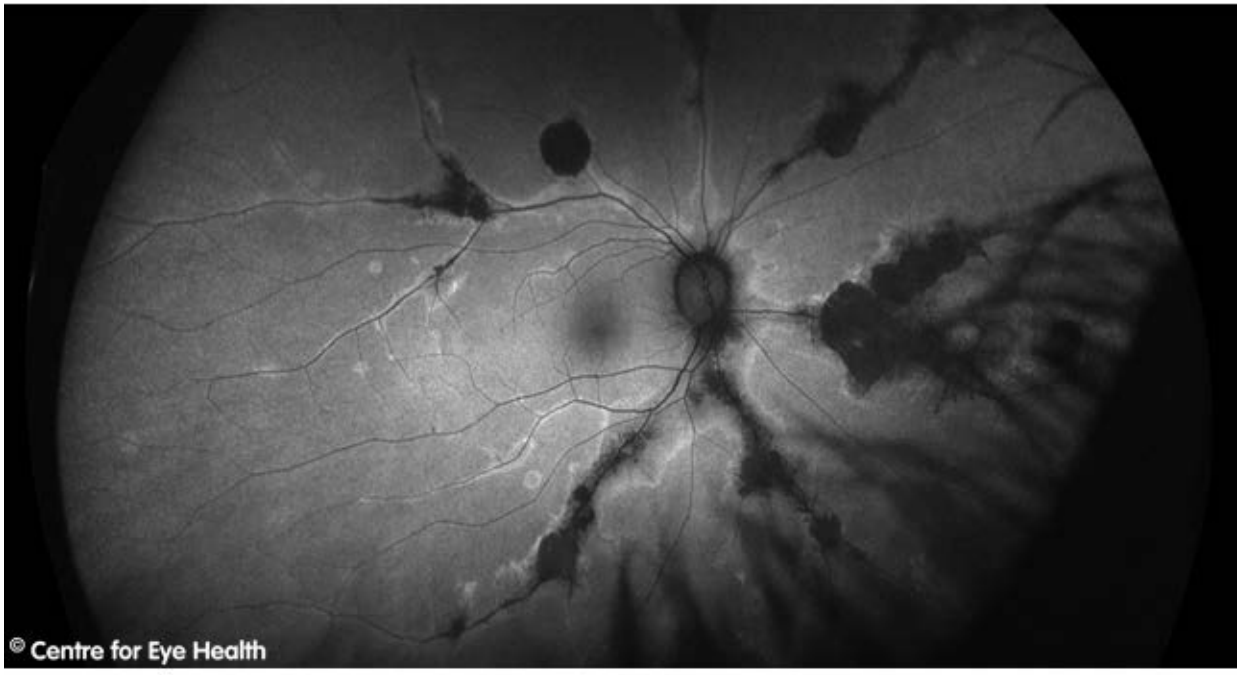




Central 30-2 Threshold Test

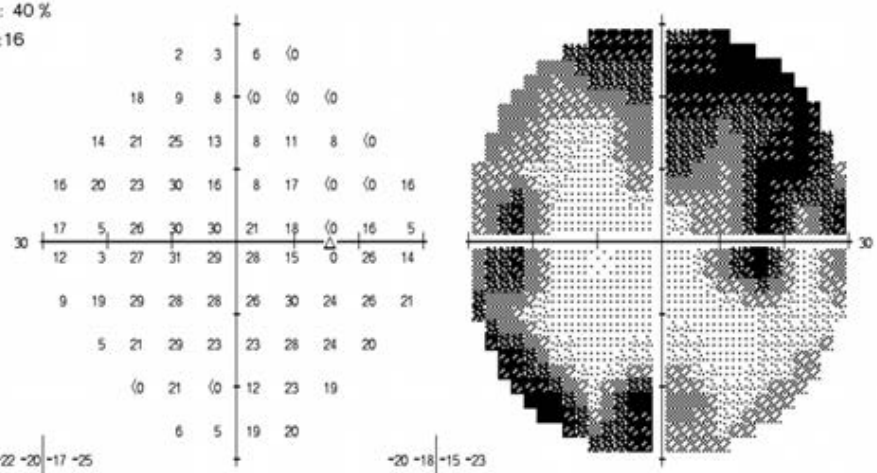
Fixation Monitor: Gaze/Blind Spot
Fixation Target: Central
Fixation Losses: 0/22
False POS Errors: 1 %
False NEG Errors: 40 %
Test Duration: 10:16

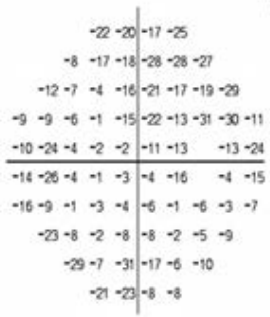
Fovea: OFF

Stimulus: III, White
Background: 31.5 ASB
Strategy: SITA-Standard

Pupil Diameter: 6.1 mm
Visual Acuity:
RX: +8.75 DS DC X

Date: 23-05-2013
Time: 11:03 AM
Age: 64





Total Deviation

Pattern Deviation

GHT
Outside Normal Limits

VFI 72%

MD -10.66 dB P < 0.5%
PSD 10.09 dB P < 0.5%

:: < 5%
∴ < 2%
▒ < 1%
■ < 0.5%

Guide Dogs - University Of NSW
Centre for Eye Health
Gate 14, Barker Street
Kensington NSW 2033

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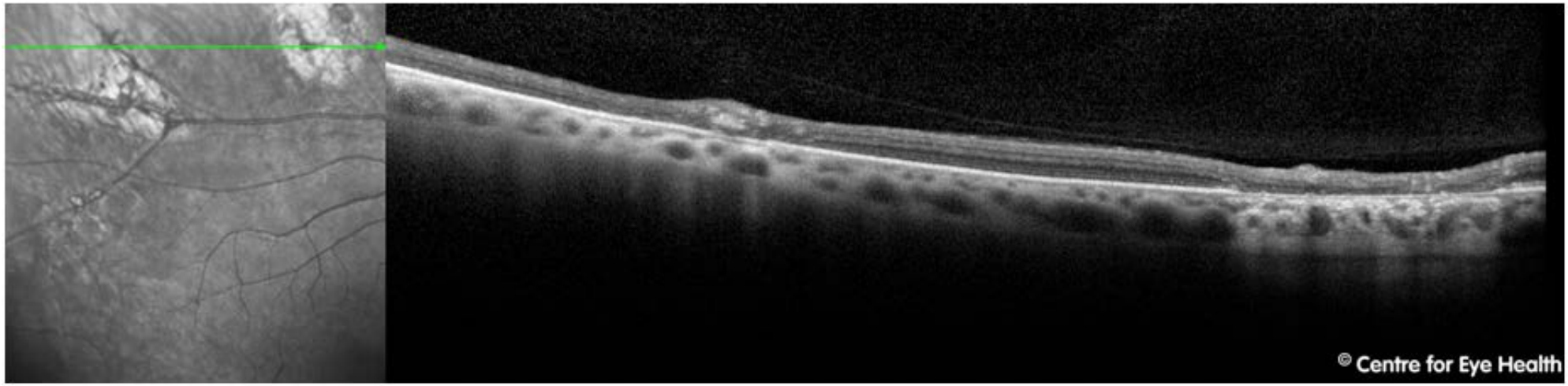

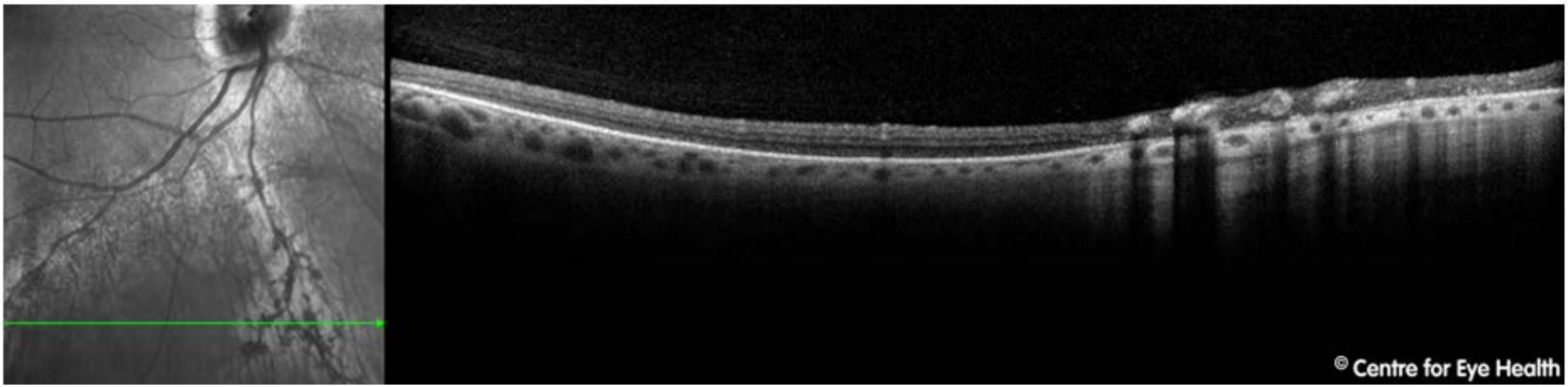

ANSWER

Pigmented Paravenous Chorioretinal Atrophy (PPCRA)

This is a bilateral, progressive condition that is characterised by pigmentary changes including RPE atrophy and pigment clumping around the blood vessels. The OCT images show a reduction in the outer nuclear layer thickness, as well as disappearance of the Ise and interdigitation zone. The hyper-reflective areas seen in the OCT images within the retina are due to RPE migration and increased reflectance seen in areas of the choroid is due to the drop out of RPE cells.

The exact aetiology of this condition is unknown, and speculation exists that it may be either genetic in nature, developmental or an immune response due to associated inflammatory conditions.

Sufferers often remain asymptomatic and the threat to vision is not high as the condition is usually stable. Recommended management is to review the fundus appearance and visual fields annually to document progression. An ERG can help with diagnosis and monitoring of this condition. In this case, both an ERG and EOG were performed at CFEH with the ERG showing a normal scotopic and reduced photopic response. The EOG showed an abnormal Arden ratio. Given the presence of significant field defect, Estermann field assessment may be required to assess suitability to drive.

This case and a series of other unusual eye conditions are presented in a video lecture "Case Studies on Unusual Eye Conditions", available for viewing through Learning for Vision and approved for 2 therapeutic CPD points. Go to www.learningforvision.com.au to purchase this and other lectures from our catalogue.