Proposing a Neurotropic Aetiology for Acute Posterior Multifocal Placoid Pigment Epitheliopathy and Relentless Placoid Chorioretinitis

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Background

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) was first described by Gass in 1968.[1] In 2000, Jones *et al* described a series of cases that resembled APMPPE but with an atypical prolonged clinical course with lesions beyond the posterior pole and coined the term *relentless placoid chorioretinitis* (RPC).[2]

While controversy continues regarding the aetiology of these conditions, the prevailing hypothesis supports a choriocapillaris ischaemia as the primary pathology.

We re-evaluate the multimodal imaging findings to explore how a primary neuronal aetiology may account for the signs and sequence of pathology observed.

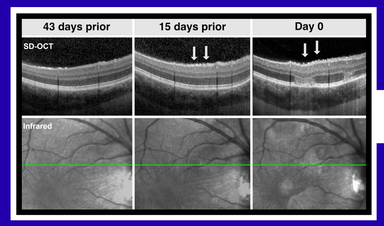
Methods

Patients with imaging in keeping with a diagnosis of APMPPE or RPC were identified from three tertiary uveitis centres in the UK between 2016 and 2020. A retrospective qualitative comparative analysis of available multimodal imaging modalities was undertaken.

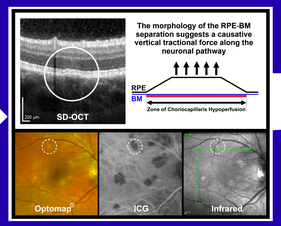
14 patients were included in our analysis. 10 patients (17 eyes) had a diagnosis consistent with APMPPE, 4 patients (6 eyes) with RPC.

Results

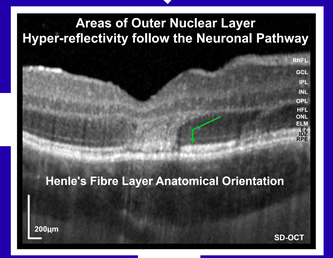
Retinal Nerve Fibre Layer Undulations Precede Outer Retinal Lesions



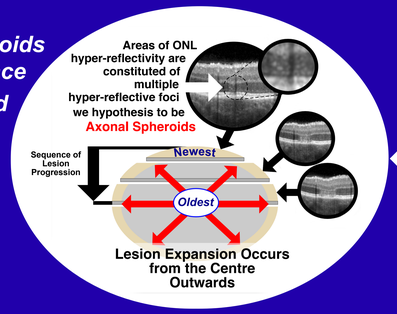
Areas of Retinal Pigment Epithelium (RPE) - Bruch's Membrane (BM) Separation Correspond to (Secondary) Areas of Choriocapillaris Hypoperfusion

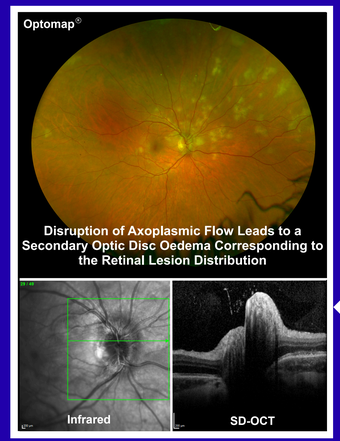


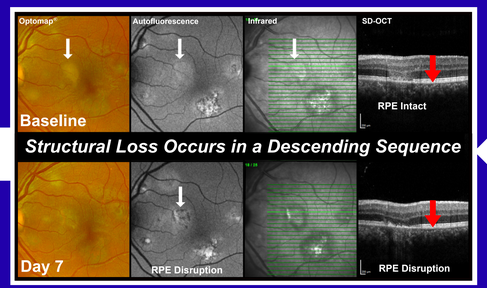
Areas of outer nuclear layer (ONL) hyper-reflectivity develop above areas of RPE-BM separation



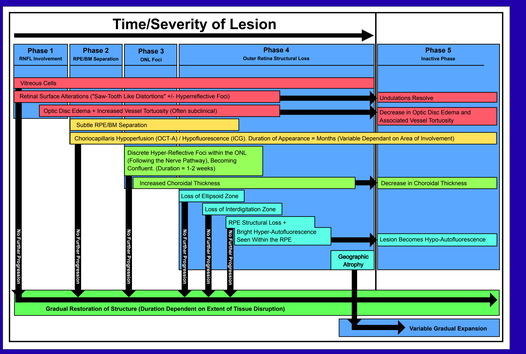
Axonal spheroids occur as a consequence of disrupted axoplasmic flow[3]







Proposed Phases of Lesion Evolution



Conclusions

Our evidence suggests a primary neuronal pathogenesis with a descending pathology through the retina. Areas of choriocapillaris hypoperfusion may occur as a secondary consequence of a physical separation between the RPE and BM induced by a vertical tractional force on the RPE along the neuronal pathway.

References

1. Gass JDM. Acute Posterior Multifocal Placoid Pigment Epitheliopathy. Arch Ophthalmol. 1968;80(2):177–185.
2. Jones BE, Jampol LM, Yannuzzi LA, et al. Relentless Placoid Chorioretinitis – A New entity or an Unusual Variant of Serpiginous Chorioretinitis? Arch Ophthalmol. 2000;118(7):931–938.
3 Beirowski B, Nógrádi A, Babetto E, Garcia-Alias G, Coleman MP. Mechanisms of Axonal Spheroid Formation in Central Nervous System Wallerian Degeneration. J Neuropathol Exp Neurol. 2010;69(5):455–472.

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