Choroidal Neovascularization Presenting in Different Stages of Best Macular Dystrophy

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Disclosures

- The authors have nothing to disclose.

Summary

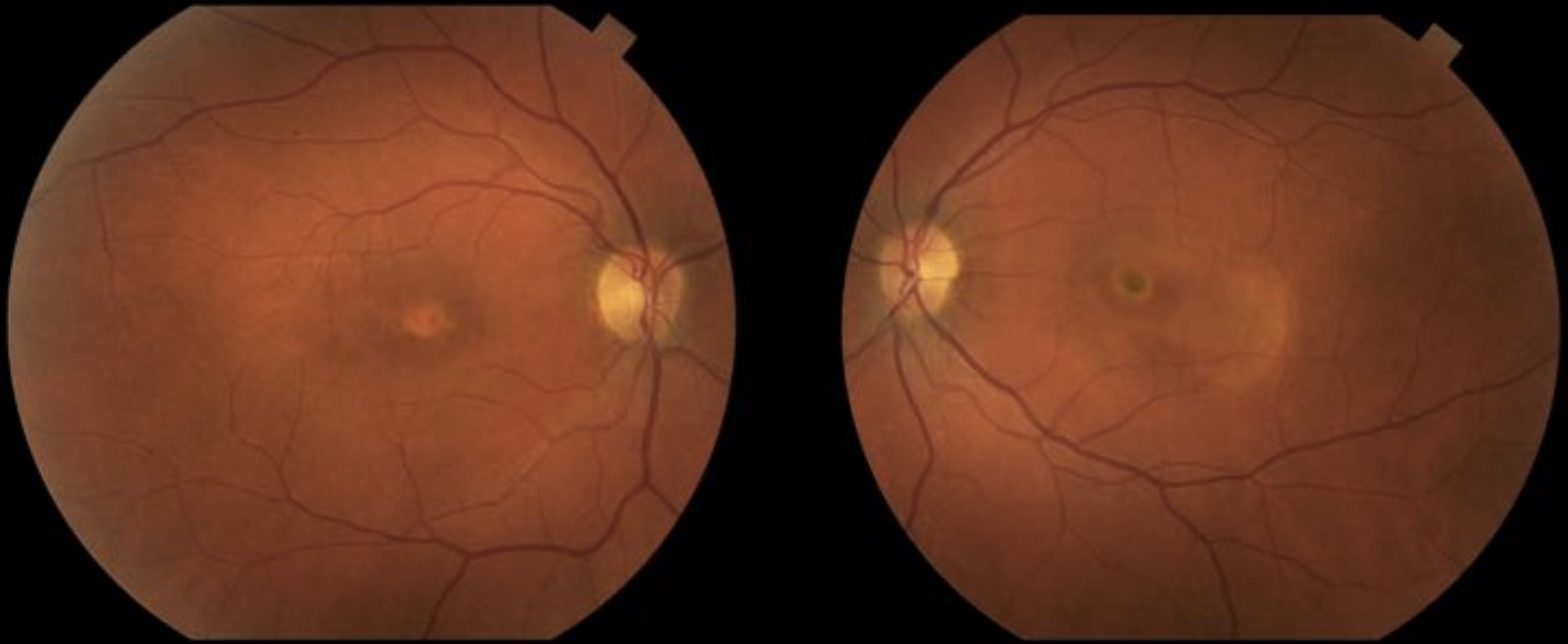
- Case series of three patients demonstrating that OCTA allows identification of CNV in more patients and in earlier stages of Best disease than previously described. This poses new clinical questions with regards to therapeutic management and follow-up.

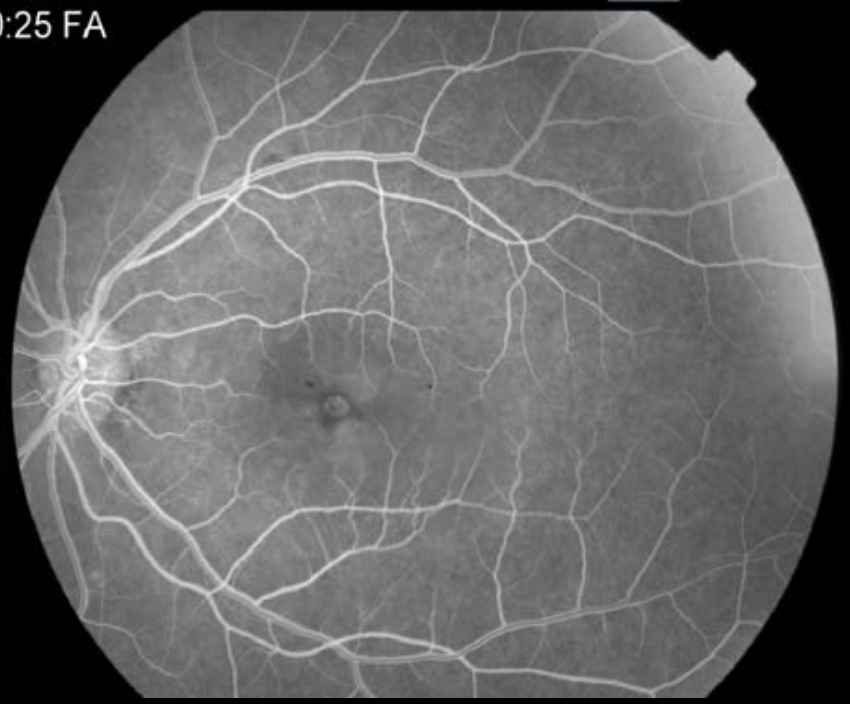
Background

- Best Macular Dystrophy (BMD): autosomal dominant condition with bilateral vitelliform lesions
- Six stages of disease: pre-vitelliform, vitelliform, pseudohypopyon, vitelliruptive, atrophic, choroidal neovascularization
- CNV: advanced disease, traditionally detected with fluorescein angiography (FA), managed with anti-VEGF agents
- OCTA has shown that CNV may occur earlier in BMD

Case 1

- 47yo F with metamorphopsia OS
- FH: two siblings with Best and other paternal family members
- BCVA 20/20 OD, 20/40 OS

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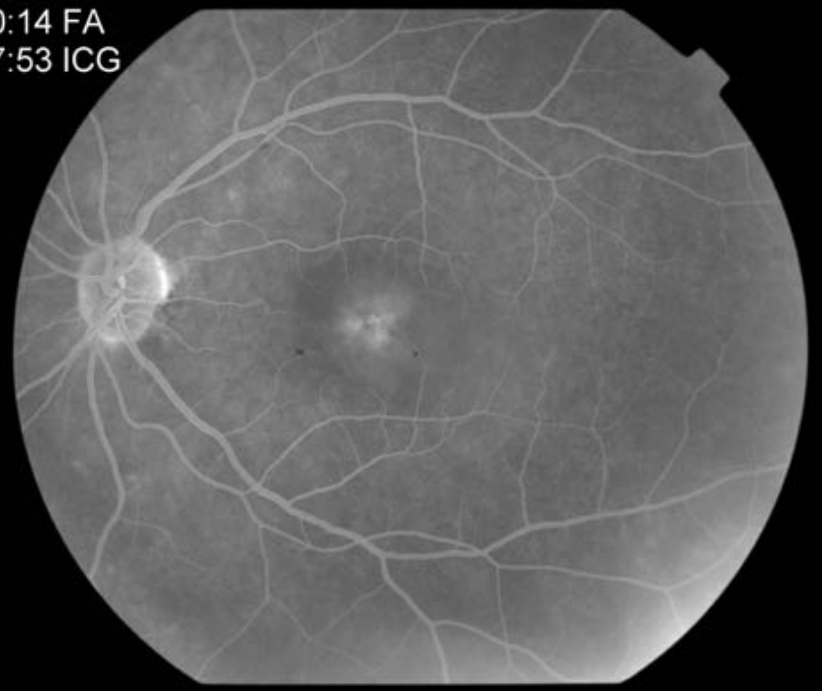
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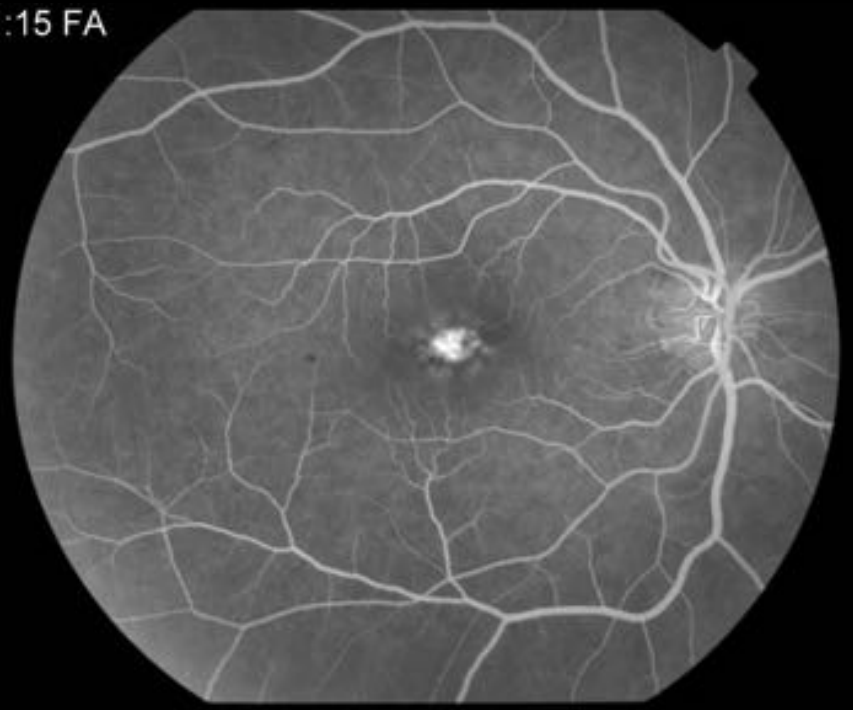
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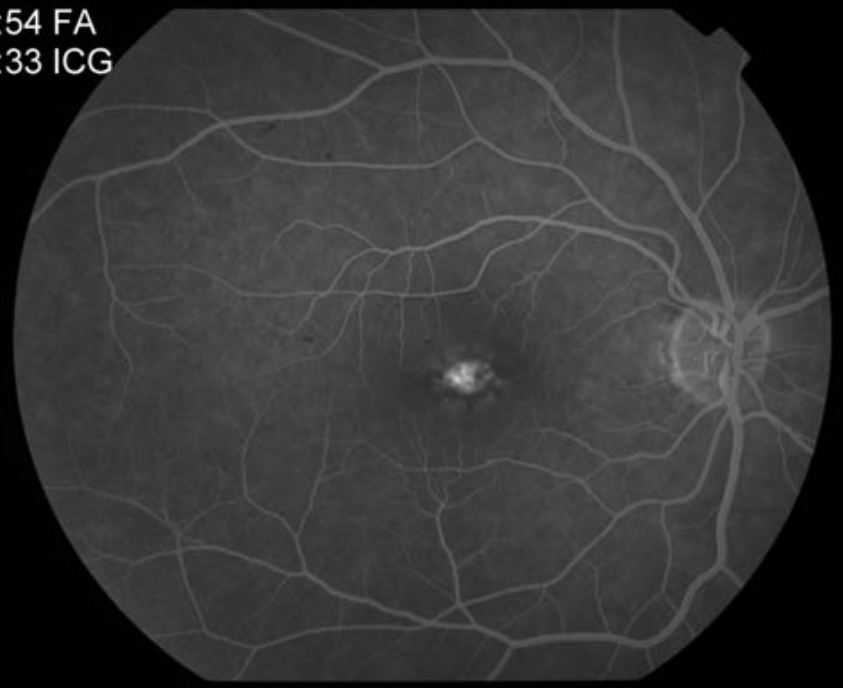
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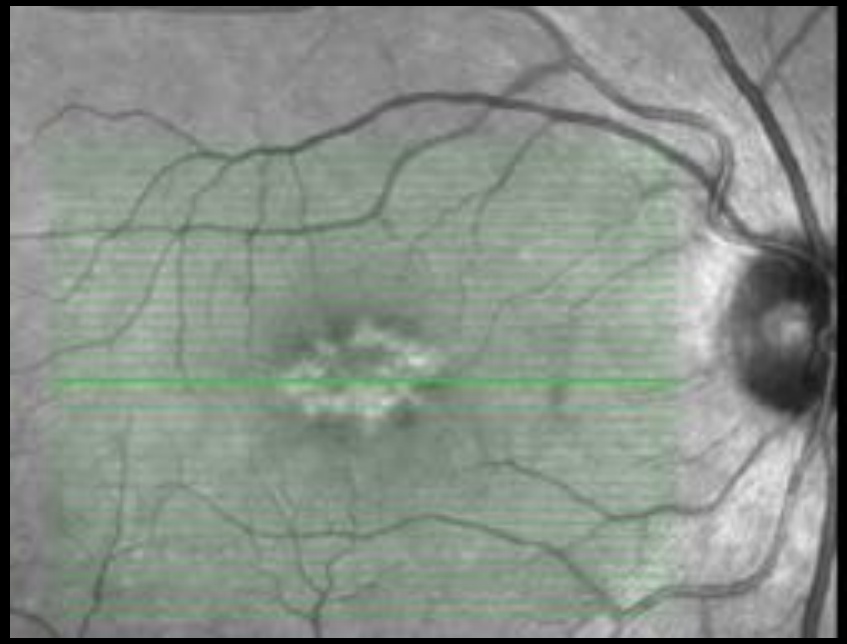

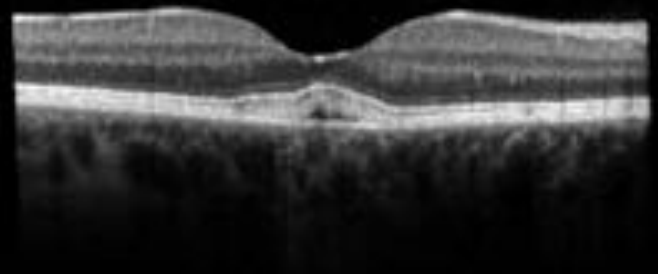

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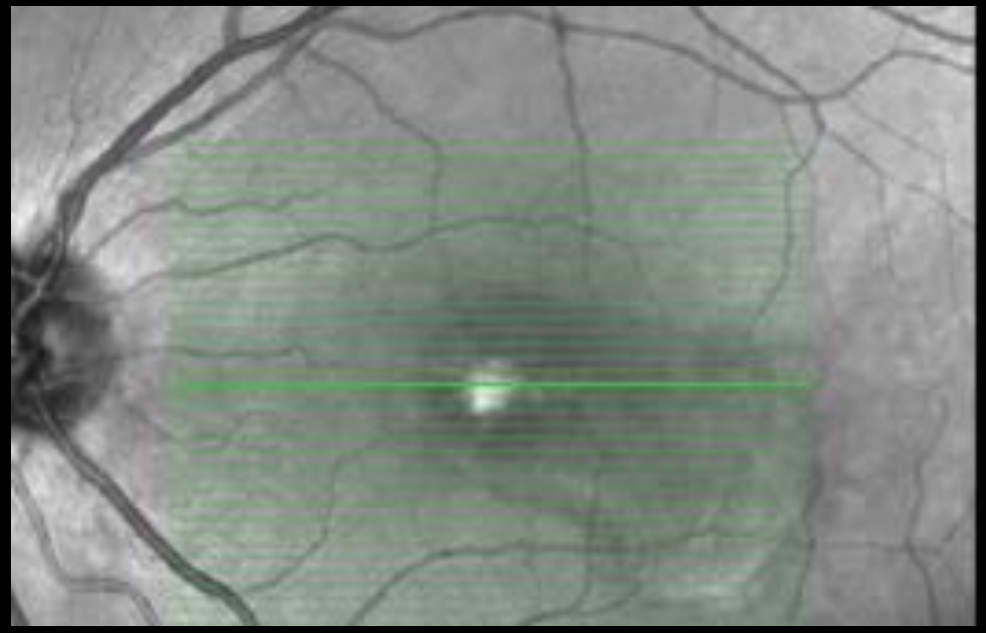

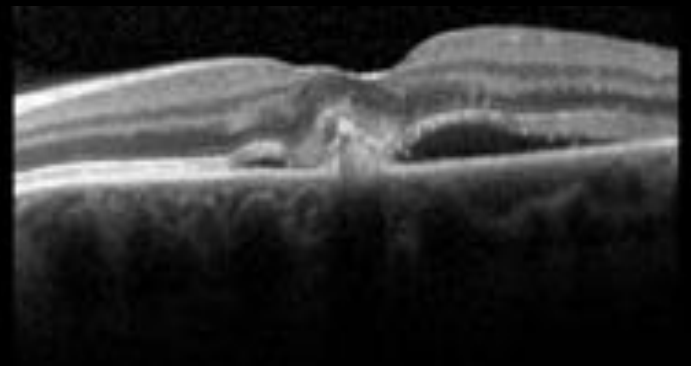

Case 1: Management

- Underwent 4 monthly bevacizumab injections for BMD complicated by CNV OS

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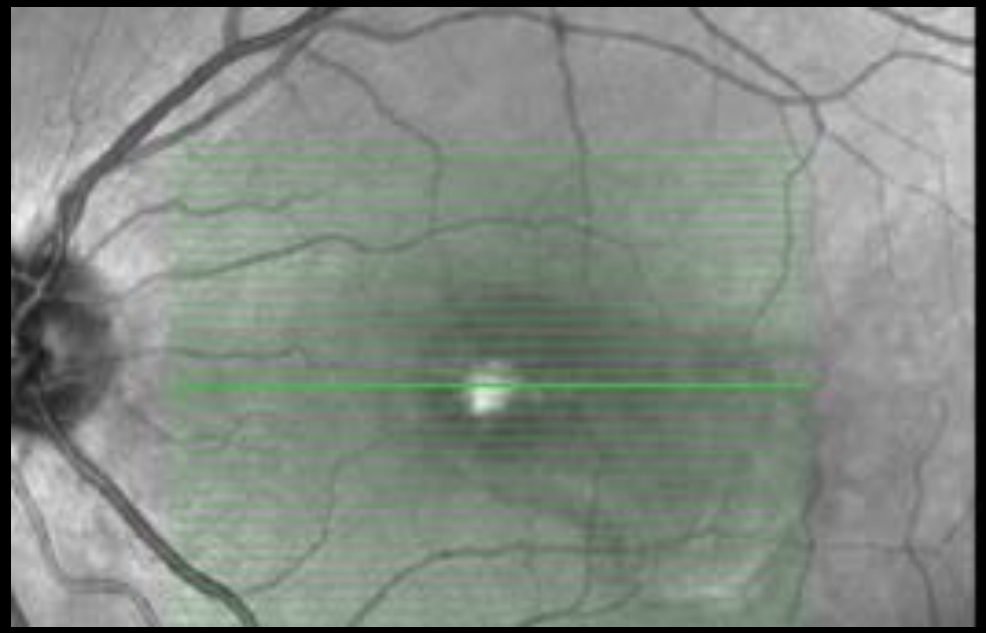

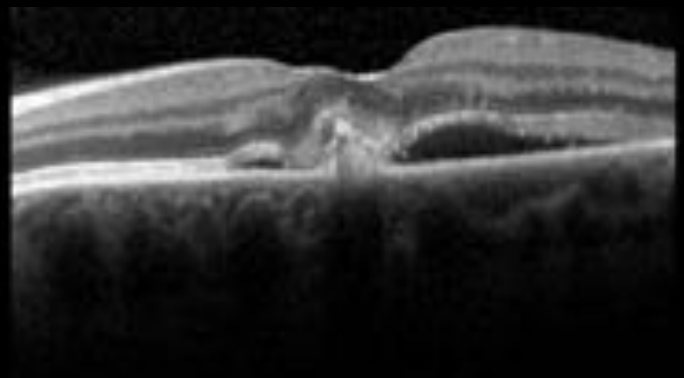

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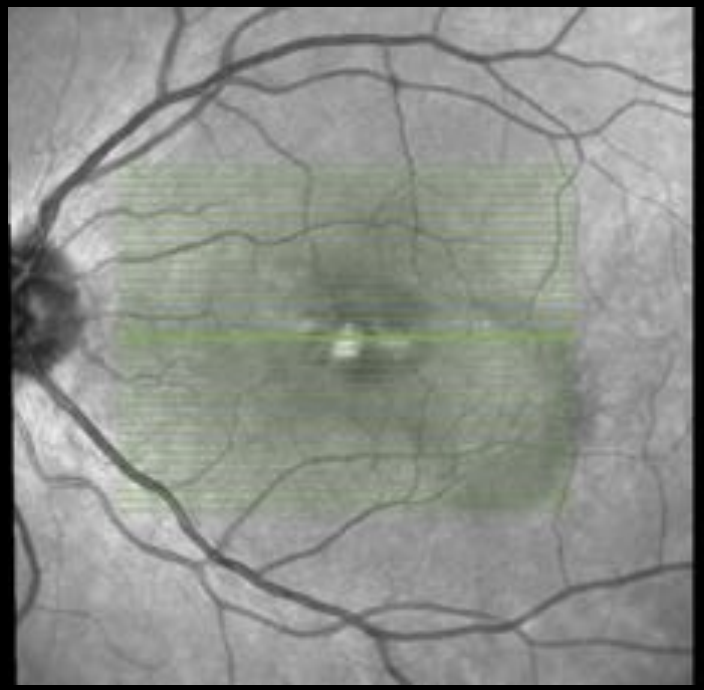

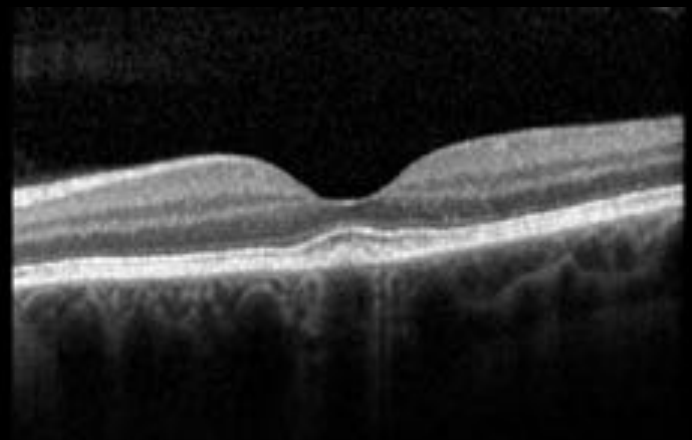

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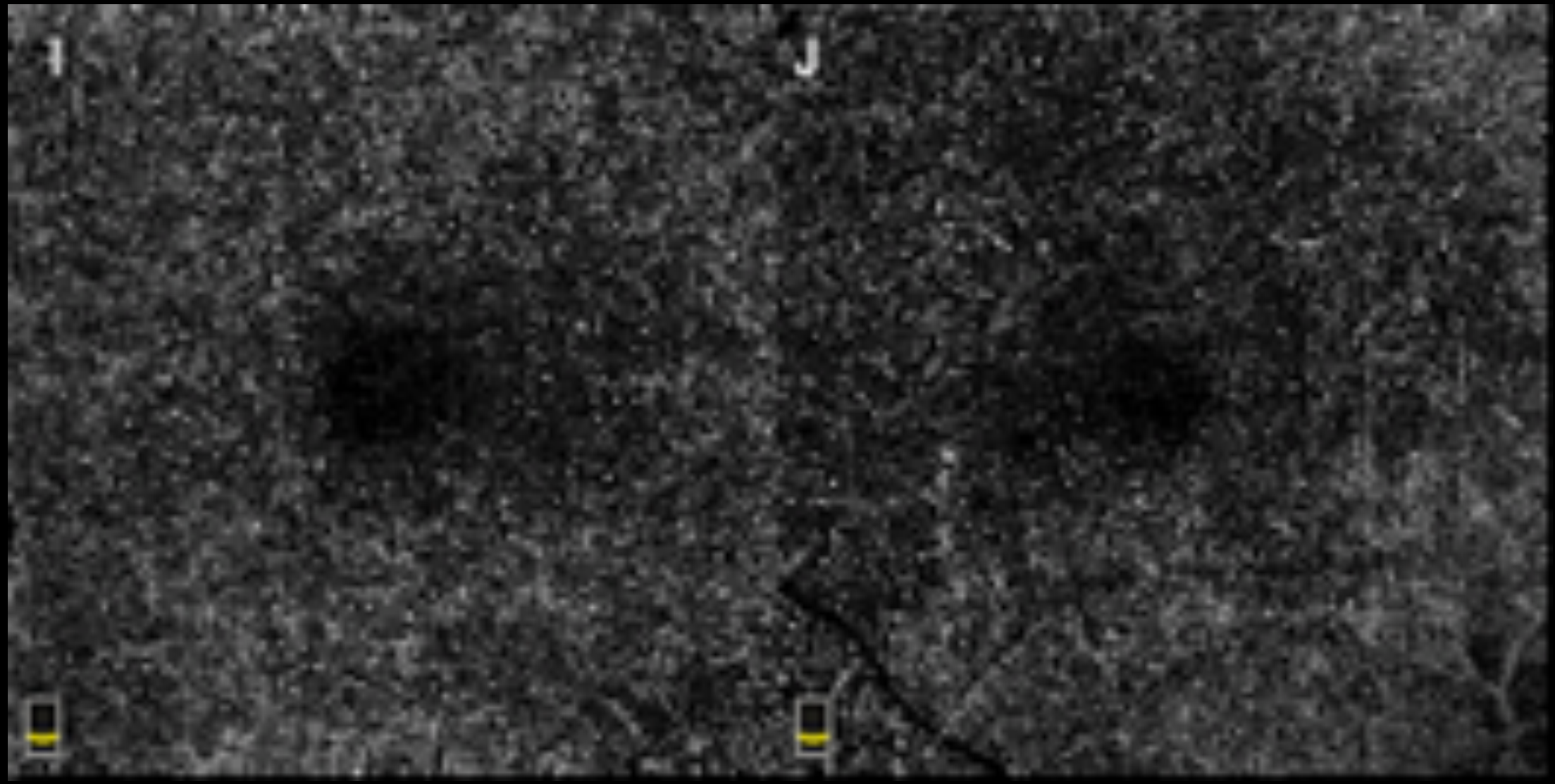
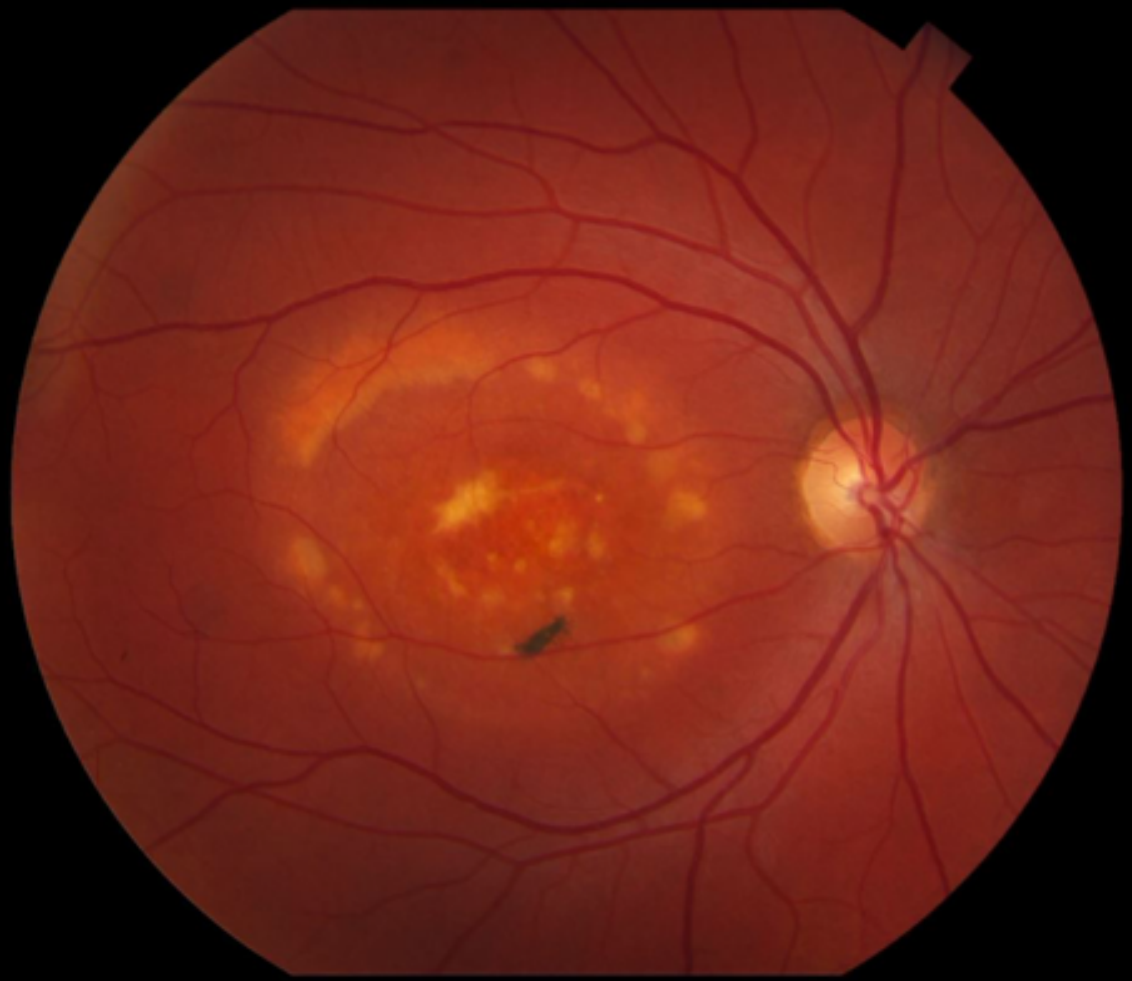
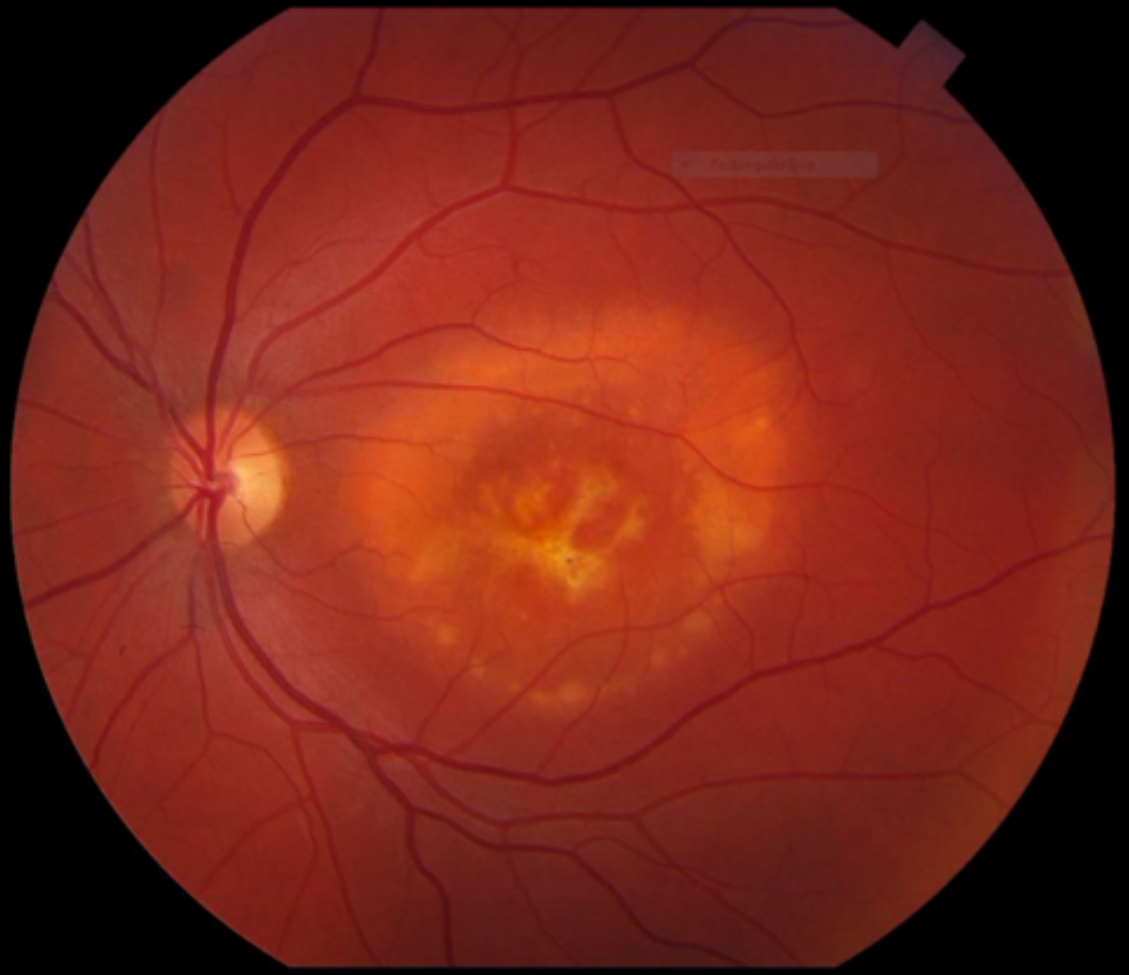


Case 2

- 34yo F with blurred vision OD > OS
- Dx with BMD since age 4
- FH: Father, paternal grandfather, great grandmother, and aunt with BMD
- BCVA 20/50 OD, 20/30 OS





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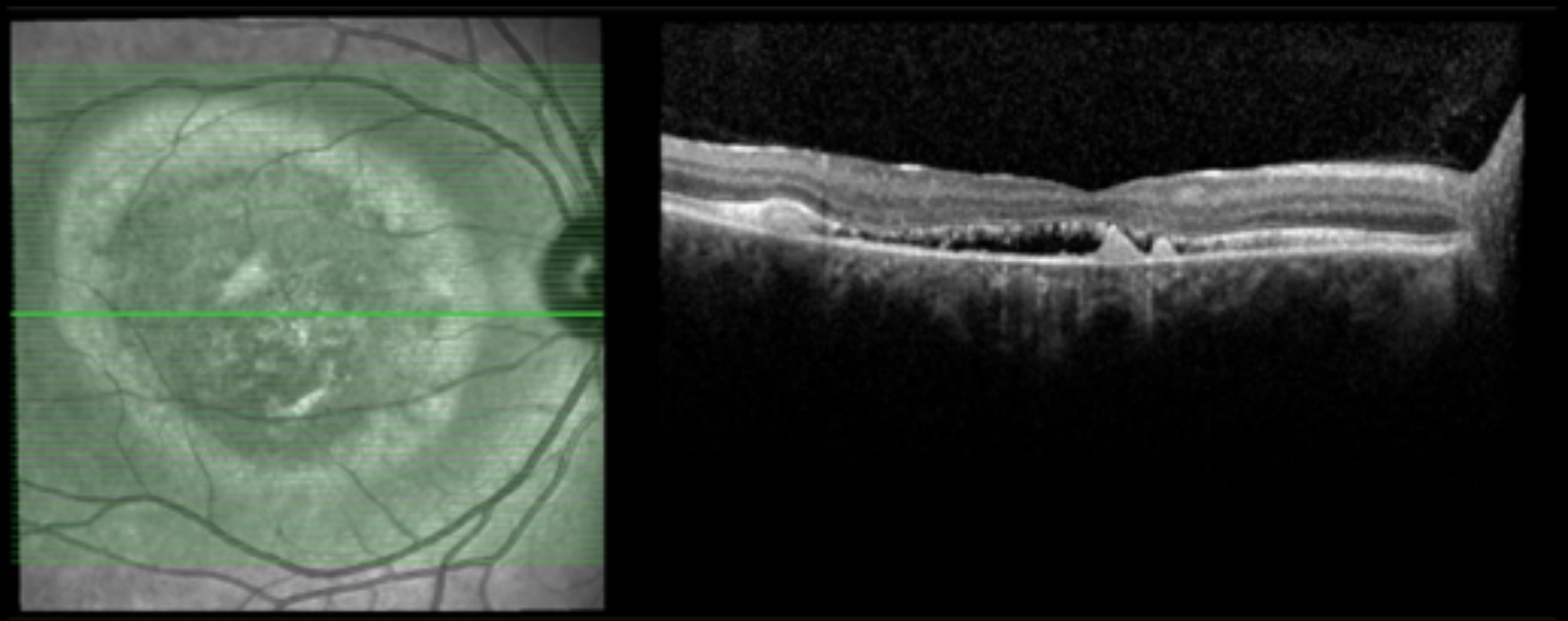

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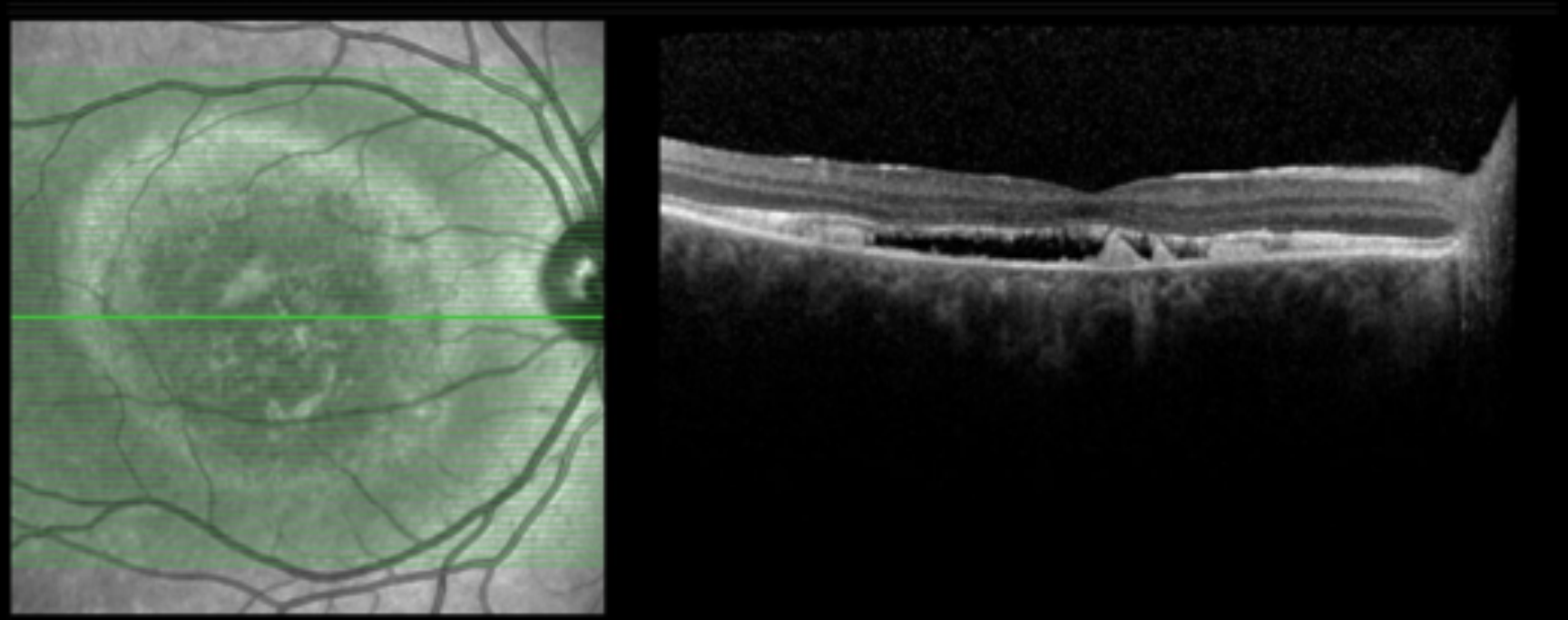

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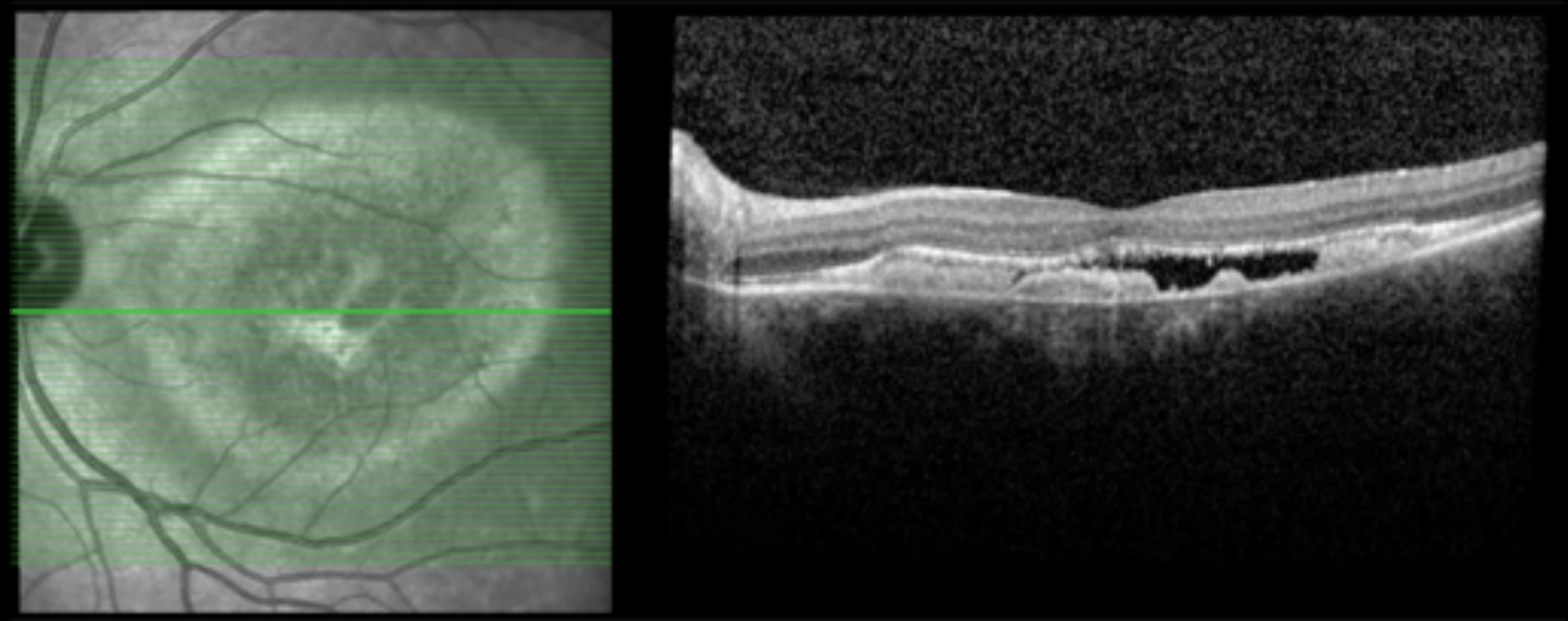

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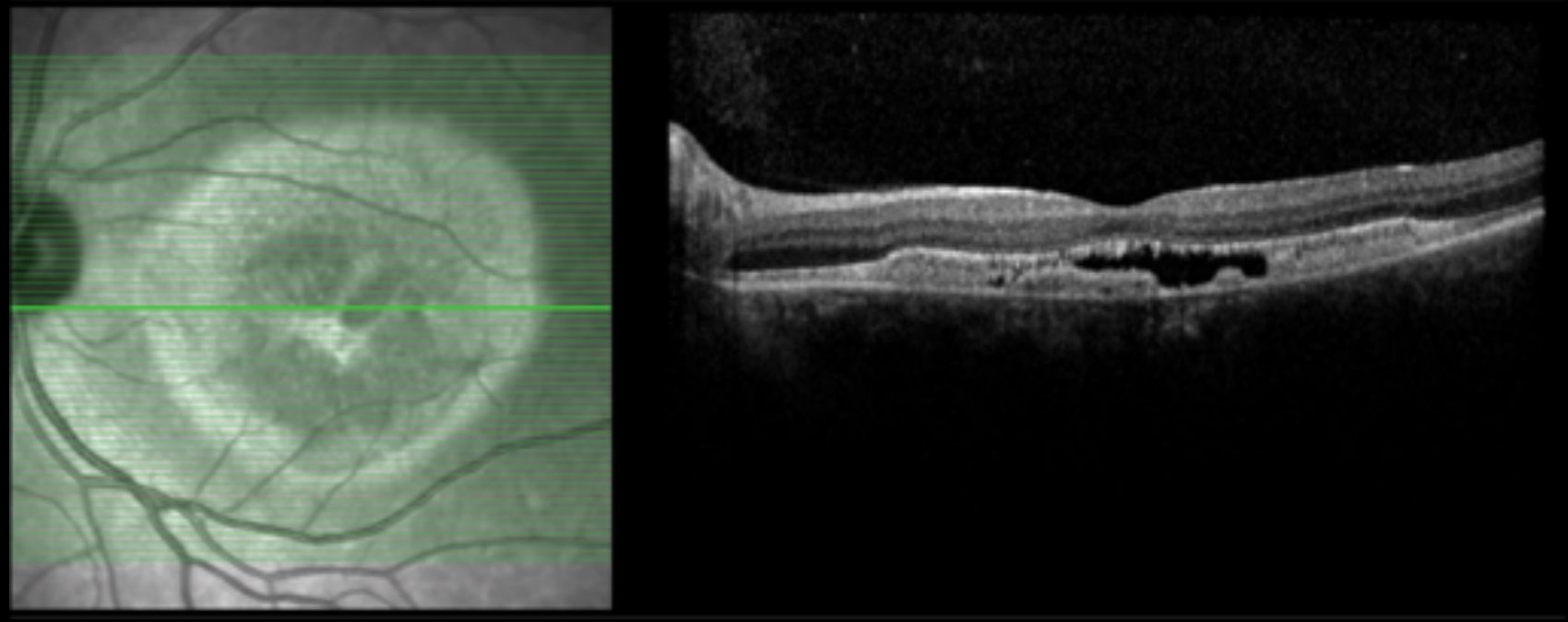

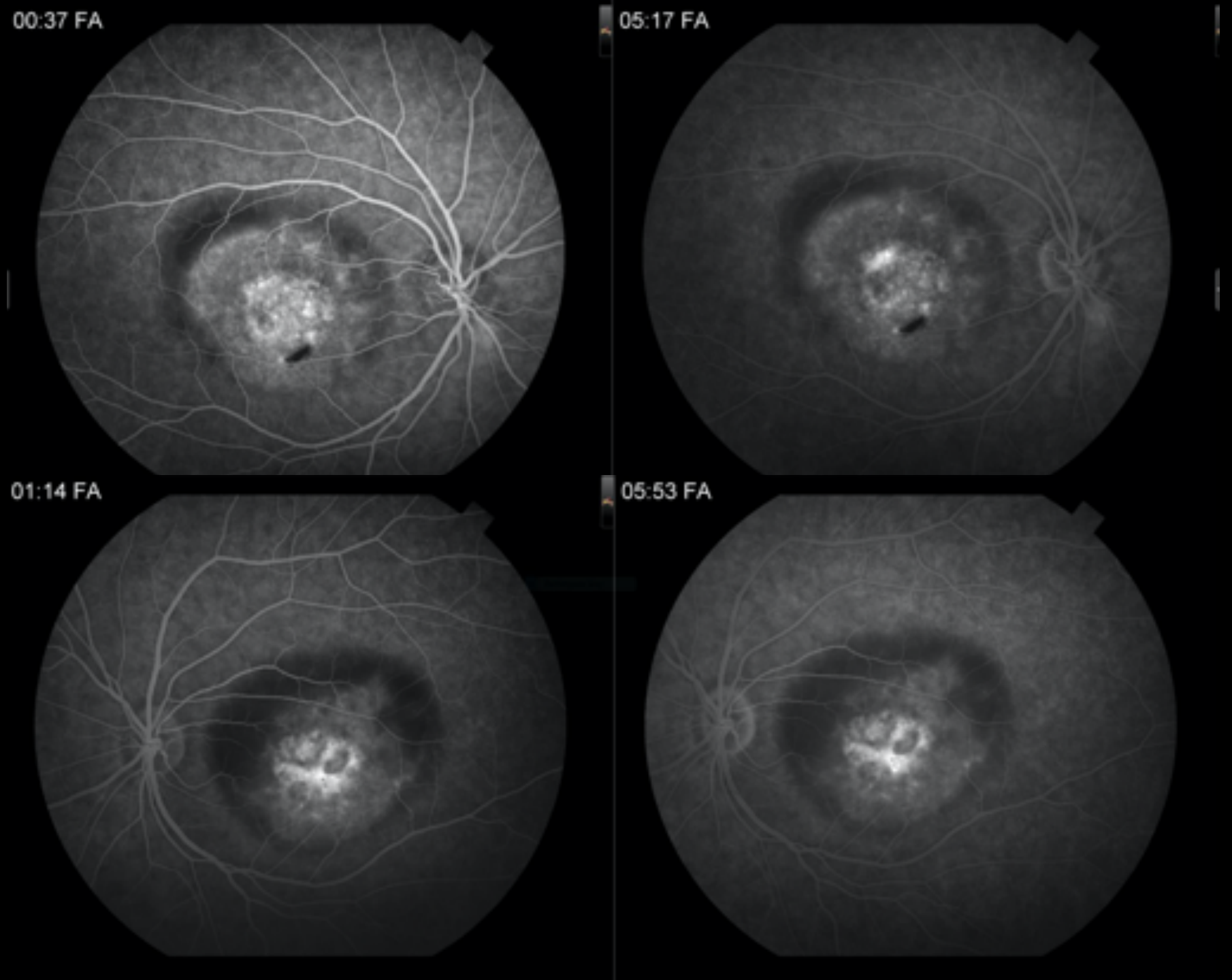
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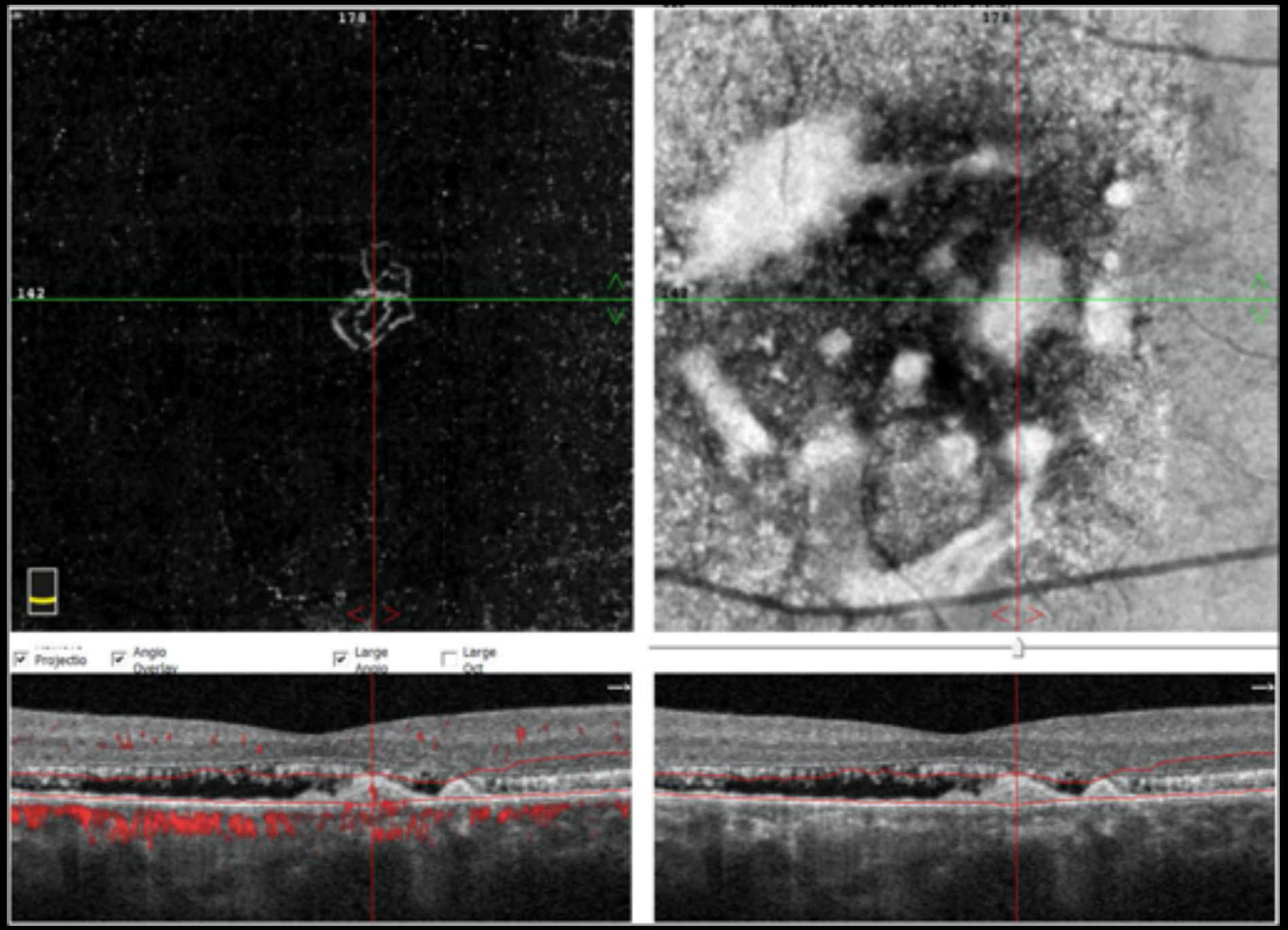
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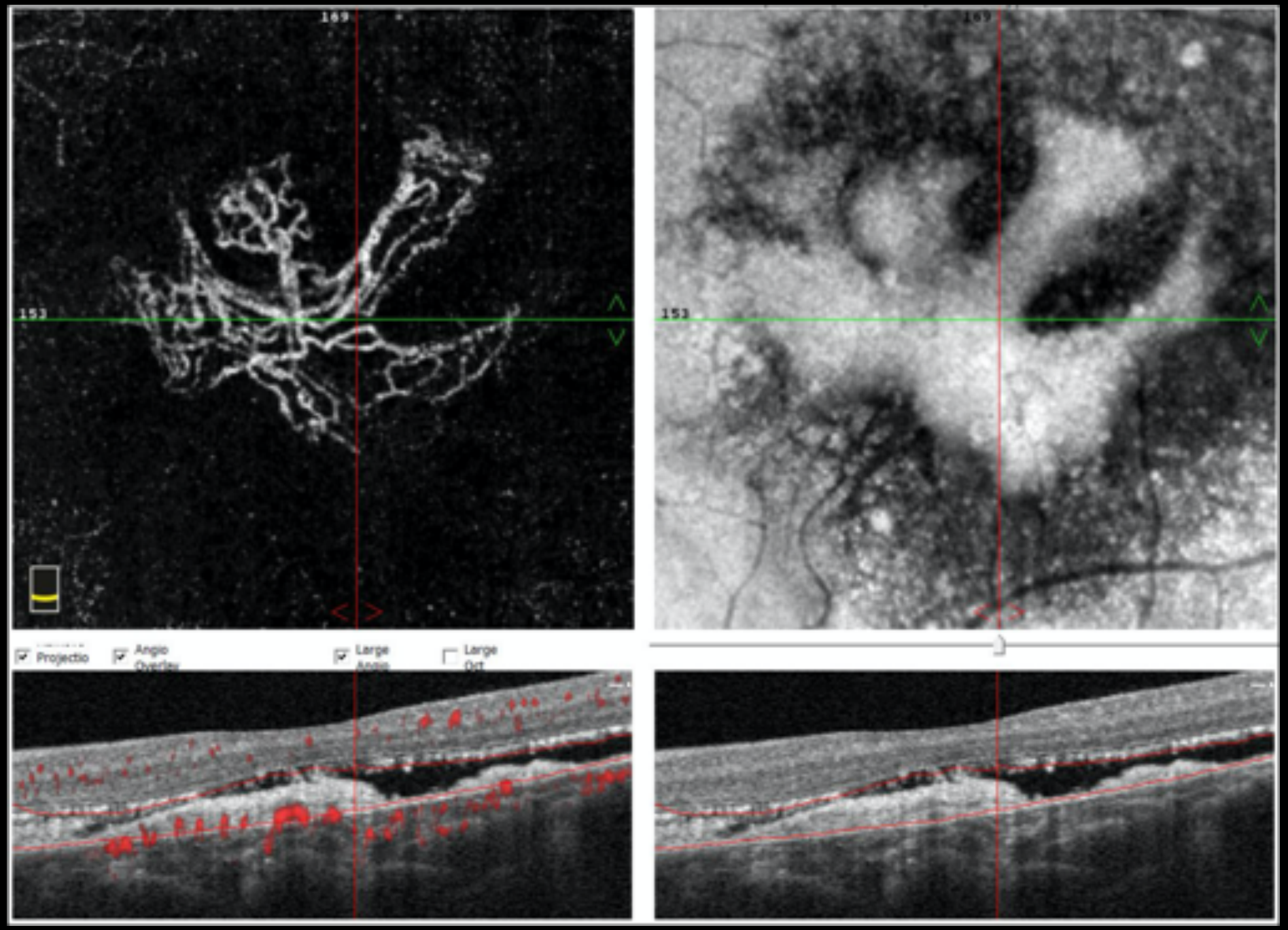
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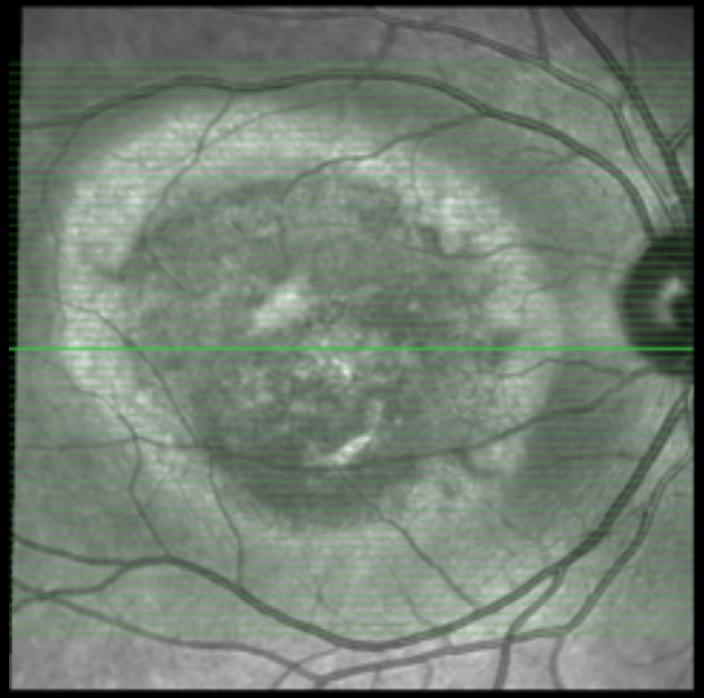
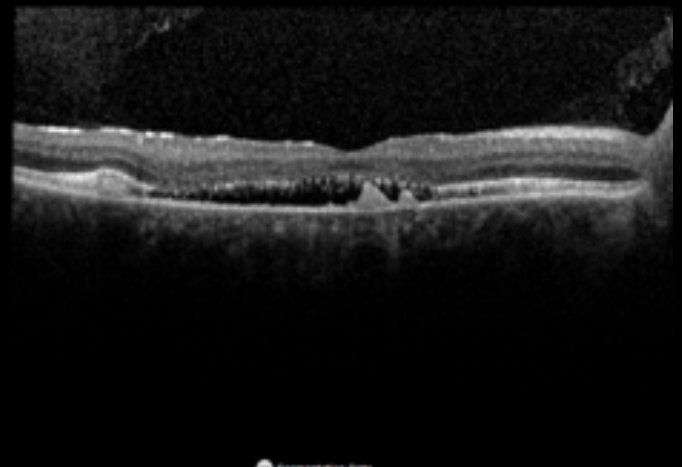
Case 2: Management

- Two bevacizumab injections OD (1/29/19 and 2/26/19) with no improvement clinically or on imaging

1/29/19





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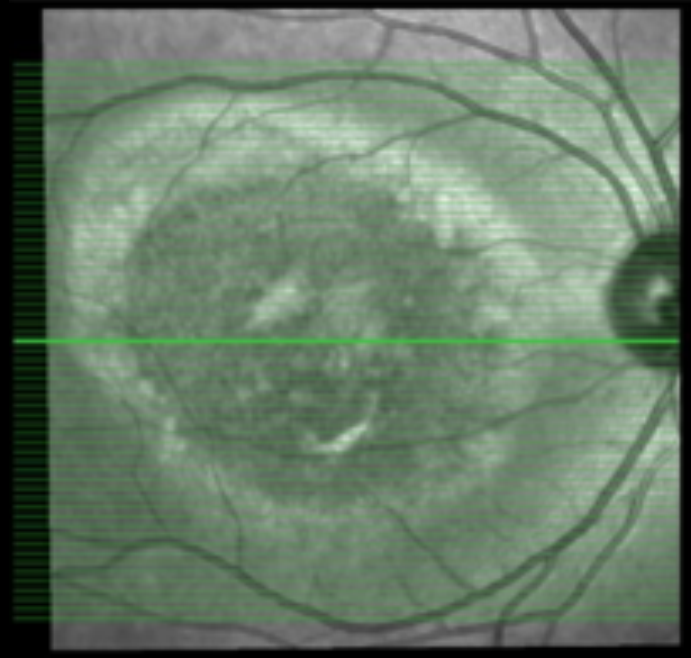

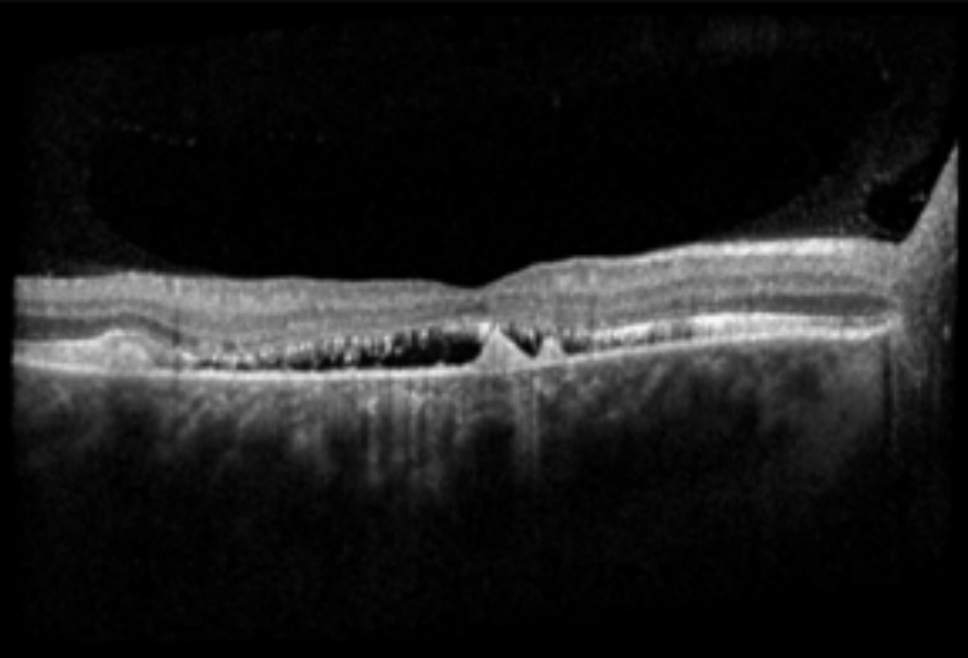

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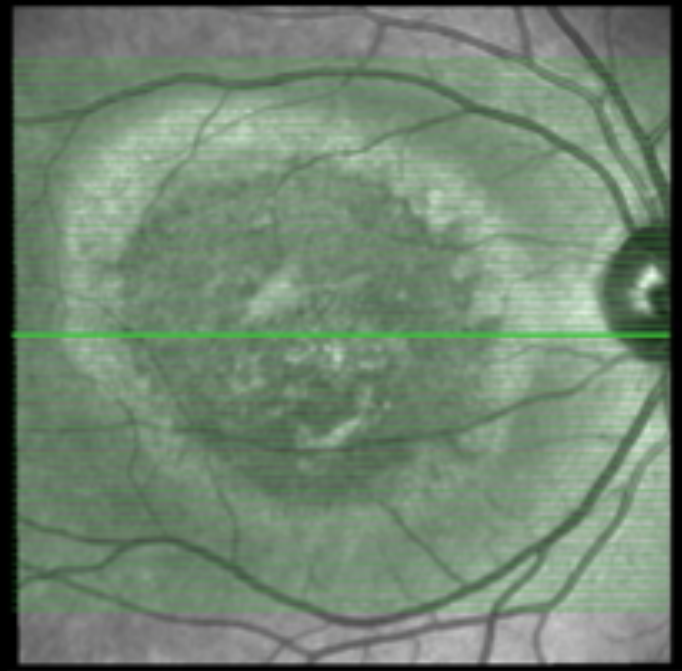
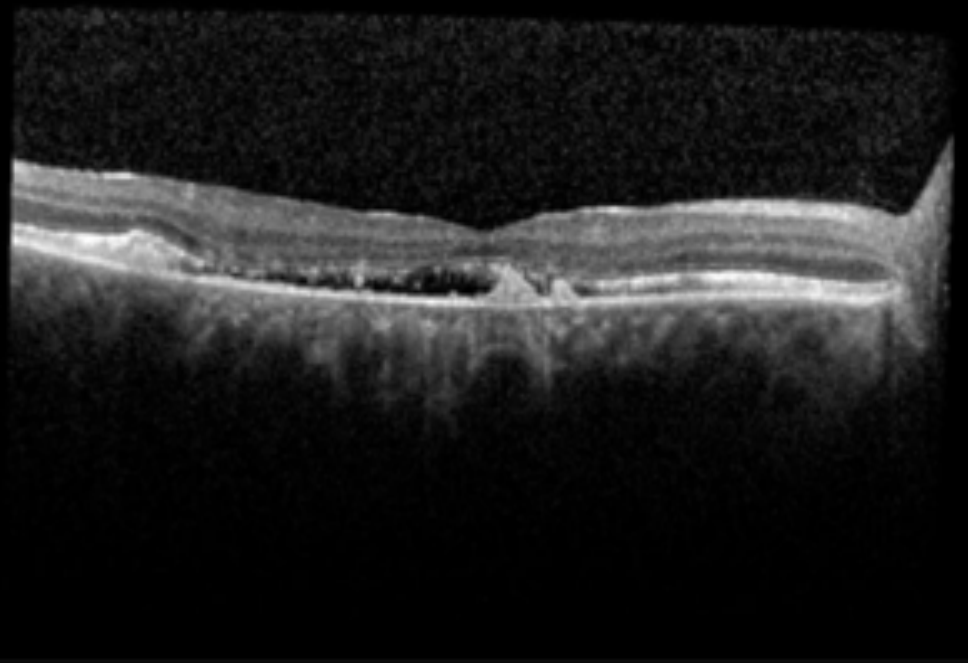
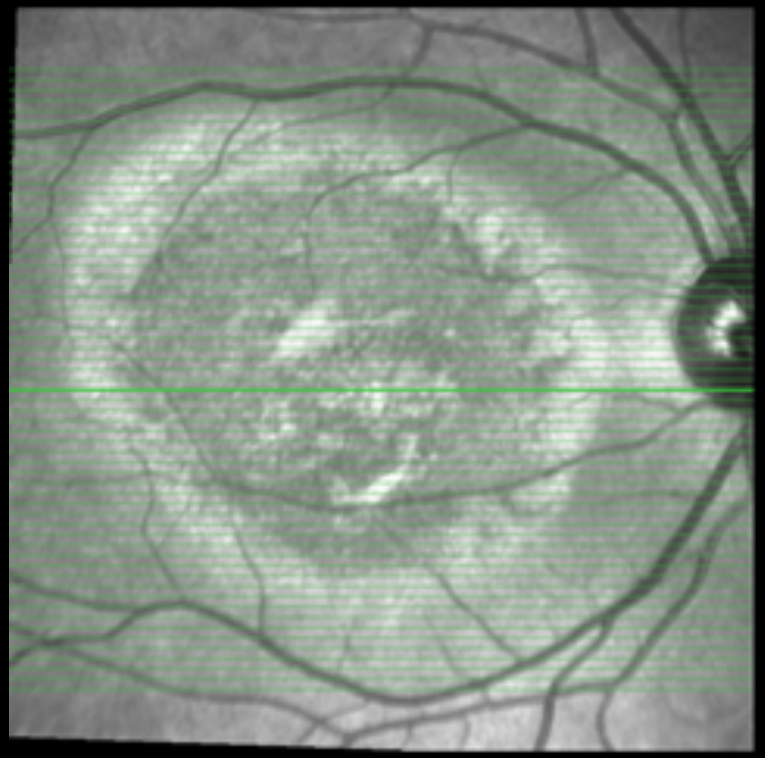
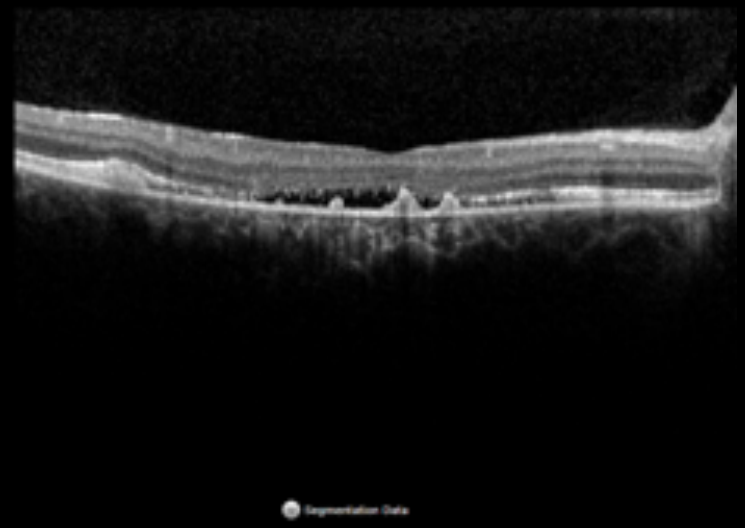


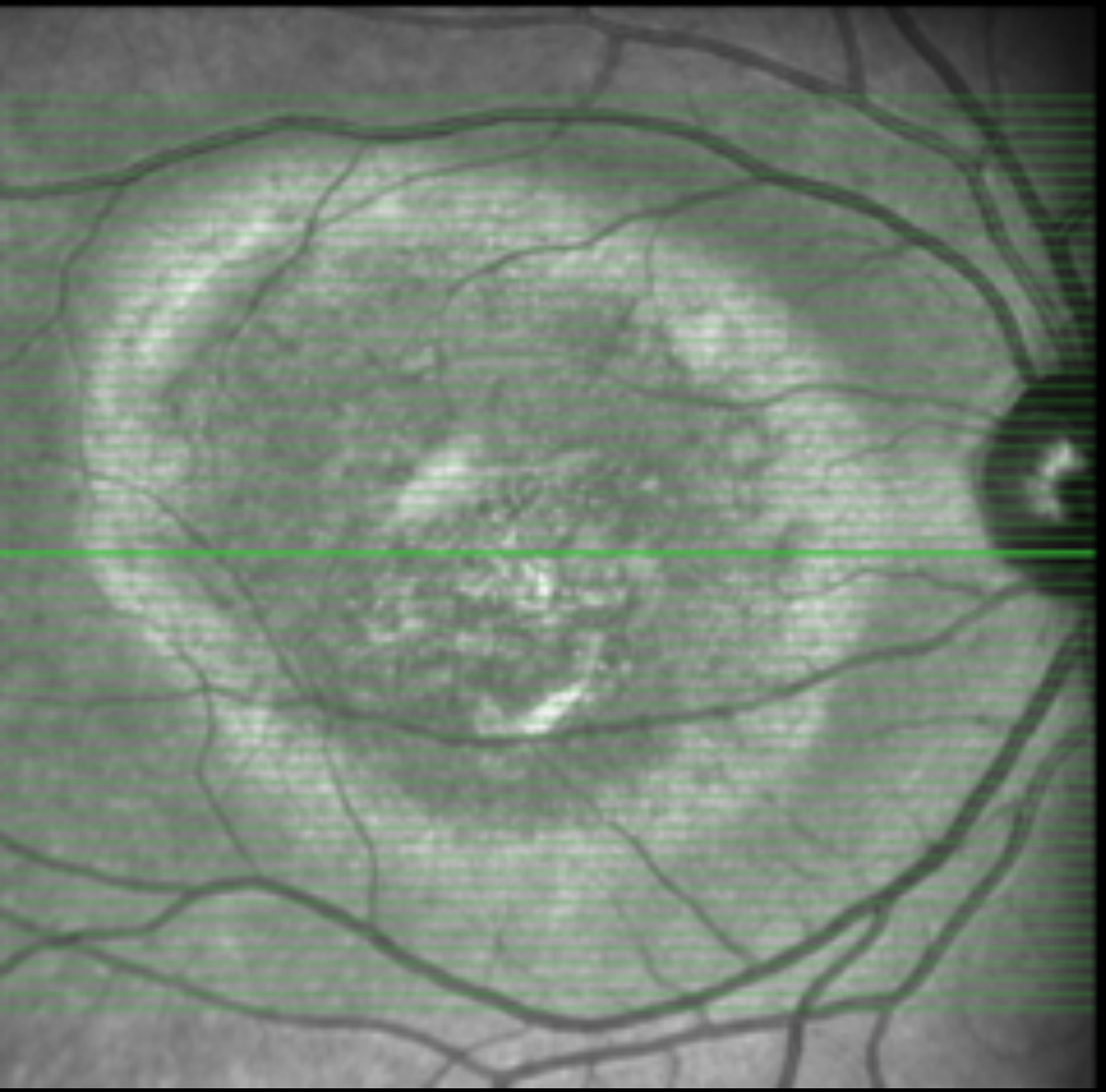


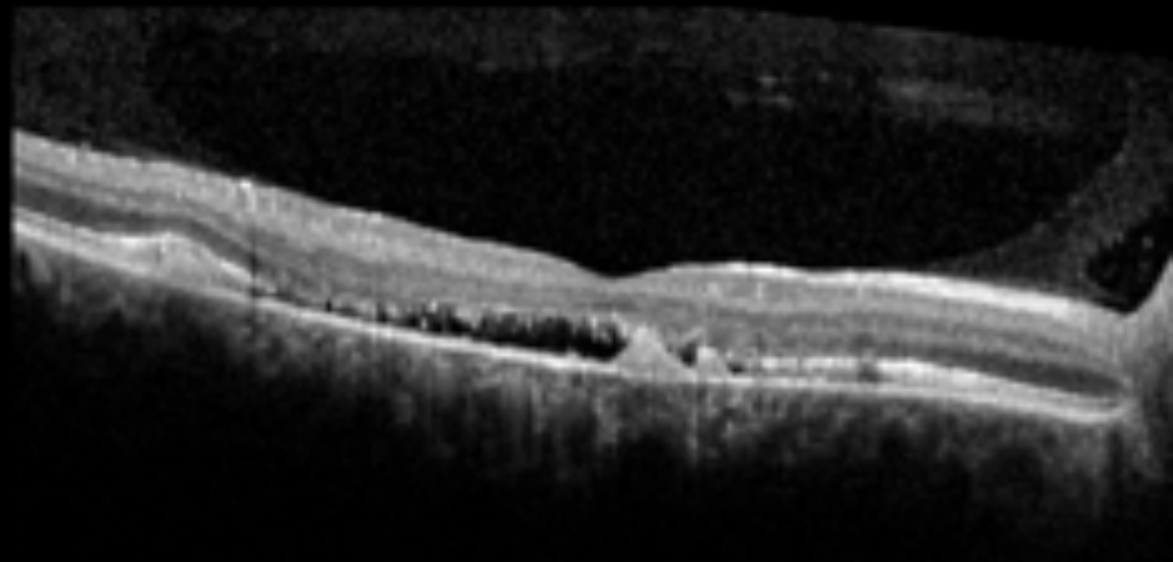
3/27/19





2/23/20





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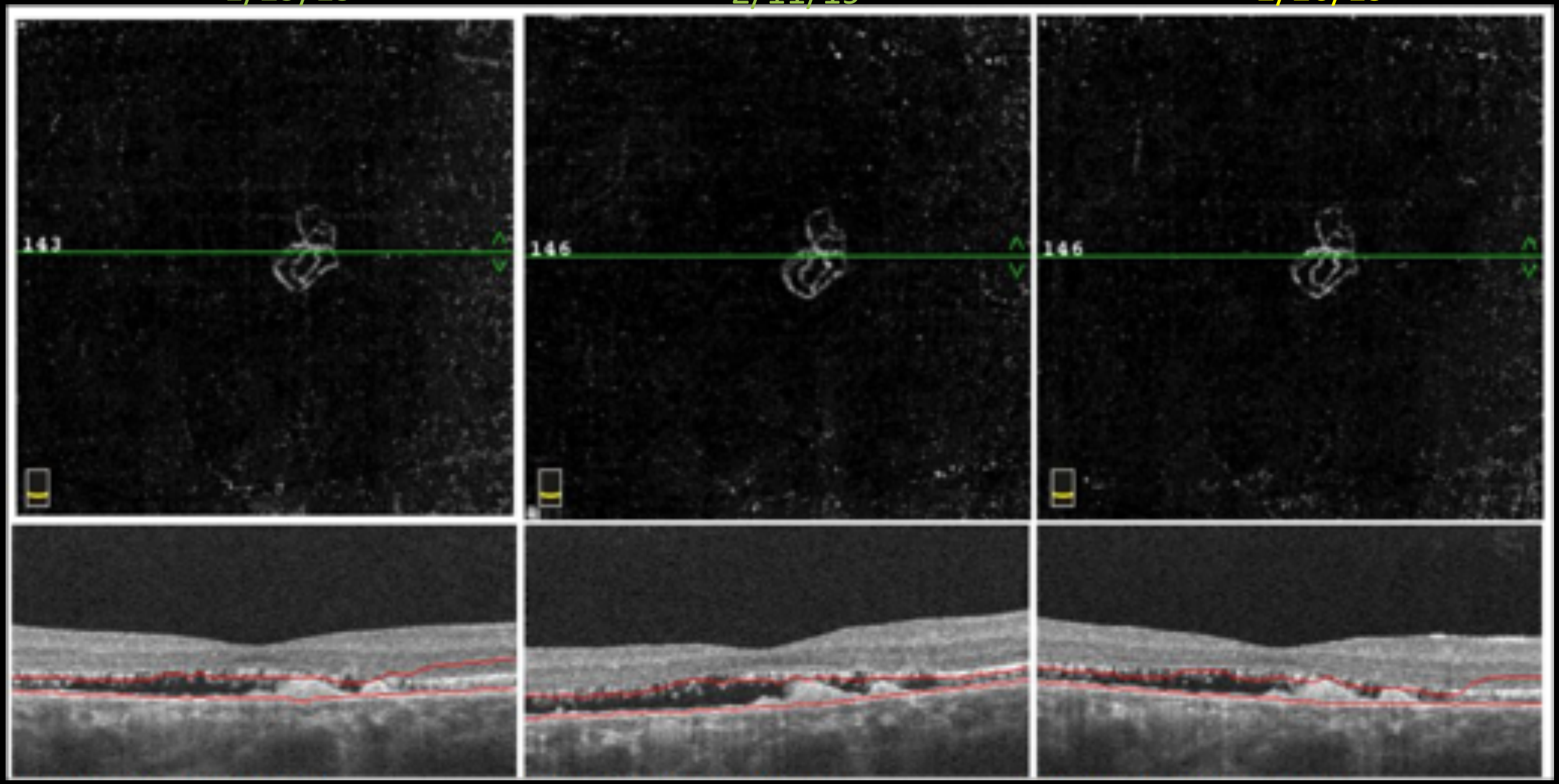

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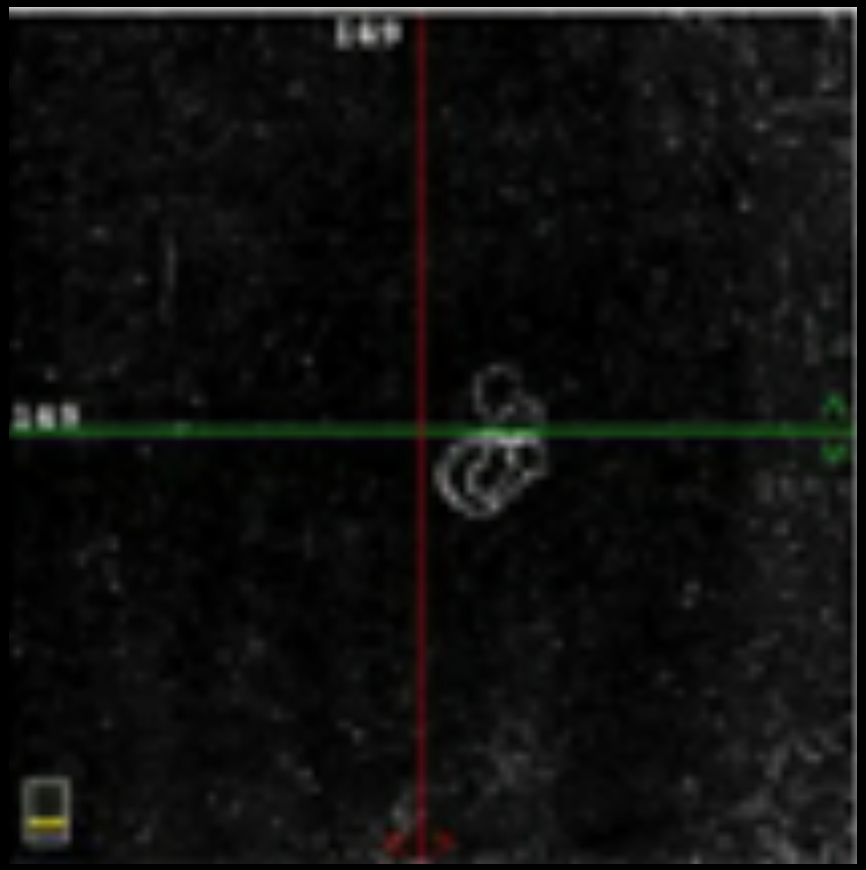

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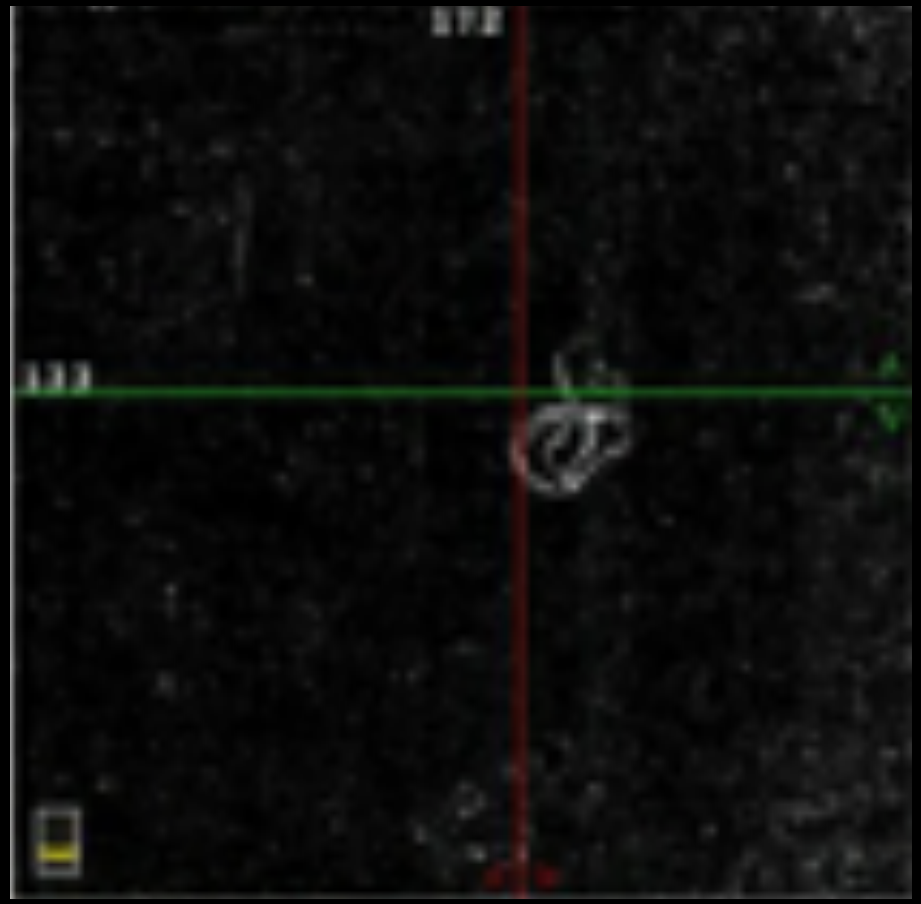
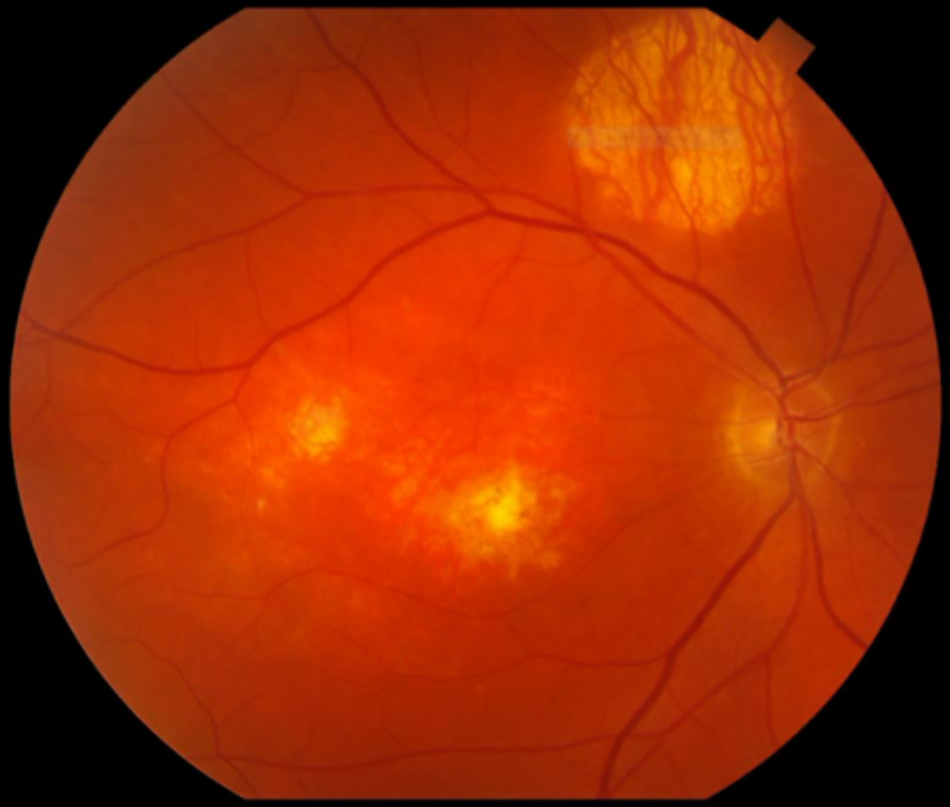
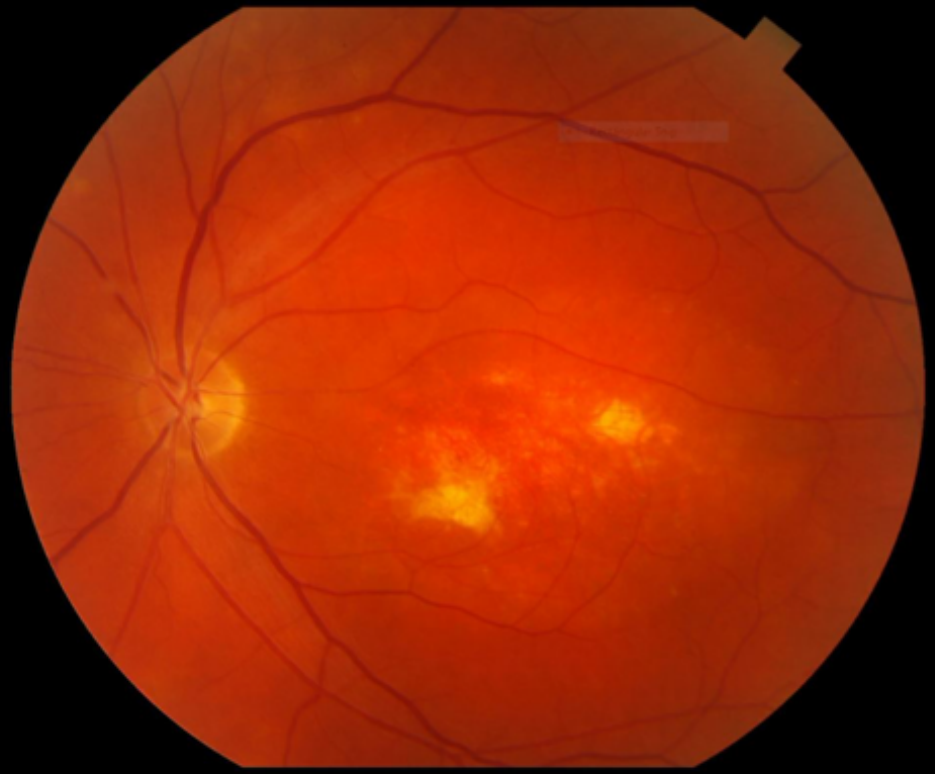
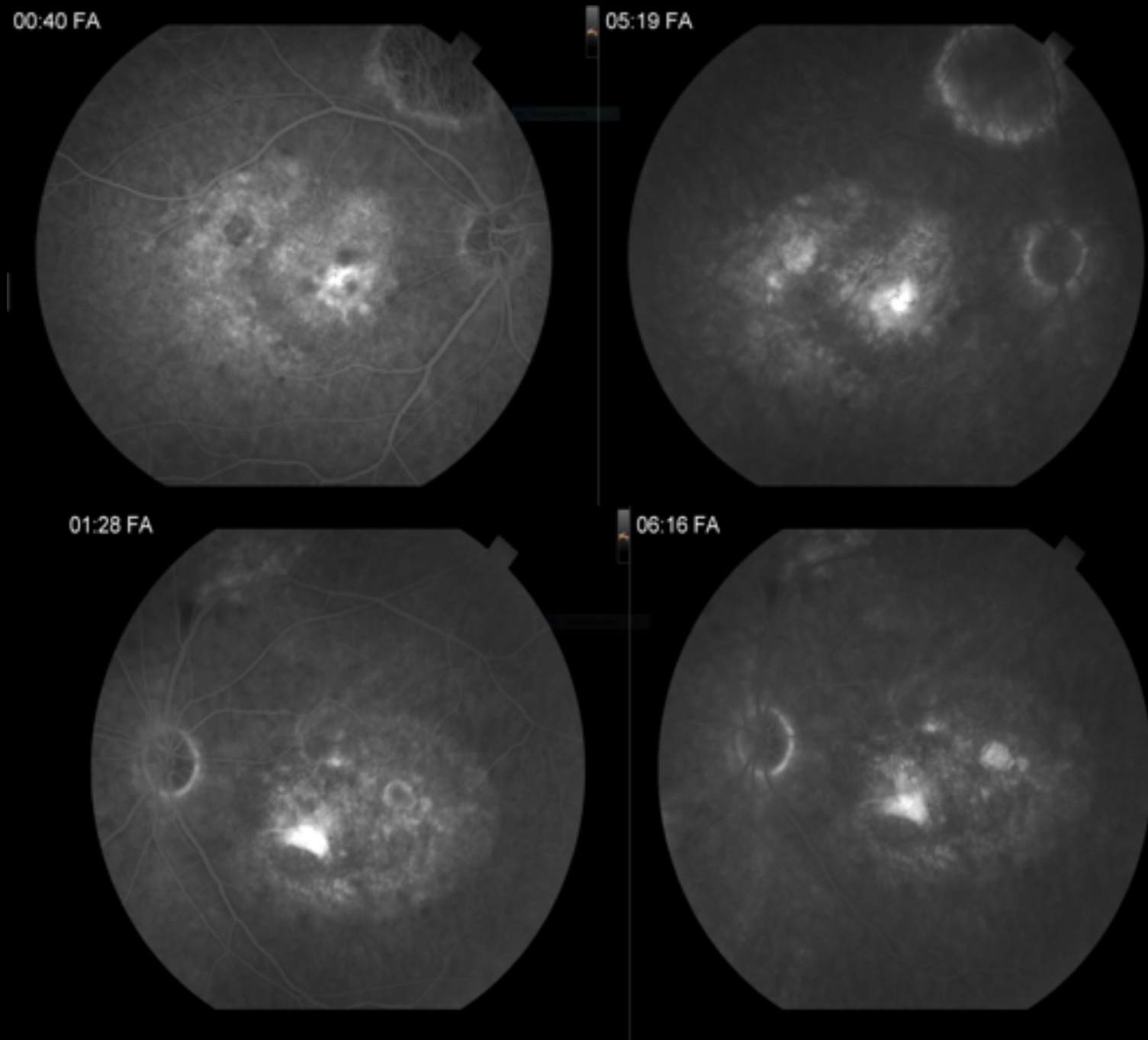


Case 3

- 71yo M with longstanding BMD diagnosis
- BCVA: 20/150 bilaterally
- Managed conservatively for 11 years



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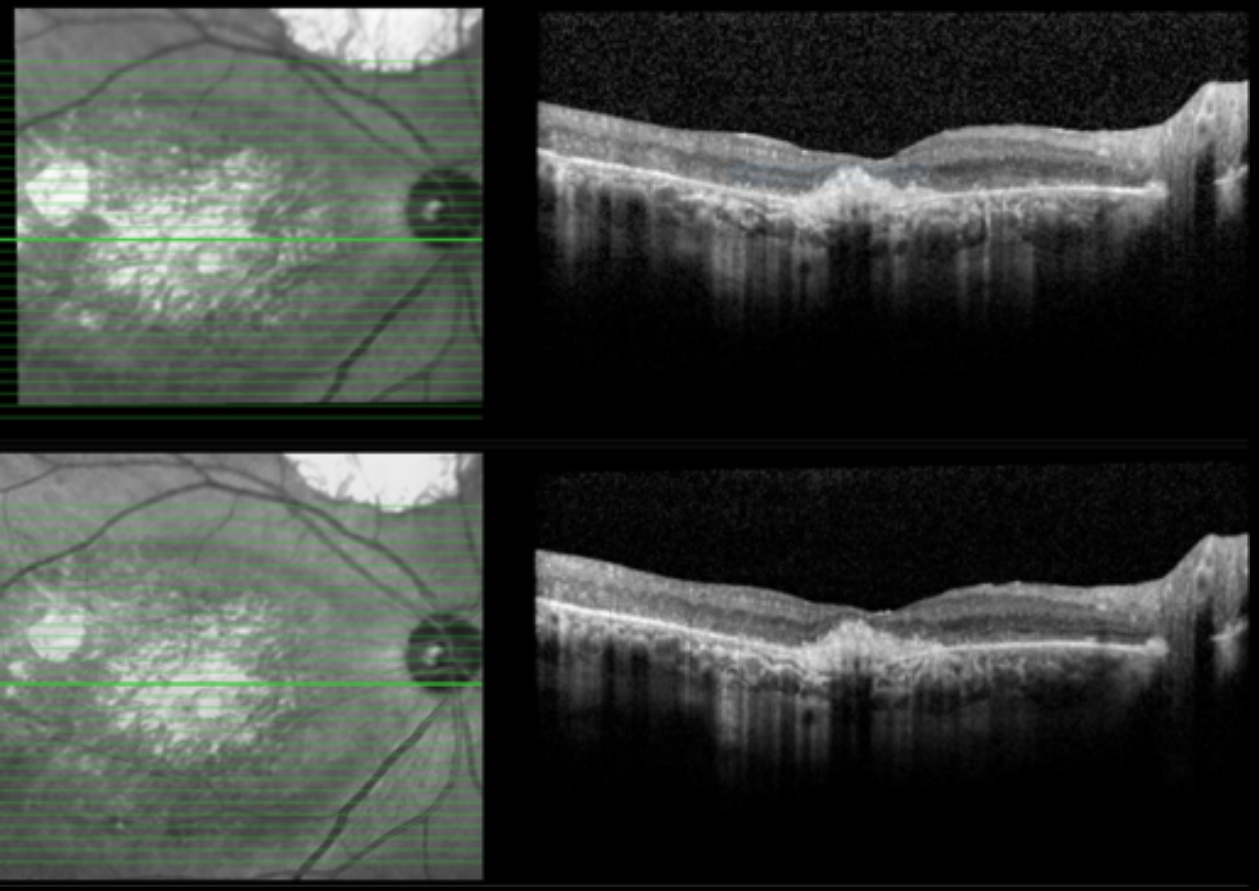
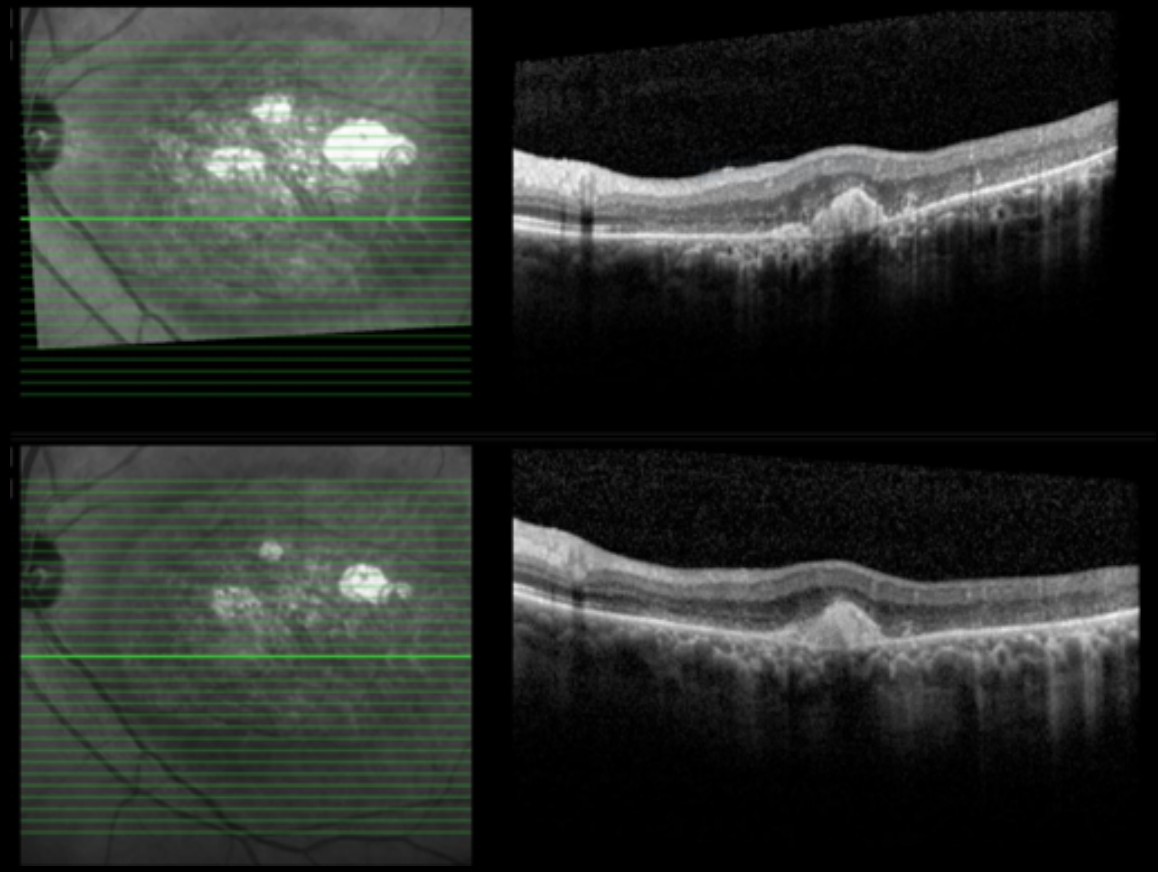
3/5/19





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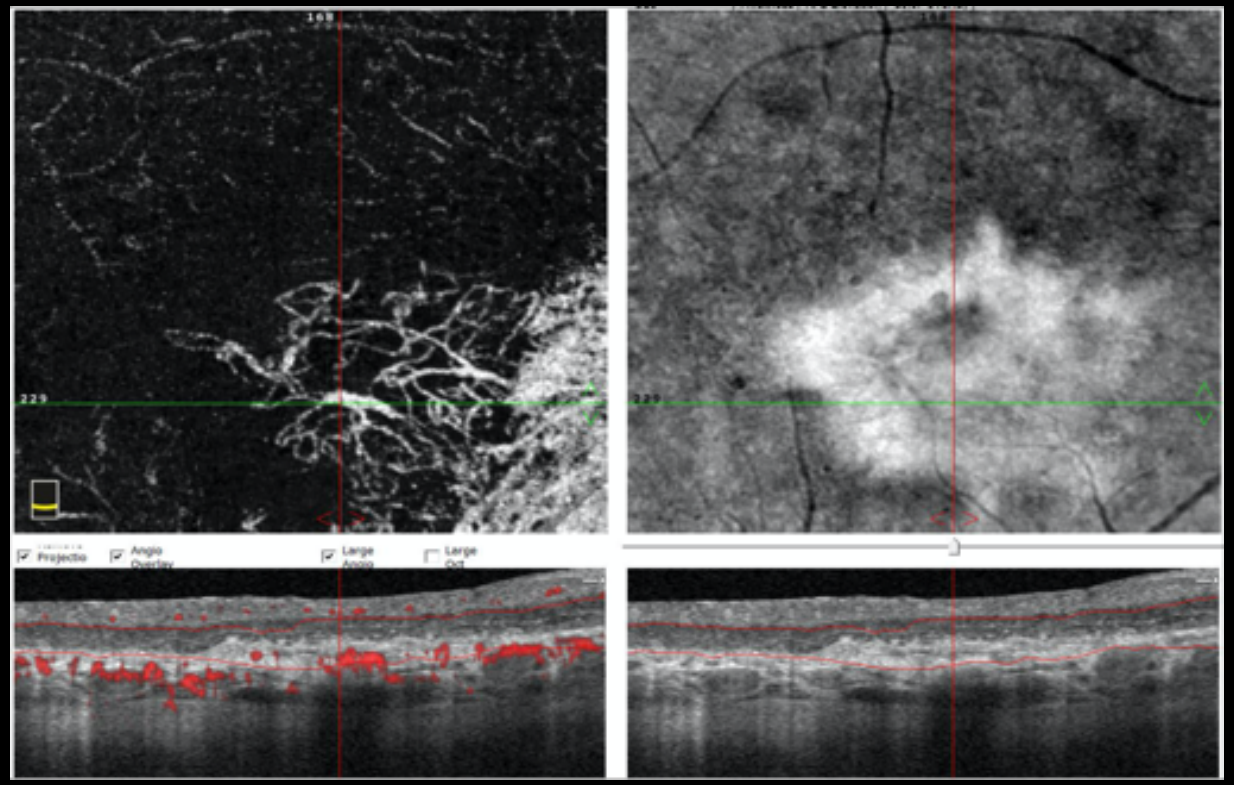
OD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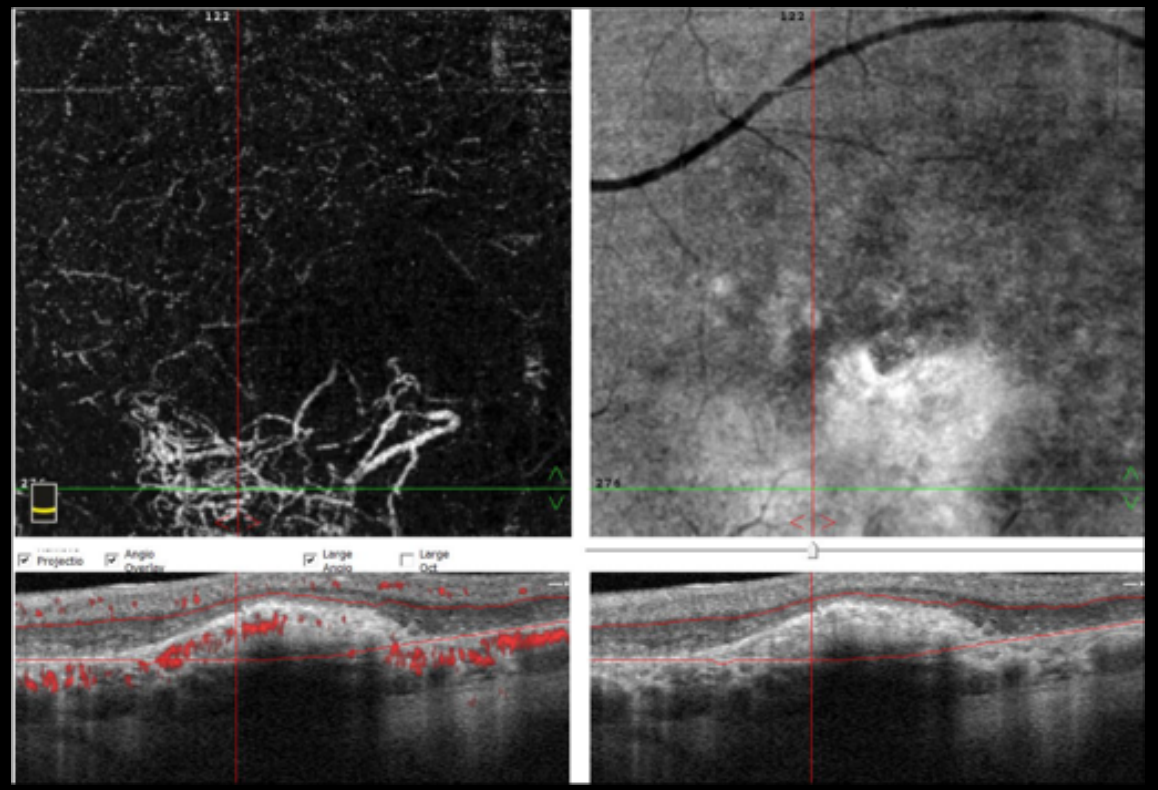
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Large Angio
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Discussion

- Six stages of disease: pre-vitelliform, vitelliform, pseudohypopyon, vitelliruptive, atrophic, choroidal neovascularization
- CNV at various stages of BMD (two vitelliruptive, one atrophic)
- OCTA: increased sensitivity in CNV detection (39% vs. 2-9% with FA)
- Reclassification of stages may be needed
- Challenges to therapeutic management as demonstrated by cases

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