



## **Brolucizumab and Multimodality Imaging in the Treatment of Angioid Streak Related Choroidal Neovascular Membrane**

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**Purpose**

*To report a case of Proactive Treat and extend usage of brolocizumab in a relatively uncommon cause of choroidal neovascular membrane with Angioid streaks and optic disc drusen in a young patient.*

**Case:** *41- yr.-old Male, presented with sudden onset metamorphopsia in Right eye.*

*The right eye presented with juxta foveal subretinal choroidal neovascular membrane with angioid streaks around the disc.*

*Multimodality imaging showed both eyes optic nerve drusen with angioid streaks and posterior pole drusens. Left eye showed similar angioid streaks but no membranes.*

*This gentleman underwent every 4 weeks, 6 mg of brolocizumab intravitreal for 12 weeks and subsequently maintained on 2 weeks treat and extend regime*

**Results:** *Following the first dose of brolocizumab the membrane size reduced, fluid reduced in all compartments. Vision improved from 6/12 to 6/6. No evidence of intraocular inflammation or any other adverse event. The improvement sustained for 3 months with continued resolution of edema.*

*The other eye closely monitored for development of CNVM.*

**Conclusion:** *Multimodality imaging and newer AntiVEGF offers extended options in terms of early detection and better management in extended spectrum of choroidal neovascular diseases. However suggest a larger case control studies to support the usage of the antivegf in Angioid streaks and to establish as a preferred practice.*

**Keywords:** *Brolucizumab, Angioid streaks, multimodality imaging, optic nerve drusen.*

## Introduction

Angioid streak (AS) is a reddish jagged line radiating from the optic nerve which represents dehiscence of Bruch's membrane following deposition of minerals and runs close to the macula or beyond.[1] AS is an ocular manifestation of Pseudoxanthoma elasticum, Paget disease, Ehlers-Danlos syndrome. [2]

Choroidal neovascular membrane (CNV) is one of the complications leading to significant vision loss and hence the need for early detection and treatment.

Several treatment modalities have been proposed in the past such as Transpupillary thermotherapy, Photodynamic therapy [3] but these treatments have not proved enough to be safe enough for the retina and these treatments have been overtaken currently by the Anti-Vascular growth factors (Anti-VEGF).

Various Anti-VEGF [4-6] like Bevacizumab, Ranibizumab, Aflibercept, Brolucizumab has been used in the past for the treatment of Age-related macular degeneration. Based on the beneficial effect of these Anti VEGF and its usage currently being extended to uncommon cause of CNV like angioid streaks also since no approved treatment currently available and increased durability.

Brolucizumab 6 (Beovu) is a VEGF inhibitor that was approved for the treatment of nAMD in October of 2019 following two, Phase-3, randomized clinical trials: HAWK and HARRIER. Based on the positive outcome of durability study with anti-VEGF for Q8W & Q12W Beovu could be a potential treatment for Angioid streaks.

## Clinical Methods

Our case of interest is a 41-year-old male who presented with recent onset of metamorphosia and scotoma in right eye for 3 weeks

He never had systemic illness in the past. On examination his skin condition was ruled out for PXE and cardiovascular and haematological conditions.

Ocular examination showed BCVA RE 6/12; LE 6/6 rest of the anterior segment examination was unremarkable. Fundus examination showed premacular and juxta-macular hemorrhage in the PMB zone & angioid streak in both eyes radiating from optic nerve. Optic nerve showed drusen more prominent in left eye with both eyes posterior pole drusen. Left eye had no sign of activity.

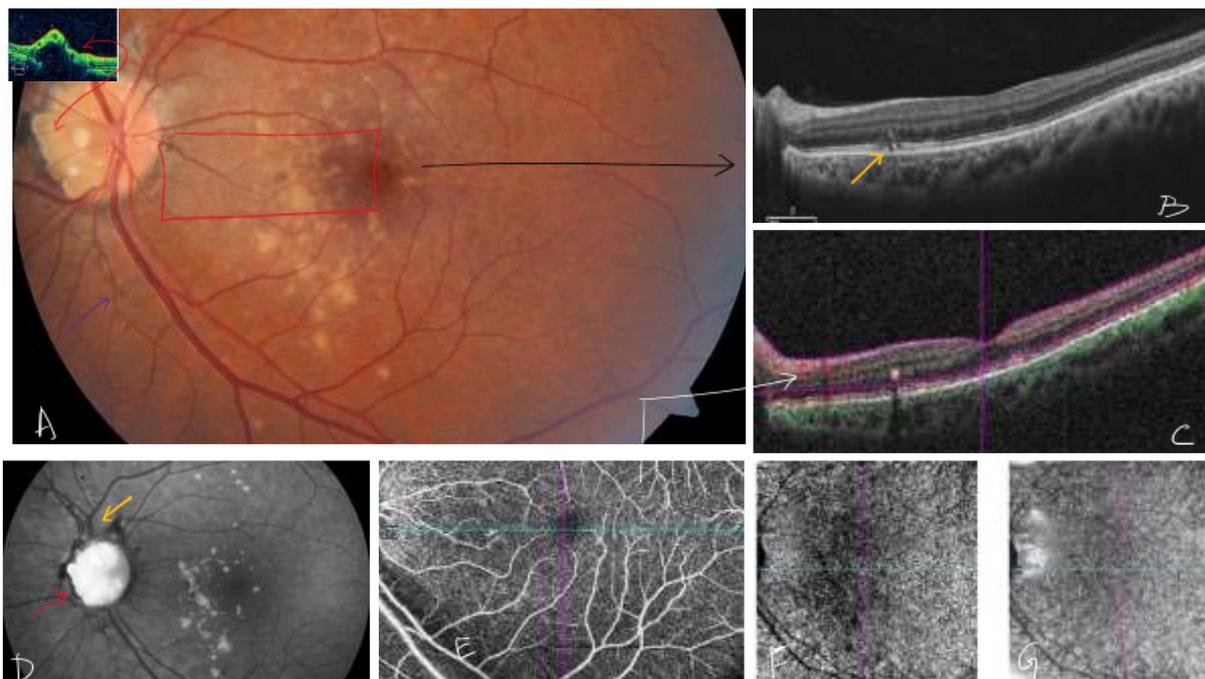
Underwent multimodality imaging with Zeiss Cirrus SD OCT 5000 & OCT angiography (OCTA), Zeiss visucam Fundus photography, (FP) autofluorescence (AF) and Fluorescein angiography. (FA)

RE found to have type 2 CNV with subretinal fluid and intraretinal drusens. LE found to have no activity which is confirmed by OCT, OCTA & FA

After explained consent and the possible adverse effects of brovacizumab and after ruling out intraocular inflammation the patient is planned for q4w 3 doses and followed by proactive treat and extend regime.

Patient underwent 3 doses accordingly and currently on 2 weeks extension. He responded well with one dose of Beovu and vision improved to 6/9 RE with resolution of subretinal fluid and premacular haemorrhage.

At the end of 3 doses his BCVA improved to 6/6 with remarkable anatomical resolution without any evidence of intraocular inflammation. He has been continuously monitored for the same.

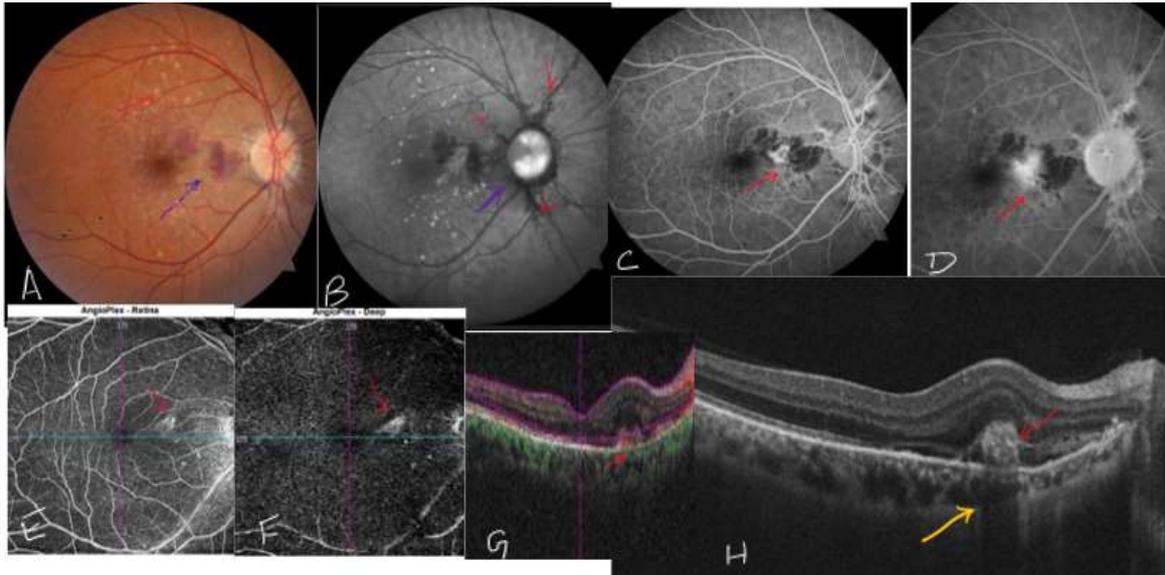


**Figure 1.** Multimodality imaging of Left eye with Angioid streaks and Optic nerve drusen

Multimodality imaging of Left eye with Angioid streaks and Optic nerve drusen:

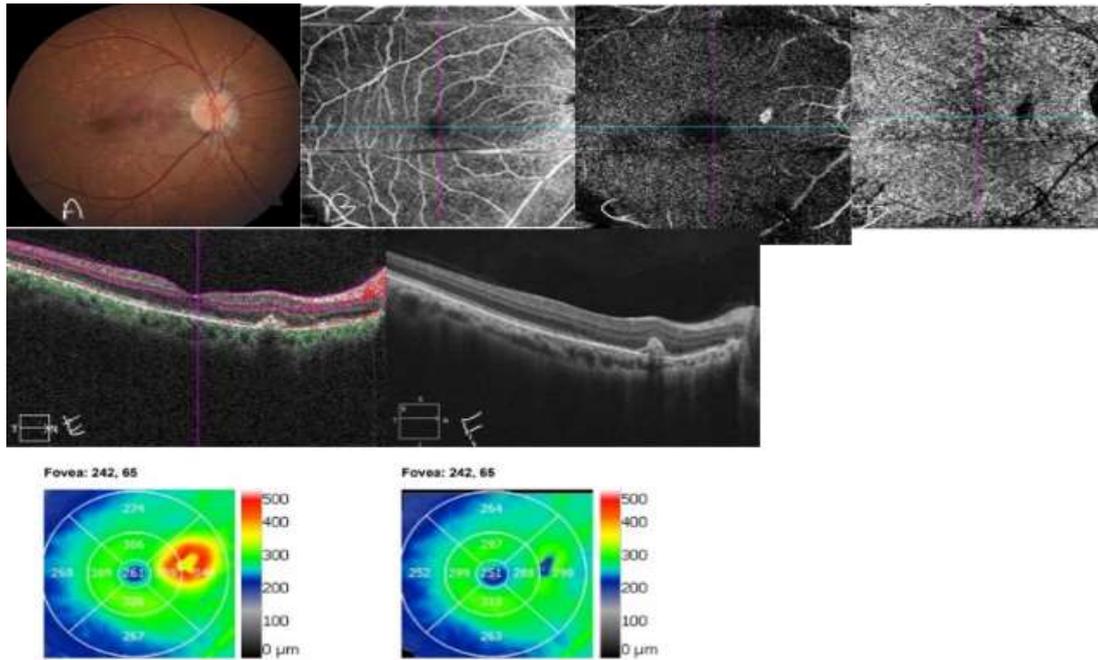
- A) Fundus photo showing optic disc drusen with angioid streaks and posterior pole drusens
- B) SD-OCT: on the angioid streak showing early Early RPE disruption and sub ELM reparative process.

- C) Angioplex Flow: No signs of activity which is a useful tool to monitor the onset of neovascularisation in a high risk setting.
- D) Autofluorescence imaging: optic nerve and posterior pole drusen with angiod streak as a jagged lines radiating from optic disc
- E) Angioflex superficial and deep plexus at the site of angiod streak which confirms quiescent state.



**Figure 02:** Multimodality imaging of Right eye: Baseline

- (A) Colour fundus photo showing Cnvm with Hemorrhage [purple arrow]Drusen[red arrow];
- (B) Autofluorescence imaging :Angiod streak[Red], Optic nerve head drusen;
- (C) FA : early and late phase showing leaking cnvm with staining do angiod streaks radiating from optic disc
- (E-F) Angioplex images with delineated close loop vascular membrane with core;(G) Flow image confirming the vascular activity [red arrow];
- (H) SD-OCT showing type 2Cnvm with serous detachment and pachydrusen [yellow]



**Figure 3:** Right eye: After 3 loading dose of Brolucizumab intravitreal injection

- (A) : Fundus photo showing resolving hemorrhages
- (B-D) : OCTA angioplex superficial to deep showing resolving membrane
- (E) : Flow angioplex shows reduced activity
- (F) : SD-OCT Shows reduced subretinal fluid and membrane
- (G) : Macular cube comparison showed reduced Juxta-foveal thickness.

## Discussion

Angioid streak related CNV has potentially blinding complication due to its aggressive nature and its systemic consequence.

The need for more efficacious and durable treatment is a long unmet need. Although other anti-VEGF treatment found to be useful in the management, the use of brolucizumab could fill the need in terms of durability and efficacy. However, a wider RCT will be required to substantiate the usage of this molecule on a long run and also taking into consideration the incidence of intraocular inflammation in the past.

With regard to the multimodality imaging the earlier detection in an asymptomatic angioid streaks will be certainly beneficial in preventing vision loss. OCTA and SD/SS OCT guides in examining the streak and the cnv pattern for prognostication.

Effective use of Anti-VEGF and multimodality imaging will certainly support in the management of angioid streak.

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