Improving clinical management of macular neovascularisation secondary to angioid streaks

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Introduction

Angioid streaks(AS) occur due to breaks in a degenerated and weakened Bruch's membrane and typically radiate outwards from the optic disc. They could be idiopathic or associated with systemic diseases such as pseudoxanthoma elasticum(PXE), Ehler-Danlos syndrome, Paget's disease, Sickle cell disease, and other hemoglobinopathies.[1] Macular neovascularisation (MNV) is a common cause of central visual loss and occurs in up to 42% to 86% of cases with bilateral involvement in up to 71%.[2] This clinical practice review aims to compare our treatment outcomes with published literature and define a protocol for optimal management AS and related MNV.

Method

Data pertaining to patient demographics, management of MNV, and visual outcomes of patients with AS-related MNV, seen in St Paul's Eye Unit, Royal Liverpool University Hospital between 2014-2022, was collected retrospectively from electronic patient records.

Results

The total number of patients with AS was 24; 6 patients without MNV were excluded. Of the 18 patients (10 males; 31 eyes) with an average age of 57.9 years, 55.6% had a diagnosis of PXE; 72.2% had bilateral MNV. Mean follow-up was 48 months (4 months–13 years). (Fig 1) The location of MNV was subfoveal, juxtafoveal, and extrafoveal in 32%, 32%, and 36% of eyes, respectively. (Fig 2)

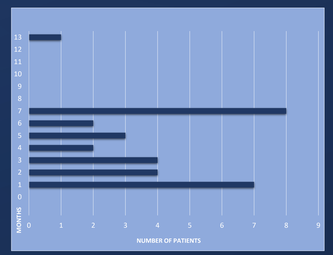


Fig 1 : Follow-up duration

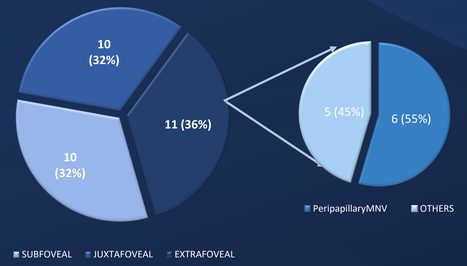


Fig 2: MNV Location

Twenty-one eyes (67.7%) received intravitreal anti-VEGF therapy, of which 11 had also received photodynamic therapy (PDT) (Fig 3). The mean number of injections per eye over the total period of follow-up was 20.6 (Fig 4), reducing to 16.7 (range 2-45) when excluding one patient who had 89 injections.

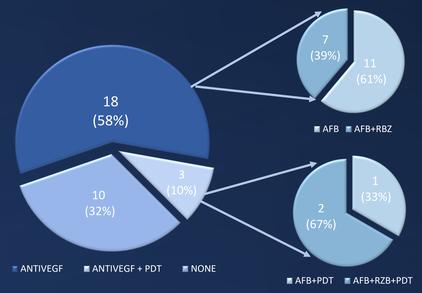


Fig 3: Treatment received

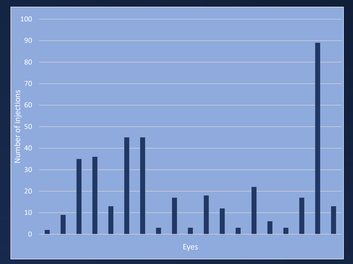


Fig 4 : Number of injections

The mean best-recorded logMAR visual acuity at baseline and final follow-up were 0.57 and 0.50, respectively. 52% of eyes gained, 29% lost, and 19% maintained baseline visual acuity at the final follow-up. One incidence of endophthalmitis with recovery to baseline visual acuity was noted. At final follow-up, central retinal thickness (CRT) on macular OCT was lower in 12 eyes (57.2%), remained stable in 8 eyes (38%), and was worse in 1 eye (4.8%) compared to baseline (Fig 5).

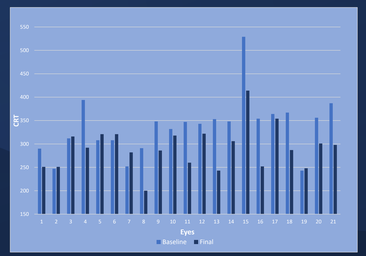


Fig 5: Change in CRT

Discussion

Although our visual outcomes are comparable to published studies[3] with different anti-VEGF agents using *p.r.n.* regime, our patients received a higher average number of injections per eye, even when OCT-derived signs of MNV activity were stable. Given the rarity of AS-related MNV, there are no established guidelines to inform treatment decisions. These results have prompted us to devise a departmental protocol to aid clinical decision-making. (Fig 6).

Conclusion

Though anti-VEGF therapy can stabilise or even improve visual outcomes in AS-related MNV, MNV can be tenacious with a high recurrence rate, often requiring long-term continuous anti-VEGF therapy, as seen in our case series. Our results are affected by the retrospective nature of the review and a high degree of subjectivity in re-treatment decisions by clinicians. We have defined precise re-treatment criteria as part of the management pathway, which we believe will prevent under or over-treatment and aim to re-audit our results in future. Given the limitations of our small sample size, further prospective studies with larger sample sizes and longer follow-up duration are required to support our conclusions.

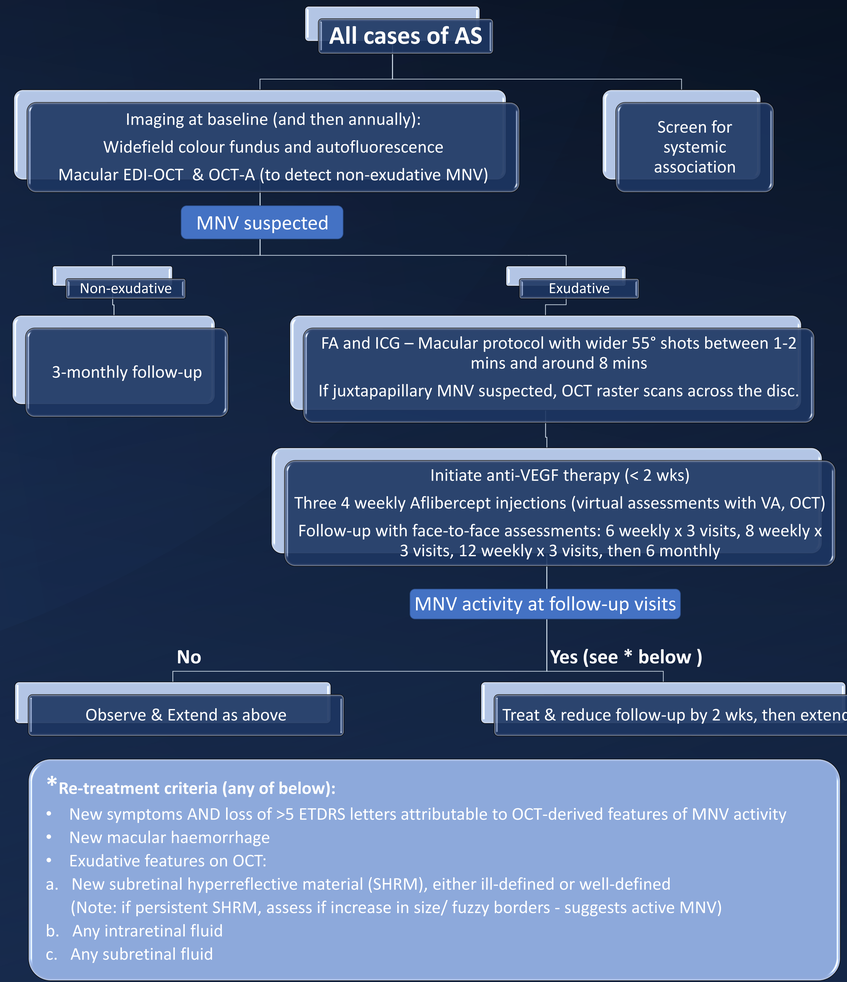


Fig 6: St.Paul`s AS-related MNV management protocol

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