Adult vitelliform macular degeneration: a new United Kingdom perspective.

Prasad Rao[1], Su Nyunt Zaw[1], Naveen Garikapati[1], Komal Godbole[1], Pratibha Veeramani[2], Lakshika Perera[1].

Shrewsbury and Telford Hospitals NHS Trust[1], Shrewsbury and Surrey and Sussex Healthcare NHS Trust, East Surrey[2].

Introduction

With the emergence of new imaging technology adult vitelliform macular degeneration (AVMD) is being diagnosed with increasing frequency over the last two decades. AVMD is often mistaken for wet age-related macular degeneration (ARMD) when the vitelliform material starts scrambling showing pseudo fluid in optical coherence tomography (OCT) scans. In addition, in some AVMD patients the vitelliform lesions in fact do develop choroidal neovascularisation (CNV) requiring urgent intravitreal injections. We aim to highlight the natural progression and treatment options in patients with AVMD.

Results

we have retrospectively analysed 123 eyes of 82 patients with AVMD who presented to this department over the last 4 years.

We identified 44 females and 38 males, 41 out of a total 82 patients had bilateral involvement at presentation.

The clinical findings varied from a classical sub foveal yellow lesion (Fig 1) to partial empty spaces within the lesion.(Fig 2), to complete atrophic scar following resorption of all the colloid material.

The presenting Visual acuity in the worst eye also varied from HM to 6/9 according to the stage of degeneration they presented with.

Thirty-nine patients were asymptomatic and AVND was an incidental finding in one of their eyes.

All patients had OCT, optical coherence tomographic angiography (OCTA), and fundus autofluorescence (FAF). Some patients in addition underwent fundus fluorescein angiography (FFA) and indocyanine green angiography (ICG) when a choroidal neovascular membrane (CNVM) was suspected.

We have also analysed serial OCT scans to study the progress whenever old scans were available.

Sixty nine out of 82 patients did not need any active intervention.

Fourteen patients were already taking antioxidant supplements.

Thirteen patients developed significant subretinal fluid (SRF) and were treated with intravitreal anti-VEGF injections.

On further review of the OCTA scans and FFA only 10 of 13 patients showed active CNVM. The other 3 patients did not have any evidence of CNVM (Table 1: Patients 2, 8, 12)

Therefore only 12 % of AVMD patients seem to have developed wet ARMD in our series.

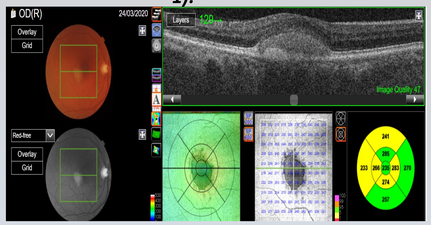
Response to the anti-VEGF injections was suboptimal in AVMD patients with CNV compared to conventional wet ARMD patients. This unfavourable response was most likely due to pre-existing long standing atrophic and degenerative changes.

In three patients who were found not to have CNV, the anti VEGF injections were ineffective as expected.

Of the other 10 patients, treatments are ongoing in 5 but have already shown a trend to declining vision. Of the remaining 5 where discontinued, 3 patients injections proved to be ineffective leaving only 2 patients with stable vision.

Contrary to earlier publications, based on family history alone we did not find any strong evidence of autosomal dominance inheritance therefore we prefer to call it a degenerative rather than dystrophic condition.

Early case of AVDM (Fig 1).



- ***Late case of AVDM showing empty space following resorption of the colloid material (fig 2).***

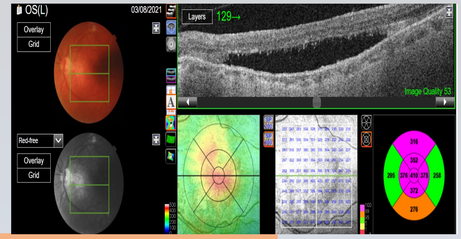


Table 1: AVMD patients treated with Anti-VGEF for secondary wet ARMD

No	Name	Age	VA R Letters	VA L Letters	Affected eye	Rx	Comment
1	GW	68	71	84	Right	3	On going
2	PW	95	20	45	Both	2 to each	No CNV, ineffective
3	AH	69	54	75	Right	3 Eylea	Ineffective
4	PW	82	64	63	Right	3 Eylea	On going
5	LF	75	69	25	Right	4 Eylea	On going; VA improved by 13 let
6	PM	85	67	69	Left	5 Eylea	Ongoing
7	SN	58	40	42	Both	4 Eylea to BE	Ineffective Rx
8	IC	67	73	71	Right	4 Eylea	Ineffective; no CNV
9	KS	65	65	84	Right	3 Eylea	On going; no improvement
10	RR	76	45	70	Right	8 Eylea	No improvement
11	GL	79	60	76	Right	20 Eylea	No improvement
12	EP	98	35	35	Right	14 Eylea	Empty space; no CNV
13	WH	86	78	78	Left	9 Eylea	stable

Conclusion

➢With increasing use of OCT scans by optometrists and primary care physicians more patients are being diagnosed with macular pathology and AVMD is one such condition. Most of these patients do not need any active intervention or long term follow up in hospital clinics. Our study shows only 12% of AVMD patients go on to develop CNV requiring intravitreal anti-VEGF injections. As our cohort shows it may not be unusual to mistake passive fluid lakes within vitelliform lesions for active wet ARMD.

➢It is important for clinicians to be aware of the possibility of AVMD with passive subretinal fluid and not to resort to unnecessary long term intravitreal anti-VEGF injections. As we have shown even in cases of AVMD with secondary CNV the prognosis is poor for long term retention of central vision.