A Case Report of Adult Vitelliform Macular Dystrophy.

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Fundoscopy: (Re)- Yellowish Oval Shaped Complaints Of Diminution Of Vision In Right Eye Since 2 Years. It Was Insidious In Lesion In The Macular Area Of About 1.5 Dd Resembling Egg Yolk .(Le) With In Normal Limits. Ffa And Oct Was Advised


Conclusion: Based On The Above Findings A Diagnosis Of Adult Onset Vitelliform Macular Dystrophy (Avmd) Was Made. Patients With Adultonsetmacular Vitelliform Dystrophy Typically Have Slowprogressive Vision Loss. However, Patients Can Develop Dramatically Decreased Vision Owing Tosubfoveal Choroidal Neovascularization (Cnv). Thus, It Is Important To Establish The Correct diagnosis And Monitor This Condition.

Keywords: Avmd, Best Disease, Macular Dystrophy, Pattern Dystrophy, Vitelliform Lesion.

I. Introduction

The Classical Vitelliform Macular Lesion Characterises Best's Vitelliform Dystrophy, But In Adults Similar Lesions Are Less Well Categorised. In 1974, Gass¹ Described A ‘Peculiar Foveomacular Dystrophy’ Which He Later Termed ‘Foveomacularvitelliform Dystrophy: Adult Type². The Hallmark Of The Condition Is A Yellow, Slightly Elevated Lesion, One-Third To One Disc Diameter In Size, And Often With Pigment In The Form Of A Spot, Figure, Or Ring. At Times The Central Pigment Clump Is The Predominant Feature, Surrounded By A Hypopigmented Halo. As A Result, Many Different Terms Have Been Applied To This Picture, Mostly Emphasising The Yellow Vitelliform Lesion,²,³,⁴ Less Commonly The Pigment.⁵,⁶,⁷

A Hereditary Trait associated With Mutations In Vmd-2 And Rds Gene With Autosomal Dominant Character Has Been Established In Some Cases But Not In Others, So That It Has Variousy Been Termed A Degeneration⁸,⁹,¹⁰ Or A Dystrophy,²,³,⁵,¹¹ In This Report It Is Termed Adult Vitelliform Macular Degeneration (Avmd). The Disease May Present Different Phenotypes In The Same Family,¹² Or In The Two Eyes Of The Same Individual,¹⁰,¹¹ Or Even In The Same Eye At Different Times.¹⁰

Vitelliform Lesions Represent The Effects Of Retinalpigment Epithelium (Rpe) Dysfunction With Accumulation Ofdegenerated Photoreceptor Outer Segments In The Subretinal Space. The Onset Of Disease Is Usually 3rd To 6th Decade With Minimal Visual Loss And Metamorphopsia. We Report A Case Of Avmd With Unilateral Presentation.

II. Case Report

A 59 Years Old Male Patient Presented With Complaints Of Diminution Of Vision In Right Eye Since 2 Years. It Was Insidious In Onset, Gradually Progressive In Nature And
Painless. It was associated with metamorphopsias in Amsler Test. No H/O of similar complaints in the family. Slit lamp biomicroscopic examination (Ou)- With in normal limits.

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1. Investigations
1.1 Dilated Fundus Examination

**Figure 1:** Right eye-yellowish oval shaped lesion in the macular area of about 1/2 Dd resembling eggolk we
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1.2 Fundus Fluorescein Angiography

Figure 2: Left Eye - Appears Within Normal Limit

Figure 3: Right Eye - Arteriovenous Phase. Central Blockage With Ambient Hyperfluorescence. (Central Hypofluorescence Surrounded By Ring Of Hyperfluorescence)
Figure 4: Left Eye- Appears Within Normal Limits.

Figure 5: Right Eye- Subfoveal Deposit. Subretinal Hyper-Reflective Echo Causing Shadowing.
1.4 EogIt Was Found To Be Normal. Ardens Ratio ≥ 1.8.

II. Discussion
“Vitelliform” Macular Lesions Can Occur In A Variety Of Diseases That Affect The Rpe. A Broad Differential Diagnosis Exists, Ranging From Nonneovascular Amd And Cd To Acute Exudative Polymorphous Vitelliformmaculopathy (Aepvm).

In Our Case Fundus Findings (Fig 1) Were Supported By Ffa (Fig 3), Oct (Fig 5) And Eog ForAvmd. Avmd Is Known To Present With Variableexpression And Reduced Penetrance. Significant Asymmetry Between Two Eyes Was Another Feature Of This Case.

III. Conclusion
Patients With Adult Onset Foveomacularvitelliform Dystrophy Typically Have Slow Progressive Vision Loss. However, Patients Can Develop Dramatically Decreased Vision Owing To Subfovealchoroidal Neovascularization (Cnv). Thus, It Is Important To Establish The Correct Diagnosis And Monitor This Condition.
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References