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CHOROIDAL NEO-VASCULAR MEMBRANE IN THE PEDIATRIC AGE GROUP

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RIYADH /SAUDI ARABIA

EOS 2025
EGYPTIAN OPHTHALMOLOGICAL SOCIETY

Case history

CASE HISTORY

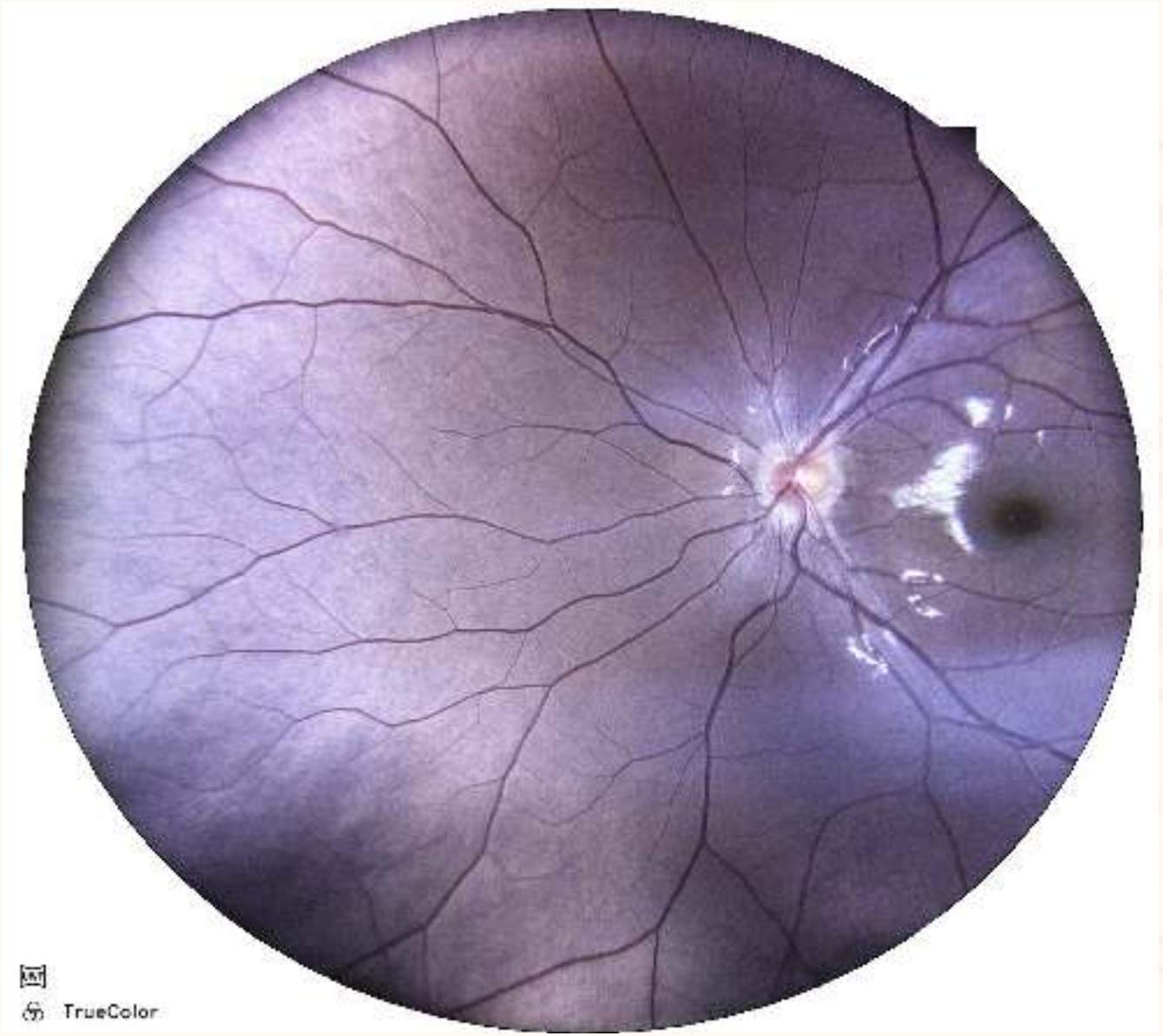
- Jouri ,l l years old healthy female
- Product of normal pregnancy and delivery
- Normal schooling and growth parameters
- No known illness current or in the past
- No family history of any ocular disease in her sibs or parents
- Complained that she noticed poor vision in both eyes few months back

EXAMINATION

- Ucva 20/100 od and 20/60 os
- SLE wnl
- CR OD +1.00 no improvement of VA
- OS -2.00-0.25@170 20/20

- Dilated fundus exam & photos

OS



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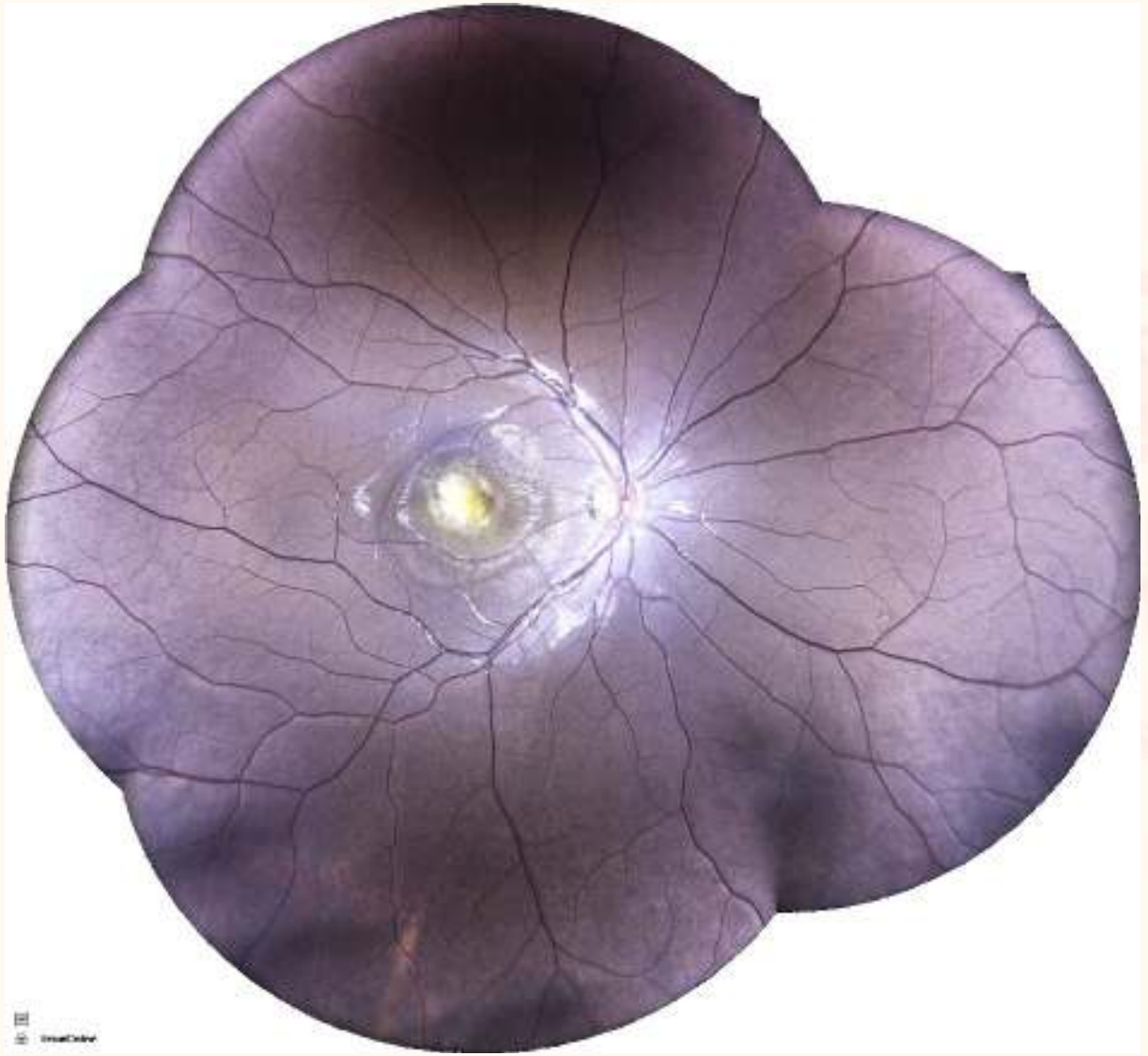

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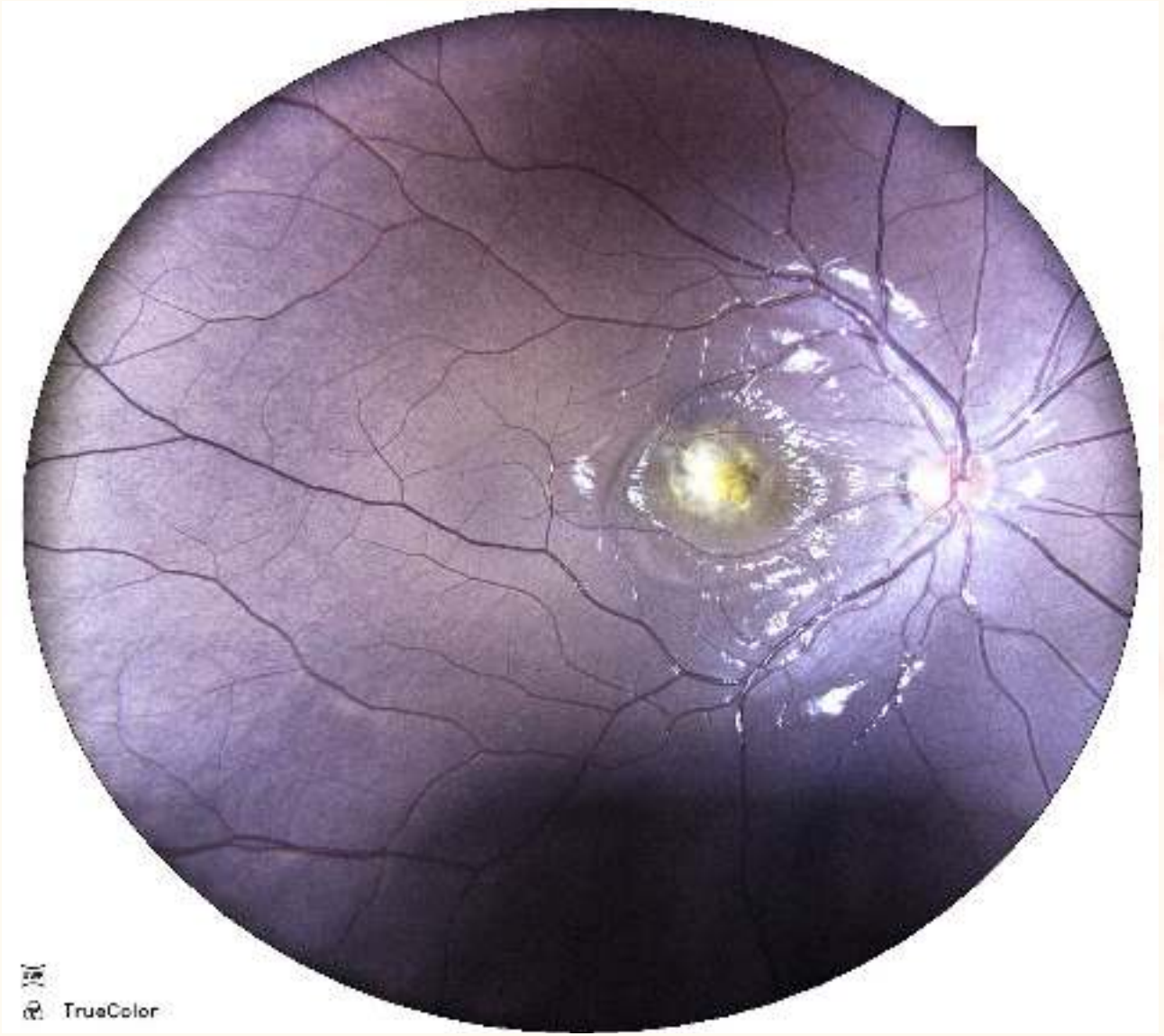

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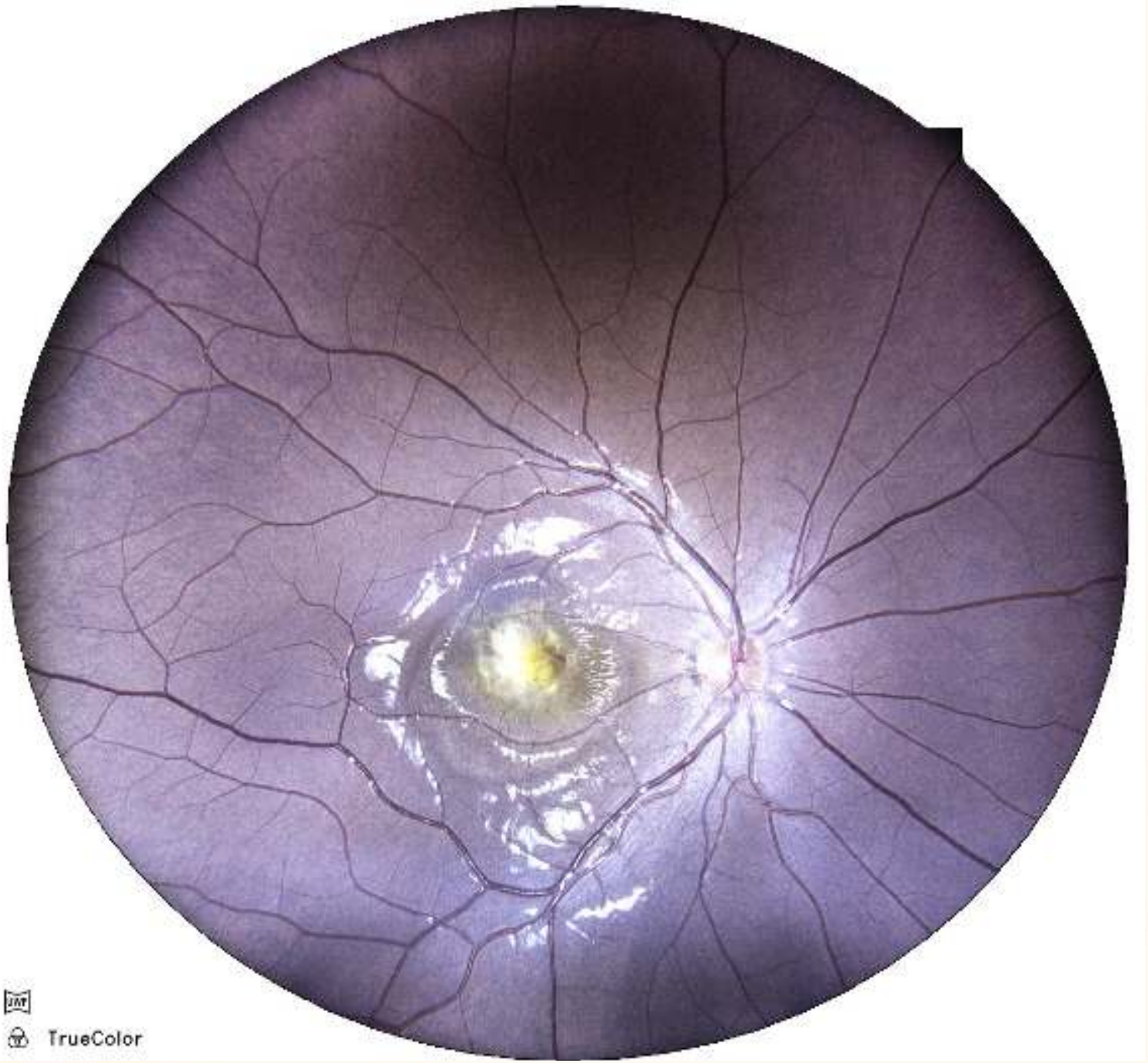

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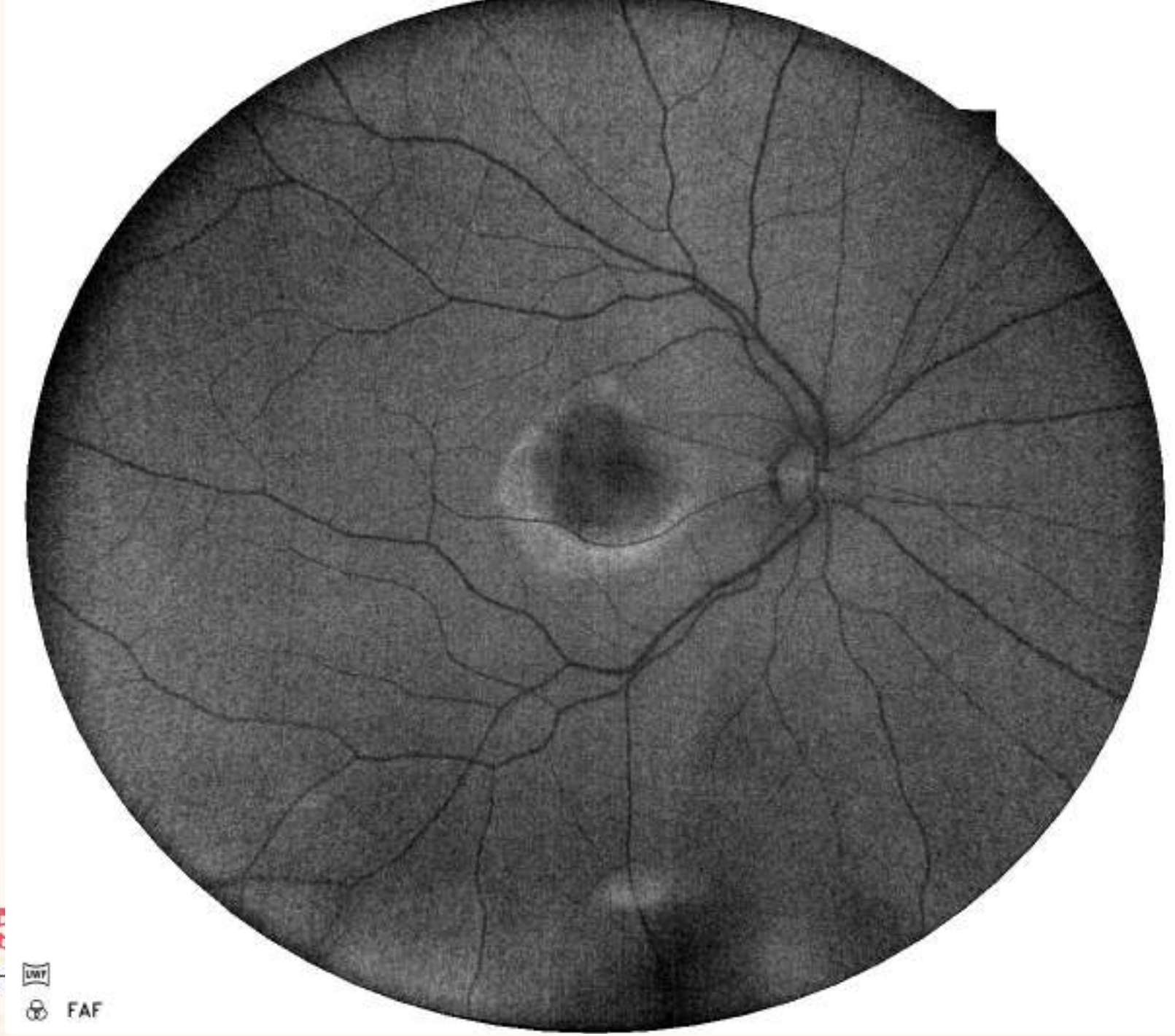
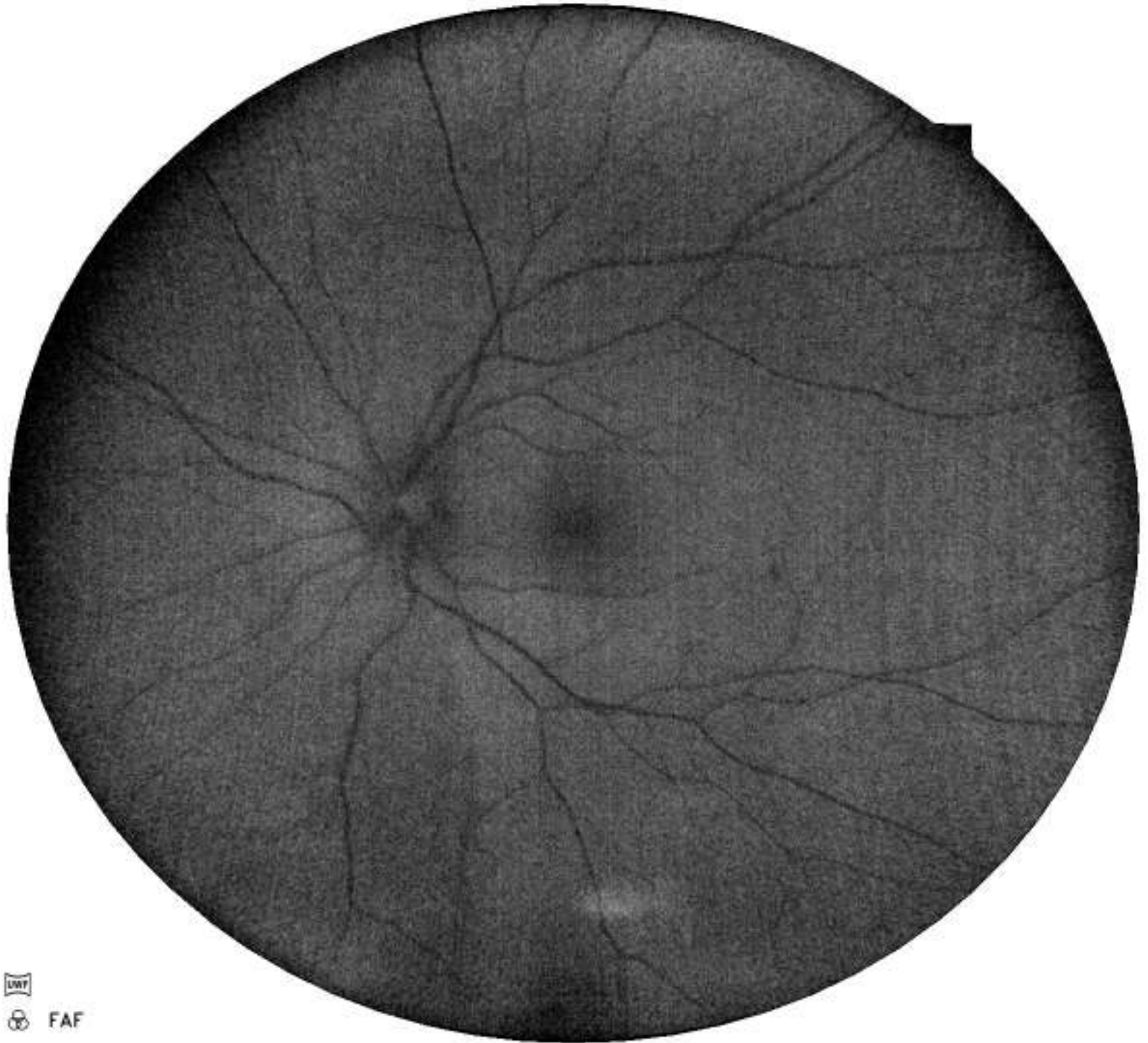


OS



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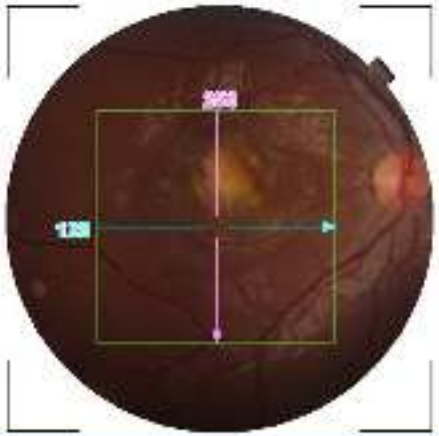
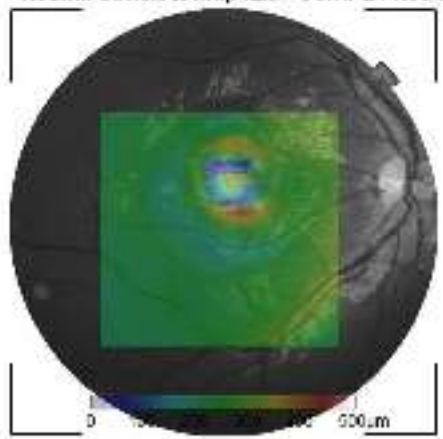
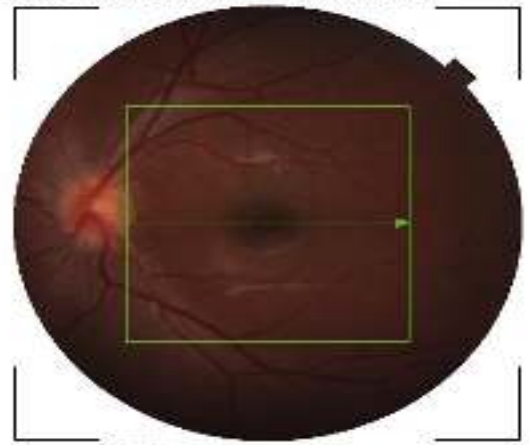
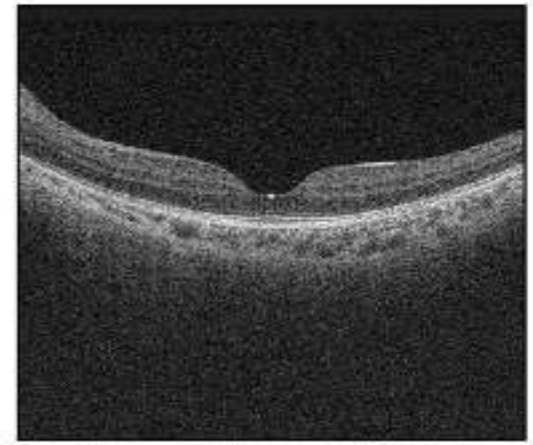


Retinal thickness map ILM - OS/RPE / Red-free



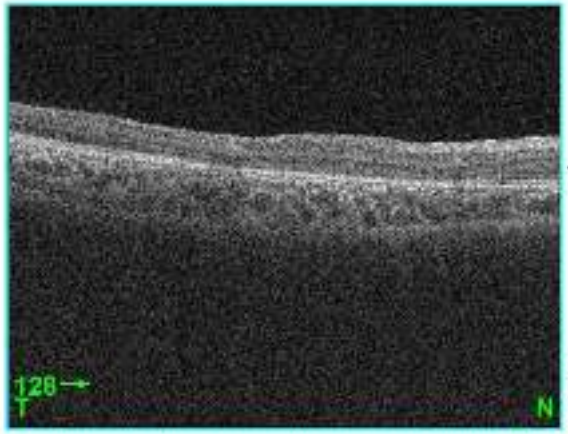




RNFL Thickness

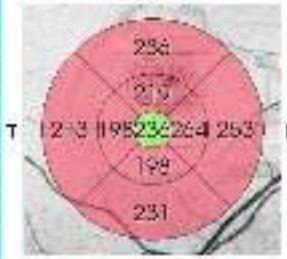
GCL+

GCL++

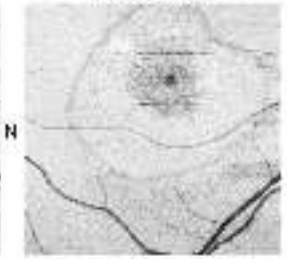


Retinal thickness ILM - OS/RPE(μm)

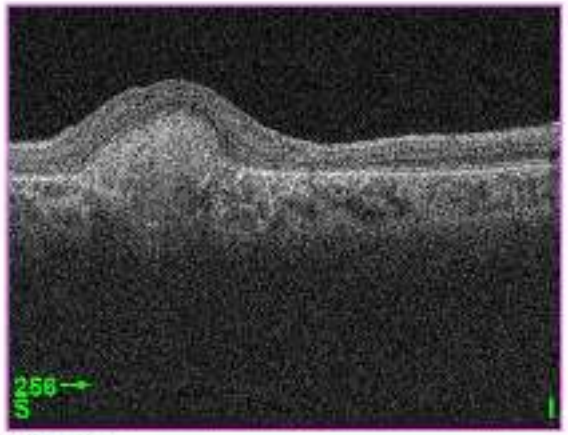
ETDRS



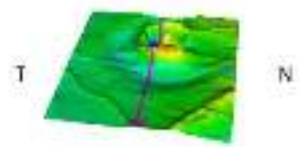
Shadowgram



Average Thickness	(μm)	230.5
Center Thickness	(μm)	237
Total Volume	(mm^3)	6.52



ILM - OS/RPE Map

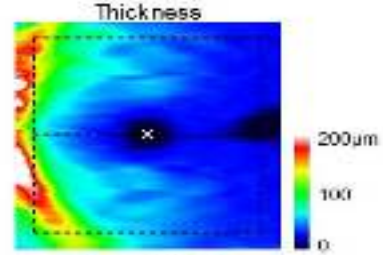


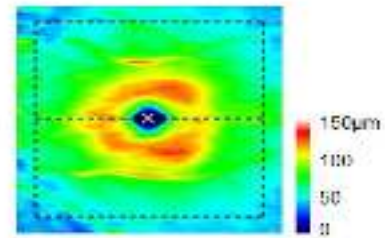
OS/RPE Surface

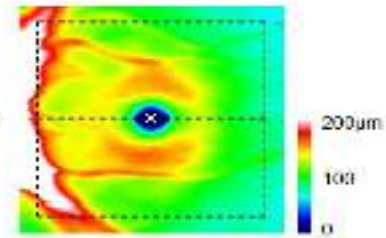
Comments :

Signature :

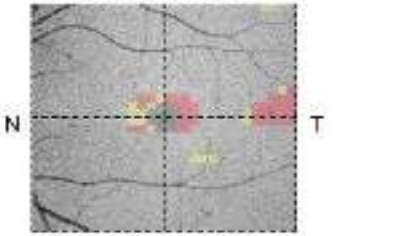
Date :

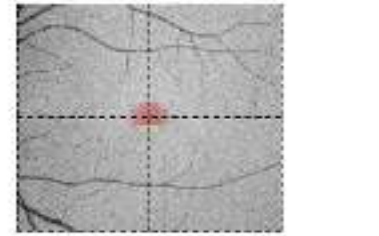


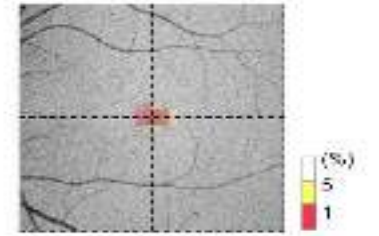




SuperPixel-200



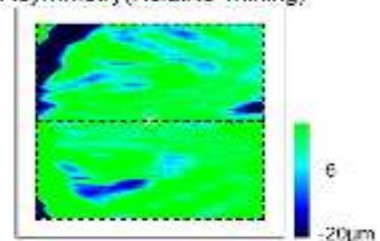


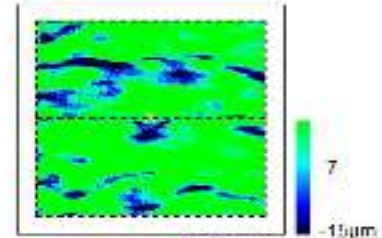


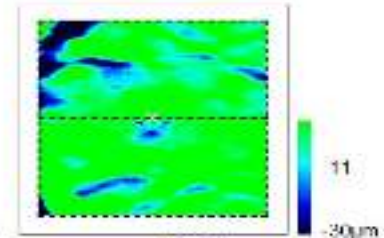
Average(6mm x 6mm)

Superior	46 um	Superior	74 um	Superior	120 um
Inferior	50 um	Inferior	75 um	Inferior	125 um
Total	48 um	Total	75 um	Total	123 um

Asymmetry(Relative Thining)

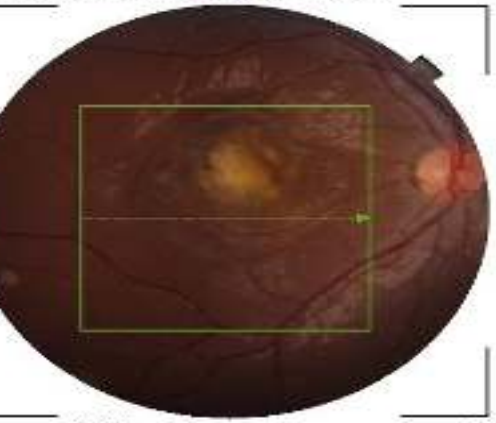


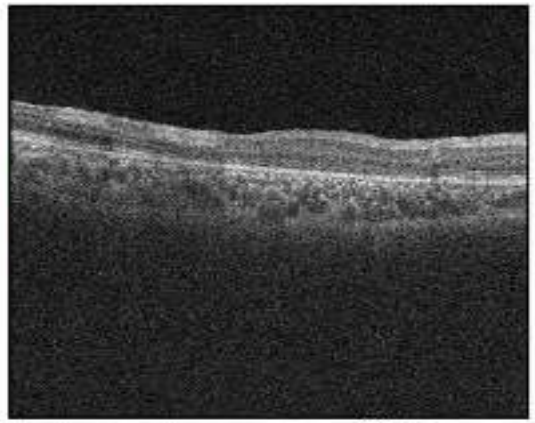




UL AZIZ ABDURABU
Gender : Female
DOB : 01-Jan-14
Fixation : OD(R) Macula
Age : 11
Scan : 3D(H)(7.0 x 7.0mm - 512 x 256)

Q Image Quality: 73 mode: Fine(2.0.7)
ture Date: 29-Jan-25 O.E.M:1.000x

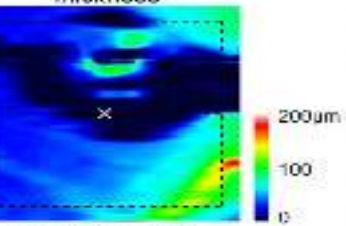


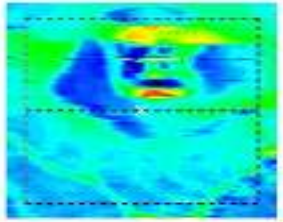


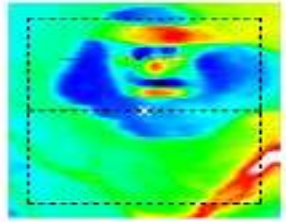
RNFL Thickness

GCL+

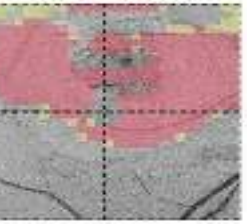
GCL++

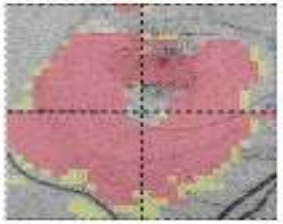


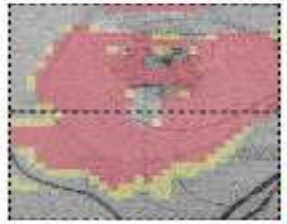




SuperPixel-200







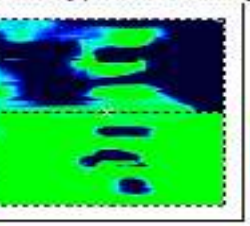
verage(6mm x 6mm)

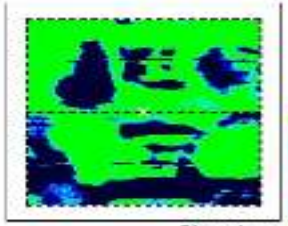
uperior	19 um
nferior	43 um
Total	31 um

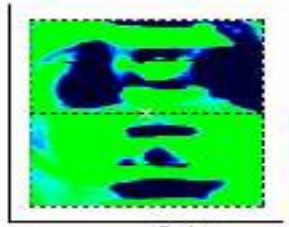
Superior	54 um
Inferior	46 um
Total	50 um

Superior	73 um
Inferior	90 um
Total	81 um

mmetry(Relative Thining)







NameJOURI ABDUL AZIZ ABDURABU
Gender : Female
DOB : 01-Jan-14
Fixation : OS(L) Macula
Age : 11
Scan : 3D(H)(7.0 x 7.0mm - 512 x 256)

OS(L)

TopQ Image Quality: 66 mode: Fine(2.0.7)
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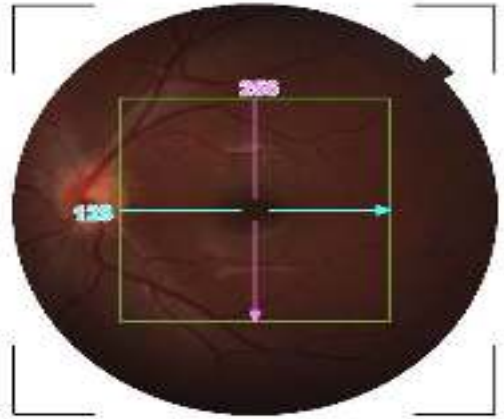
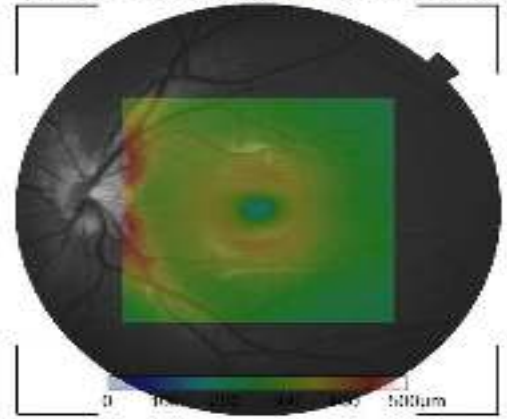
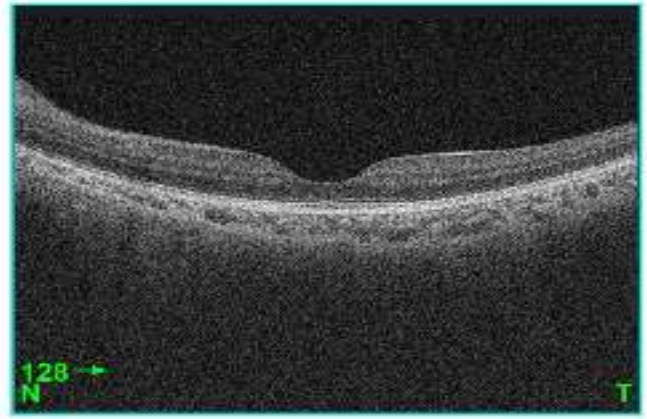


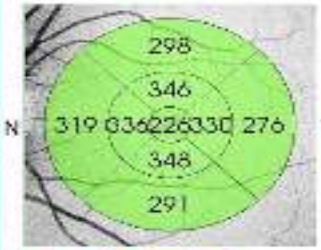
Retinal thickness map ILM - OS/RPE / Red-free



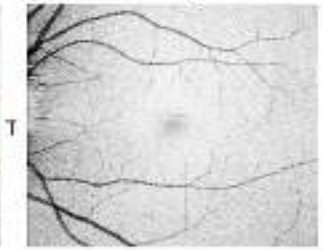
Retinal thickness ILM - OS/RPE(µm)



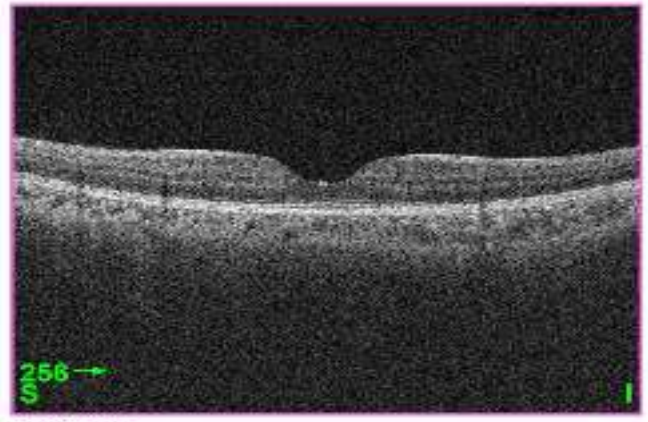
ETDRS



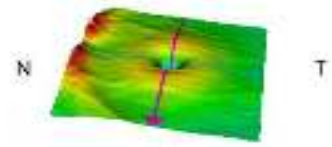
Shadowgram



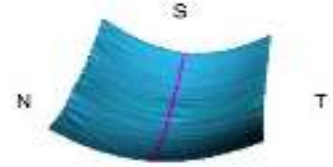
Average Thickness	(µm)	303.9
Center Thickness	(µm)	172
Total Volume	(mm^3)	8.59



ILM - OS/RPE Map



OS/RPE Surface



Comments :

Signature :

Date :

WORK UP

- ERG /EOG
- B-SCAN
- BLOOD TEST FOR TOXOPLASMOSIS & TOXOCARIASIS
- UVEITIS CONSULTATION
- RETINA CONSULTATION
- DX INACTIVE IDIOPATHIC CNVM OD

INTRODUCTION

- Choroidal neovascularization (CNV) is a common pathologic lesion that occurs in various chorioretinopathy In adults . it is the most common cause of visual loss in age-related macular degeneration, followed by CNVM secondary to pathologic myopia ,
- It is the leading cause of blindness in Europe and North America in the adult population

INTRODUCTION

- In children, the incidence of CNV is quite rare , however, its impact in view of the number of blind years lived is tremendous and the lesion have a severe impact on visual acuity and quality of life.

- Furthermore, young children may have no symptoms or complaints , thus hindering early diagnosis

- Several but very few studies have been published on this topic and is mainly in the Retina journals ,the last study was by sandra hoyek and associates (AJO ,Am Ophthalmology 2024;261: 76–84.)

- Pediatric ophthalmologist awareness about this disease is very limited as they will be eventually managed by the medical retina.

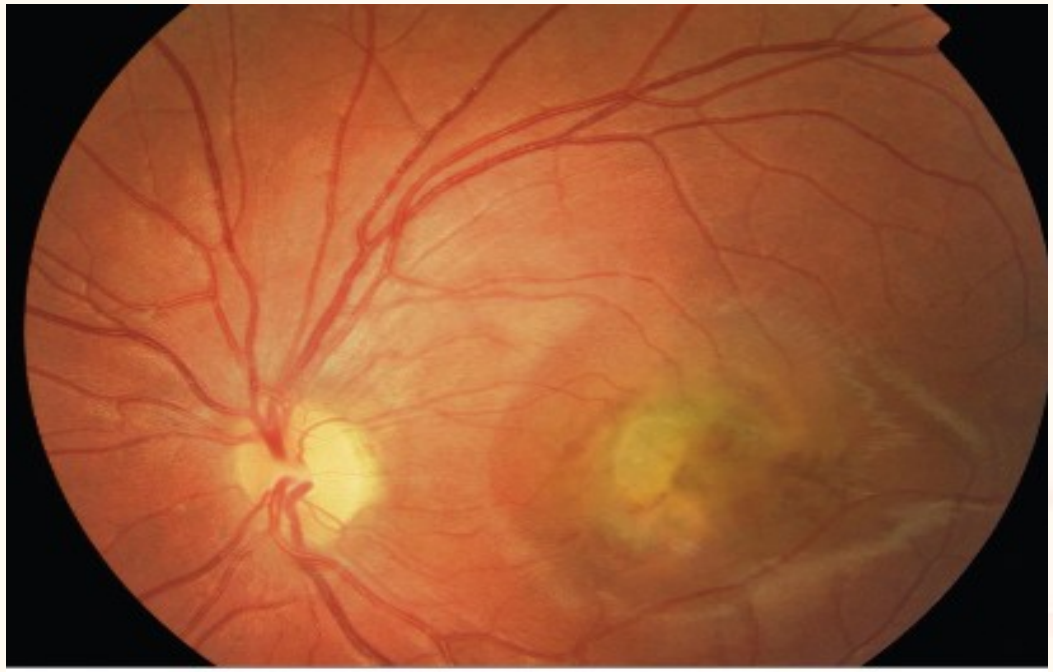
- The prevalence of CNV in children is not really known and varies from one region of the world to another.

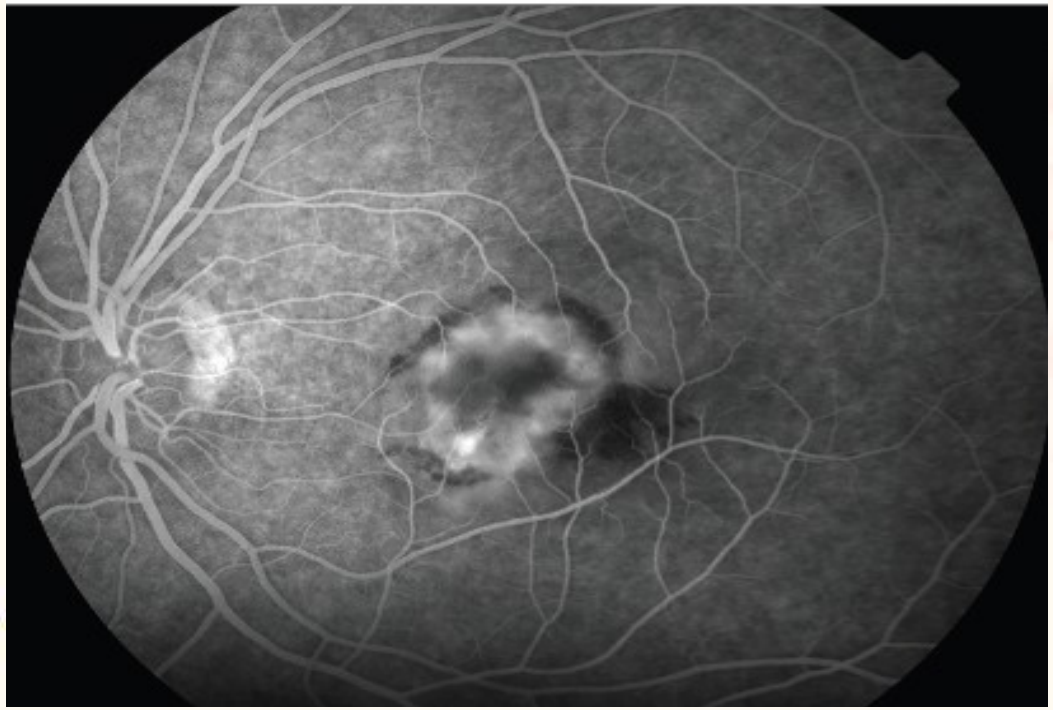
- In several studies there was more female preponderance

DEFINITION

- Choroidal neovascularization (CNV) is characterized by the growth of new blood vessels that originate from the choroid through a break in Bruch's membrane into the sub-retinal pigment epithelium (sub-RPE) or subretinal space.

- The neovascular membrane usually occurs at the macula or at the margin of the optic disc and often leaks blood and fluid, resulting ultimately in photoreceptor cell death.





ETIOLOGY

- A case series that includes 33 eyes of 30 patients in the Chinese population was reported by Ting Zhang and associated

The most important etiological factors in his group was:

- 1-congenital/developmental anomalies(16%)of cases
- 2- inflammatory retina-choroidopathy (2% of cases in JRA)
- 3- Best vitelliform macular dystrophy (bilateral)
- 4-High myopia
- 5-optic nerve head drusen, optic disc hamartoma, morning glory disc anomaly.
- 6- retinitis pigmentosa.

- Idiopathic (most common) in Sandra study 48%

- Inflammation (Unclassified choroiditis post inflammatory , Unclassified choroiditis, Serpiginous choroiditis. Vogt–Koyanagi–Harada disease ,toxoplasmosis, congenital rubella , toxocariasis ,sarcoidosis, Presumed ocular histoplasmosis syndrome, Viral retinopathy) **Sandra study 20%**

- Myopia CNVM can occur at any age, although it is rare in the pediatric age group. This may be because predisposing factors such as RPE atrophy and lacquer cracks take time to develop

- Retinal dystrophies, including ,Bests disease Stargardt disease, choroideremia, North Carolina macular dystrophy, and other macular dystrophies

- Optic N head anomalies(drusen, chronic papilledema, pseudo papilledema, idiopathic intra -crânial hypertension, malignant hypertension,idiopathic &congenital anomalies (coloboma ,optic n pit ,morning glory)
- Post trauma with choroidal rupture (esp. if the rupture is close to fovea)
- fewer rare causes: Choroidal osteoma, Combined hamartoma

DIFFERENCE BETWEEN ADULT AND PEDIATRIC CNV

- 1- Rarity of macular degeneration and myopic fundus changes
- 2-lack of calcification and thickening of Bruch's membrane & absence of basal laminar deposits.
- 3- single blood vessel growth site, whereas age-related macular degeneration related CNVM often have multiple such sites.
- 4- presence of solitary subretinal in-growth sites unlike adult cases in which multiple in-growth sites are common
- 5-Usually, unilateral in 80% of cases in most studies
- 6- Most of the CNVs were found to be classic on FFA and type 2 on OCT and had a sub- foveal location.
- 7- spontaneous regression of CNV in children making surgical excision technically complete and recurrences less likely.

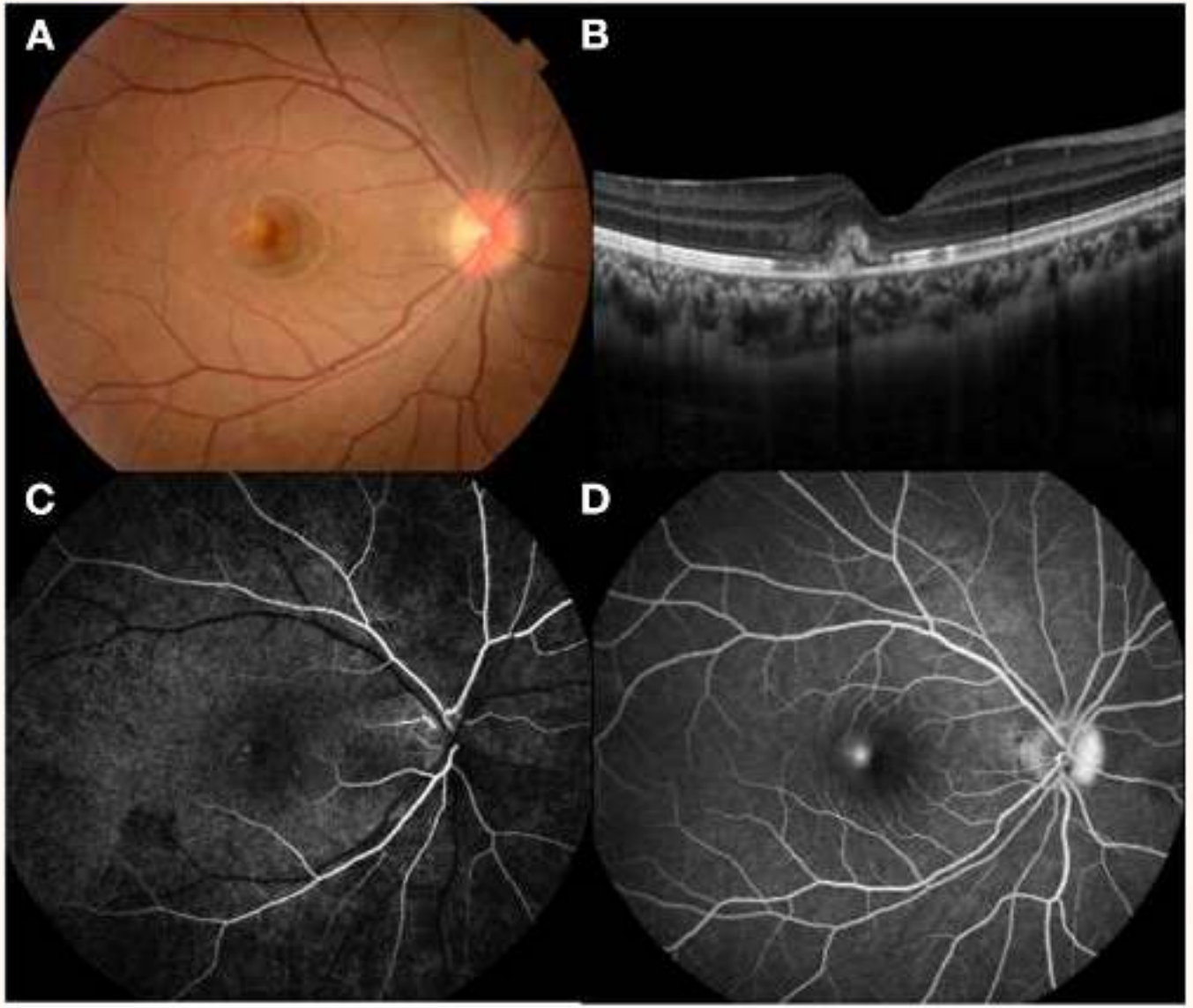
HISTOPATHOLOGY

- On histopathologic examination, the most common components of pediatric CNVM are retinal pigment epithelium (RPE), fibrocytes, vascular endothelium, and collagen.

- in one case of idiopathic CNVM in a 21-month-old boy that was removed surgically, there were no inflammatory cells (inflammatory cells are often seen in CNVM removed from adults).

DIAGNOSIS

- Fundus examination
- FFA remains the gold standard but is technically difficult in children
- OCTA is becoming the preferred alternative test as itis noninvasive diagnostic technique without risks and resources associated with FFA dye use.it provides high-resolution, depth-resolved images and may be particularly beneficial in the diagnosis and monitoring and follow –up of children with CNVM
- OCTA (study by saaly ong and associates from USA/Graefe's Arch Clin Exp Ophthalmology. 2020) and (Sandra Hoyek study 2024)This study was a 2-center retrospective consecutive case series of pediatric patients with CNVM at Massachusetts Eye and Ear and Boston Children's Hospital from 2005 to 2022 they reported data of 30 eyes of 25 pediatric patients).using the OCTA

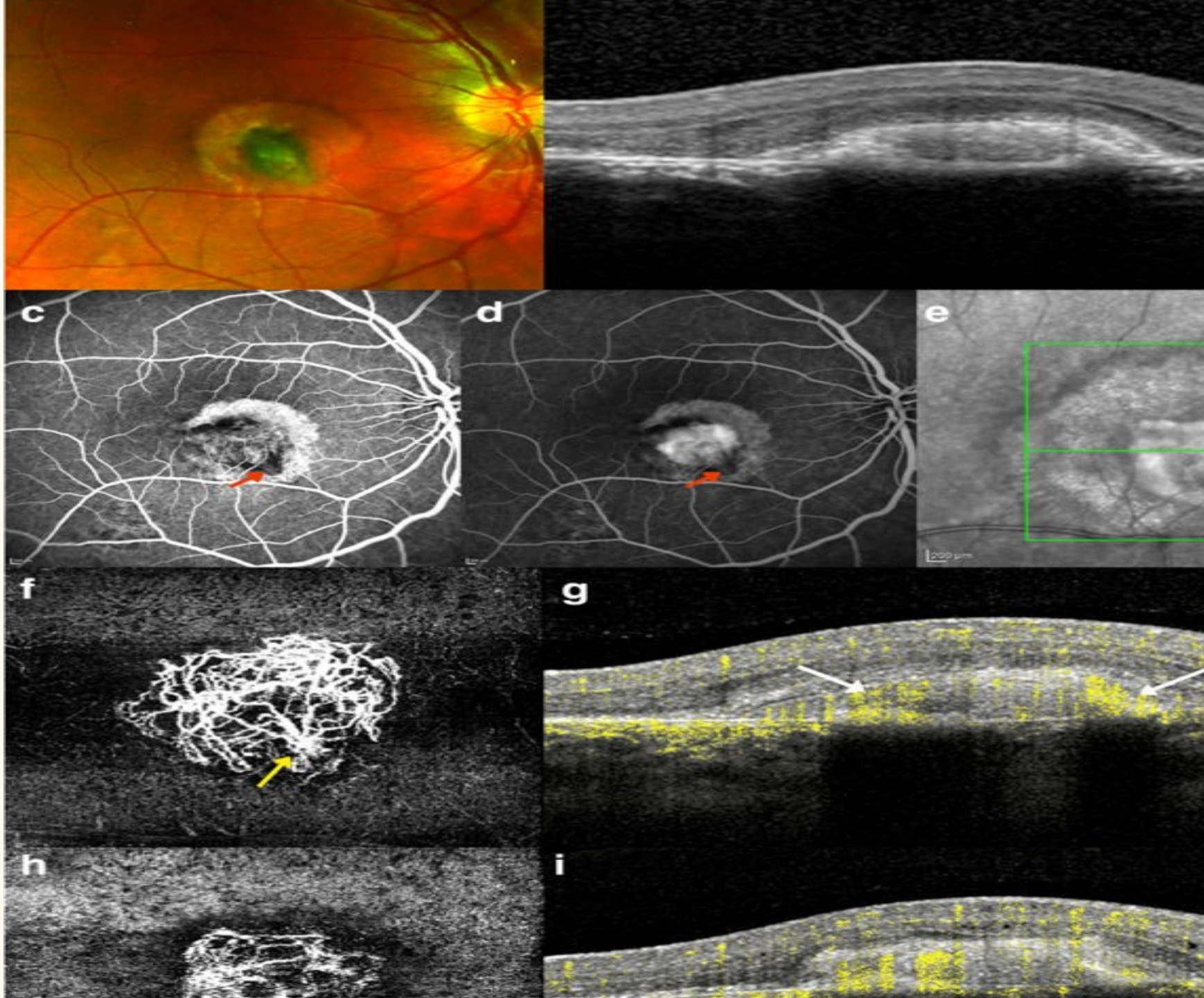


Multimodal imaging of a 14-year-old female with idiopathic choroidal neovascularization. (A) A parafoveal yellowish lesion was noted on fundoscopy. (B) OCT showed a hyperreflective area located in subretinal space with surrounding serous retinal detachment. (C,D) Hyper fluorescence in the early phase and leakage in the late phase FFA revealed an active lesion parafoveally.

DIFFERENCE BETWEEN ACTIVE AND INACTIVE MEMBRANE

- Hemorrhage is seen & numerous dense, fine capillaries with frequent anastomoses and vessel loops
- After treatment there will be lack of angiographic leakage and the OCTA demonstrated a loss of fine capillaries, anastomoses, and vessel loops when compared with imaging before treatment and resolution of subretinal hyperreflective material corresponding to hemorrhage and subretinal fluid

e 1 idiopathic CNV in patient 1. **a** At entation, color photo demonstrates a erpigmented CNV complex with norrhage just inferonasal to the foveal ter. **b** OCT shows the subretinal ment epithelium CNV complex. The ent was treated with intravitreal anti-GF injections. **c** One year after entation, the patient had a recurrence CNV activity. **c** Early and **d** late nes of fluorescein angiography strate occult leakage of the CNV plex, blocked fluorescence from norrhage, and circumferential window ect from retinal pigment epithelium phy. A feeder vessel is also visible arrows). **e** The green box and green w in the infrared image depict the captured by the en-face (**f**) and cross-ional (**g**) OCTA, respectively. **f** En-OCTA shows a large circular CNV plex with a main trunk (yellow w), multiple dense thin capillaries aching from the main trunk in a tree-manner, and frequent anastomoses in



DIAGNOSIS

- OCTA is a non-invasive device that can demonstrate differences in morphological characteristics between active and quiescent CNV.

- Therefore, it can be a useful tool to monitor intra-individual changes in CNV morphology across time. This can be especially helpful in the early detection of CNV recurrence in children .

TREATMENT

- The decision whether to treat CNV in children is particularly difficult because of the late presentation, paucity of natural history data, and lack of clinical trials in children.

The four main options

- 1- Observation(spontaneous resolution have been reported but most studies showed worst VA if untreated)
- 2- laser photocoagulation,(in a case series of 23 patients Wise et noted a rapid progression of the CNV towards the macula and recommended early photocoagulation.
- 3-submacular surgery (CNV in children usually develops from a solitary site and is subretinal rather than sub-RPE). This result in a favorable outcome with surgical management.
- 4-photodynamic therapy (PDT) with verteporfin.(few studies in the literature with small n of patients)

TREATMENT MODALITIES

- Recent therapeutic paradigm is shifting toward the use of anti-vascular endothelial growth factor (VEGF) Several studies have reported the usefulness of anti-VEGF in pediatric CNV, but most are retrospective case series reports (the last was in Korea by Soon and associates and was published in the journal of retina 2023)
- In Sandra study the most common first-line treatment was intravitreal injection of an anti-VEGF drug (retreatment rate was 52.2%)There was an overall positive response reported in many studies (no more than two injections were required)
- CNV in children probably requires fewer anti-VEGF injections for stabilization than does CNV in adults. this may be due to the health of the RPE pump in young people

TREATMENT OUTCOMES

- Different clinical outcomes could be identified by subtype.

- tumor-associated CNV had poor outcomes, whereas CNV associated with inflammation, idiopathic and trauma had positive outcomes.(choi study)

- in another study by Kozak , CNV secondary to uveitis and idiopathic Causes showed good visual response, but CNV secondary to dystrophies did not show significant improvement after anti-VEGF treatment.

VEGEF USE IN PEDIATRICS

- A concern about using anti-VEGEF agents in children is lack of data about drug metabolism; this may be different in children from adults, and adverse events may occur that are not predictable from the adult experience.
- The average number of injections is different in different studies but is 2-3 injections and is generally less than adult injections .
- Observation of CNVs in children might be a reasonable option; but so far it is difficult to assess which CNV would regress or progress.
- In one non-controlled study, visual outcomes in eyes with successfully treated sub foveal CNVM was noted to be better than in eyes with spontaneously regressed sub foveal CNVM, hence the probable importance of early diagnosis and treatment of pediatric CNVM.
- studies have reported that children are able to maintain gains in vision for 2 years (Sandra study shows the durability of the treatment up to 13 years)

CONCLUSION

- CNVM in pediatric patients are most often idiopathic and unilateral.
- CNVM in the pediatric population differed from that in the adult population according to etiology, angiographic characteristics, and treatment response.
- OCTA is a useful adjunct that may replace dye-based angiography to monitor regression and guide management as it allows early visualization of CNVM, detection of its response to treatment and recurrence
- Intravitreal injection of an anti-VEGF leads to visual improvement, chronic treatment is rarely required.

CONCLUSION

Awareness of the pediatric ophthalmologist is warranted as the patient might present first to them rather than to retina

- Early intervention and treatment might save the patient vision especially if treated before the amblyopia age group
- The need for multicenter case studies is indicated to look at the treatment results all over the world .

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