

# Multimodal imaging of torpedo maculopathy in a Chinese woman: a case report

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## Case report

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# Abstract

**Background:** Torpedo maculopathy is a rare, benign, and congenital macular lesion that typically appears in 'torpedo-shape' and located at the temporal macula region. The purpose of this article is to describe in detailed regarding the torpedo maculopathy in a Chinese woman using multimodal imaging. **Case presentation:** A 30-year-old Chinese woman occasionally had an yellowish-white macular lesion in her right eye during a routine examination. She had no symptoms, and the best-corrected visual acuity of both eyes was 6/6. Funduscopic examination revealed a torpedo-shaped and mild hyperpigmentation lesion in the temporal macular area of her right eye. Infrared fundal (IR) photograph showed that the lesion contour was visible, transverse elliptical, a tip pointed towards the central fovea of the macula. Microperimetry visual field appeared normal. The spectral-domain optical coherence tomography (SD-OCT) showed a normal inner, mildly thinned retinal pigment epithelium (RPE), and increased choroidal signal. OCT angiography (OCTA) choroid capillary segment revealed increased density of the choroidal vasculature. With fundus autofluorescence (FAF), the lesion showed slight hypoautofluorescence, and marginal mild hypoautofluorescence. Fundus fluorescence angiography (FFA) of the lesion showed variegated fluorescence and no leakage and change in the morphology during the whole imaging process. **Conclusions:** This is the first report to put forward a thorough and detailed description of torpedo maculopathy simultaneously using fundal photograph, IR, microperimetry visual field, OCT, OCTA, FAF, and FFA. Multimodal imaging provides precious and detailed information to further clarify the characteristics and development of this rare disease.

## Background

Torpedo maculopathy is a rare, benign, and congenital retinal pigment epithelium (RPE) disease, which typically appears as a 'torpedo-shaped' lesion at the temporal macula. In 1992, it was first reported by Roseman and Gass [1] as an asymptomatic, solitary hypopigmented nevus of the retinal pigment epithelium. In the classic fundus, it manifests as a solitary hypo-pigmented lesion that is oval in shape, resembling a 'bullet' or 'torpedo,' with a wedge-shaped tail extending outward and pointing towards the foveola along the horizontal raphe [2]. In 1993, torpedo maculopathy was named by Daily [3] due to its typical appearance. The typical fundus can be distinguished from other lesions, such as toxoplasma scar, traumatic injury, congenital hypertrophy of the RPE (CHRPE), and congenital RPE hypertrophia associated with Gardner syndrome, allowing for diagnosis.

However, the etiology of torpedo maculopathy still remains unknown. Many imaging devices have been used to observe the lesion. We herein used multimodal imaging for describing torpedo maculopathy in a Chinese woman. To our knowledge, this is the first report in the world to observe the torpedo maculopathy simultaneously using fundal photographs, IR, microperimetry visual field, OCT, OCTA, FAF, and FFA. We hypothesized that this case might be in a very early stage or a mild type of torpedo maculopathy.

## Case Presentation

A 30-year-old Chinese woman had a yellowish-white macular lesion in her right eye during a routine examination. She had no other symptoms. There was no pain or vision loss in her right eye. The patient denied any traumatic history. Her past medical history and ophthalmic history were negative. The initial best-corrected visual acuity (BCVA) was 6/6 for both eyes. The cornea was clear, and the anterior segment was normal. Pupils were equal, round and reactive to light with no afferent pupillary defect. There was no cataract in both eyes. The initial intraocular pressure (IOP) was 14mmHg in the right eye and 13 mmHg in the left eye. Funduscopy examination of the left eye was unremarkable. In the right eye, a spindle-shaped yellowish-white and hypo-pigmented lesion, which was about 0.5 disc diameter vertically by 1 disc diameter horizontally, was located in the temporal macular area with a tip pointed towards the central fovea of the macula (Figure 1, A). IR photograph showed that the contour of the lesion was visible, and transverse elliptical and was consistent with the colorful fundus photograph (Figure 1, B). Microperimetry visual field was basically normal (Figure 2). The SD-OCT showed a normal inner retina, mildly thinned outer retina and RPE in the temporal macular area, with correspondingly increased choroidal reflectivity (Figure 3). Other OCT findings included disorganization of the outer retina, and significant atrophy of an intact ellipsoid zone. OCT angiography (OCTA) of choroid capillary segment revealed increased density of the choroidal vasculature, corresponding to the area of the lesion ((Figure4). With FAF, the lesion showed slight hypoautofluorescence with marginal mild hyperautofluorescence (Figure 5, A). FFA of the lesion showed variegated fluorescence and no leakage and change in the morphology during the whole imaging process (Figure 5, B,C,D). Based on these findings, the patient was confirmed to have torpedo maculopathy clearly.

The treatment of this patient was closely observed. The patient was followed-up every 3 to 6 months. Her BCVA was still 6/6 in both eyes, and the fundus lesion remained intact during 15 months followed-up.

## Discussion And Conclusions

Torpedo maculopathy is a rare, benign, and congenital maculopathy, which is usually asymptomatic, but occasionally found during routine examination. It often occurs in patients without any relevant medical history and commonly unilateral, although bilateral cases have been reported previously[4]. The torpedo maculopathy appears as a transverse oval, yellowish-white hypo-pigmented lesion, and is located in the temporal macular area, with a tip pointed towards the central fovea.

Till now, the etiology of the disease is still unclear. Few studies have been reported to explain the pathogenesis of the lesion. Pian et al[5] assumed that it might be a developmental defect within the nerve-fiber layer at the horizontal raphe. Shields[6] suggested a persistent defect in the development of RPE during the fetal temporal bulge might be the reason for the cause of the lesion. Golchet et al[7] hypothesized that the lesion may be related to dysmorphia of the emissary canal of the long posterior ciliary artery and nerve.

Microperimetry can precisely reveal the correlation of retinal sensitivity and fundus lesion, and also detect microscotoma within the central visual field that may not be detectable using standard perimetry

methods[8]. Published reports demonstrated that scotoma is frequently associated with torpedo lesion[9-11]. Focal RPE atrophy results showed that reduced metabolites and oxygen supply for the inner retina, secondary choriocapillaris loss and photoreceptor degeneration are associated with the reduction of retinal sensitivity[12]. This case was different from the earlier reports, and there was no microscotoma seen, the retinal sensitivity appeared normal, and the patient had good visual function. Therefore, we speculated that the degree of RPE atrophy was very light in this case, and does not harm the function of overlying photoreceptors.

Using SD-OCT, the outer retina was disorganized, and significant atrophy of RPE with an intact ellipsoid zone was seen in this case. We thought that RPR still preserved the function of ingesting photoreceptor cell outer segment, so the ellipsoid zone still remained intact and the patient preserved the normal visual function. Evan et al[13] identified two patterns of abnormality: type 1, attenuation of outer retinal structures without outer retinal cavitation; and type 2, those with both attenuation of outer retinal structures and outer retinal cavitation. According to their theory, this patient was included under type 1 torpedo maculopathy. Besides, they also observed that the patients with type 1 lesion (age, 4–37) tended to be younger than those with type 2 lesion (age, 13–73) [13]. This patient was 30, and was consistent with the characteristics summarized by them.

OCTA non-invasively detected the movement of red blood cells to reveal the retinal and choroidal vascular system. Therefore, alterations in the choriocapillaris can be visualized using OCTA. Papastefanou and his colleagues[14] described the OCTA features of torpedo lesions with OCTA, and revealed choroidal vascular segmentation with hypo-reflectivity (atrophy), which was correlated to the OCT of the subretinal cleft. While there was no subretinal cleft in our patient, and the OCTA findings were different. OCTA choroid capillary segment revealed increased density of choroidal vasculature, revealing thinner RPE as the optical signal transmission for the increased choroidal thickness. Comparison of our case with the previous reported cases[14], our case could be considered as an early stage according to the OCT classification.

Autofluorescence signal is predominantly derived from lipofuscin within the RPE[15]. In our case, FAF showed slight hypoautofluorescence with marginal mild hyperautofluorescence. The possible explanation for this was that the hypoautofluorescence of thinner RPE within the lesion provides a sharp contrast to the physiological hyper-autofluorescence within the marginal region.

FFA of the lesion showed variegated fluorescence and no leakage and change in the morphology during the whole imaging process. This demonstrated no choroidal neovascularization (CNV), while few cases showed the existence of CNV[16]. Lesions in our case were still limited to RPE without CNV.

In conclusion, we herein presented a case of torpedo maculopathy using fundal photograph, IR, microperimetry visual field, OCT, OCTA, FAF, and FFA simultaneously for the first time. We demonstrated it to be present in a very early stage or a mild type of torpedo maculopathy. The natural development of torpedo maculopathy is still unclear, whether it does not develop or develops very slowly. Multimodal imaging provides precious and detailed information to easily diagnose and deeply understand this rare

disease. This disease is very rare, and needs more case reports worldwide and longer follow-up to further understand regarding the etiology, characteristics and development of this lesion.

## **Abbreviations**

BCVA: best-corrected visual acuity; CNV: choroidal neovascularization; CHRPE: congenital hypertrophy of the RPE; FAF: fundus autofluorescence; FFA: Fundus fluorescence angiography; IOP: initial intraocular pressure; IR: Infrared fundal; OCTA: OCT angiographic; RPE: retinal pigment epithelium; SD-OCT: spectral-domain optical coherence tomographic

## **Declarations**

### **Funding**

Not applicable.

### **Availability of data and materials**

All data and materials of the case are included in this published article.

### **Authors' contributions**

Dr. Ding collected the data of the patient, consulted literatures and wrote the manuscript; Dr. Yao edited the manuscript, and dealt with the figures. Dr. Ye also gave useful suggestions on writing and improved the manuscript. Dr. Yu was the Consultant in charge for the case, established the diagnosis and approved the submitted version. All authors read and approved the final manuscript.

### **Acknowledgement**

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### **Ethics approval and consent to participate**

Not applicable.

### **Consent for publication**

Informed written consent had been obtained from the patient.

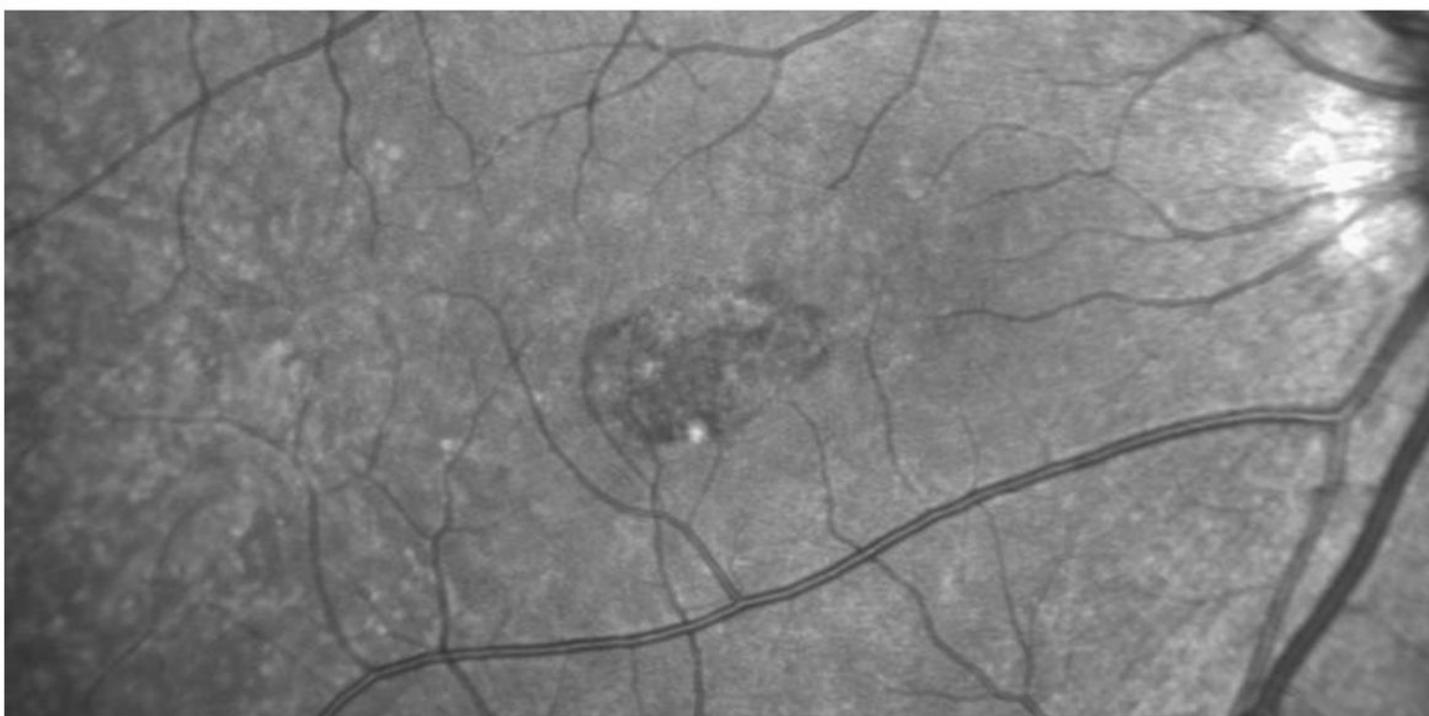
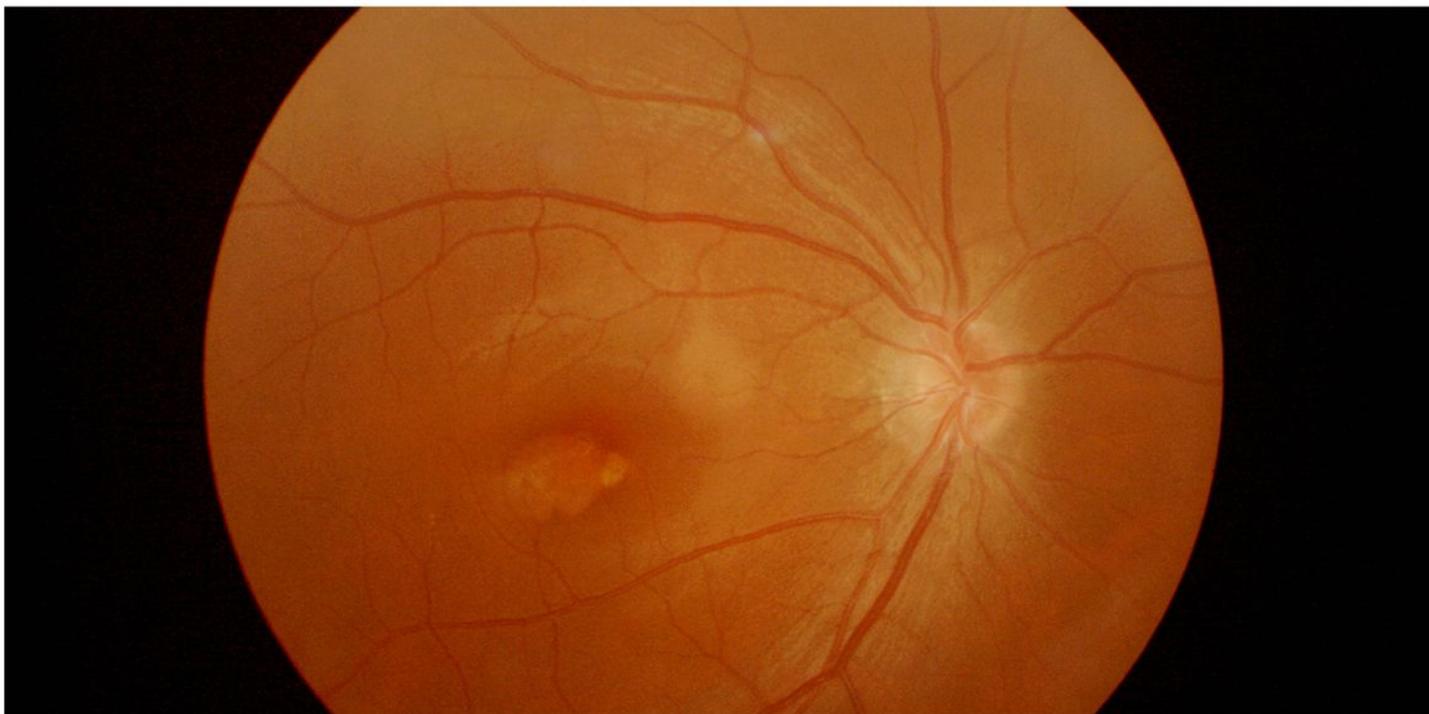
### **Competing interests**

The authors declare that they have no competing interests.

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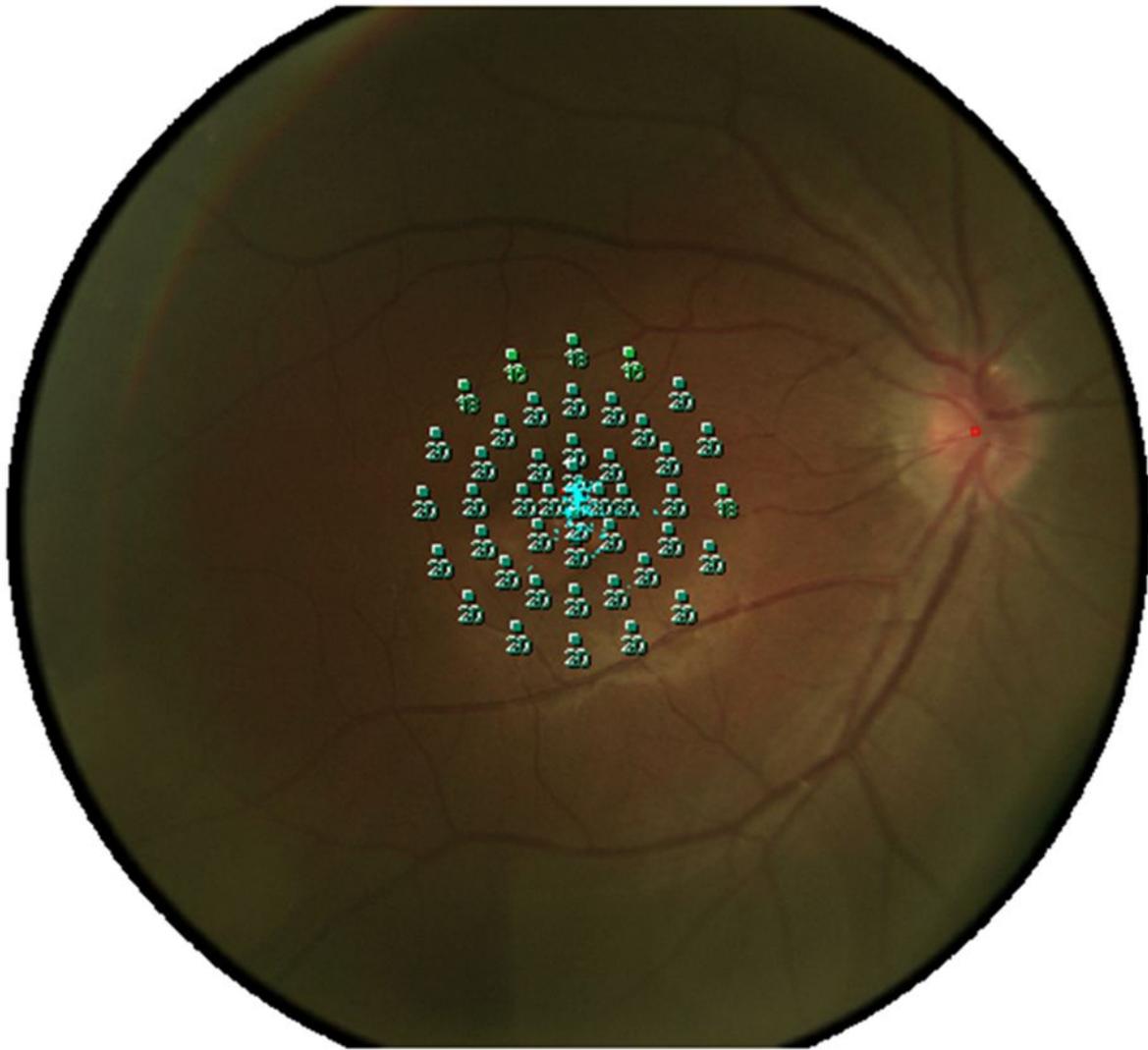
## Figures



## Figure 1

In the right eye, a spindle-shaped yellowish-white and hypo-pigmented lesion, which was about 0.5 disc diameter vertically by 1 disc diameter horizontally, was located in the temporal macular area with a tip pointed towards the central fovea of the macula (Figure 1, A). IR photograph showed that the contour of the lesion was visible, and transverse elliptical and was consistent with the colorful fundus photograph (Figure 1, B).

Note:



Test	Background	Fixation target	Reliability test	Pattern	Thresh. strategy	Stimulus	
Test 1	White	Single cross, 1	(0/3)	macula 12° 0dB	4-2	Goldmann (R00 ms)	○

**Legenda**

- Not seen at ... dB
- Seen at ... dB
- △ Not projected

Attenuation scale (dB)

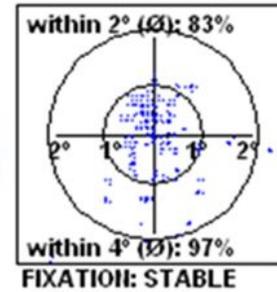
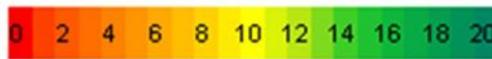
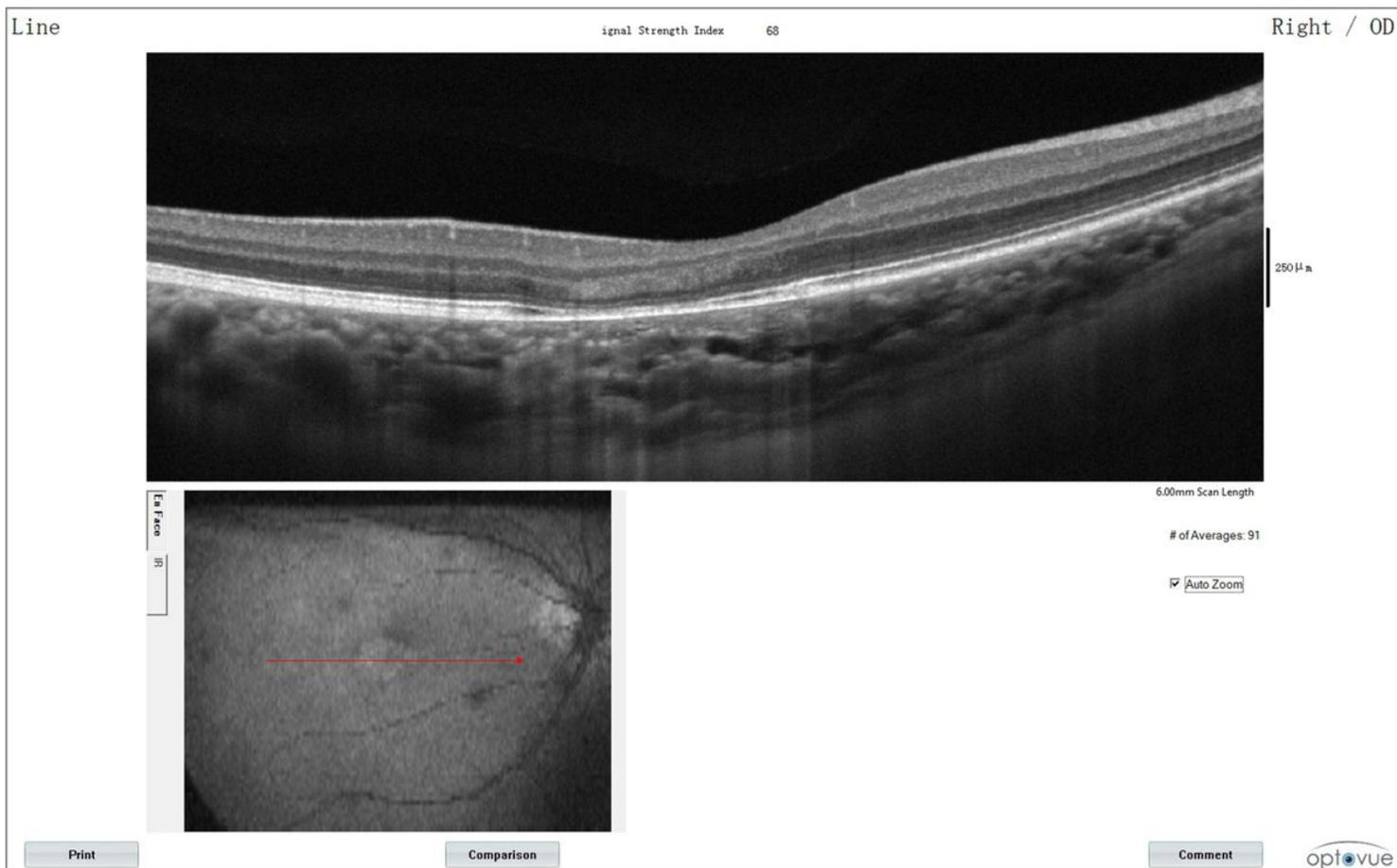


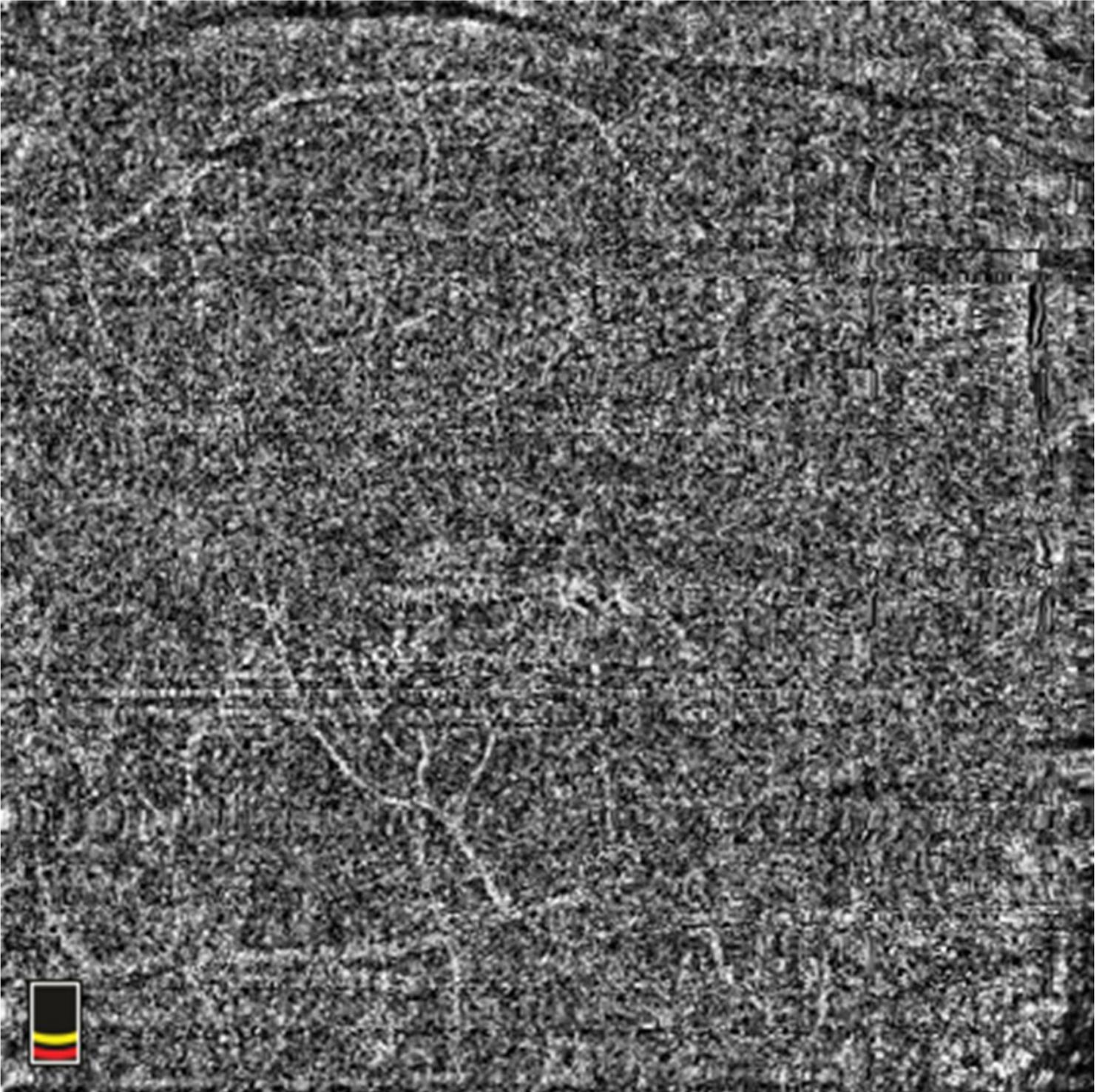
Figure 2

Microperimetry visual field was basically normal.



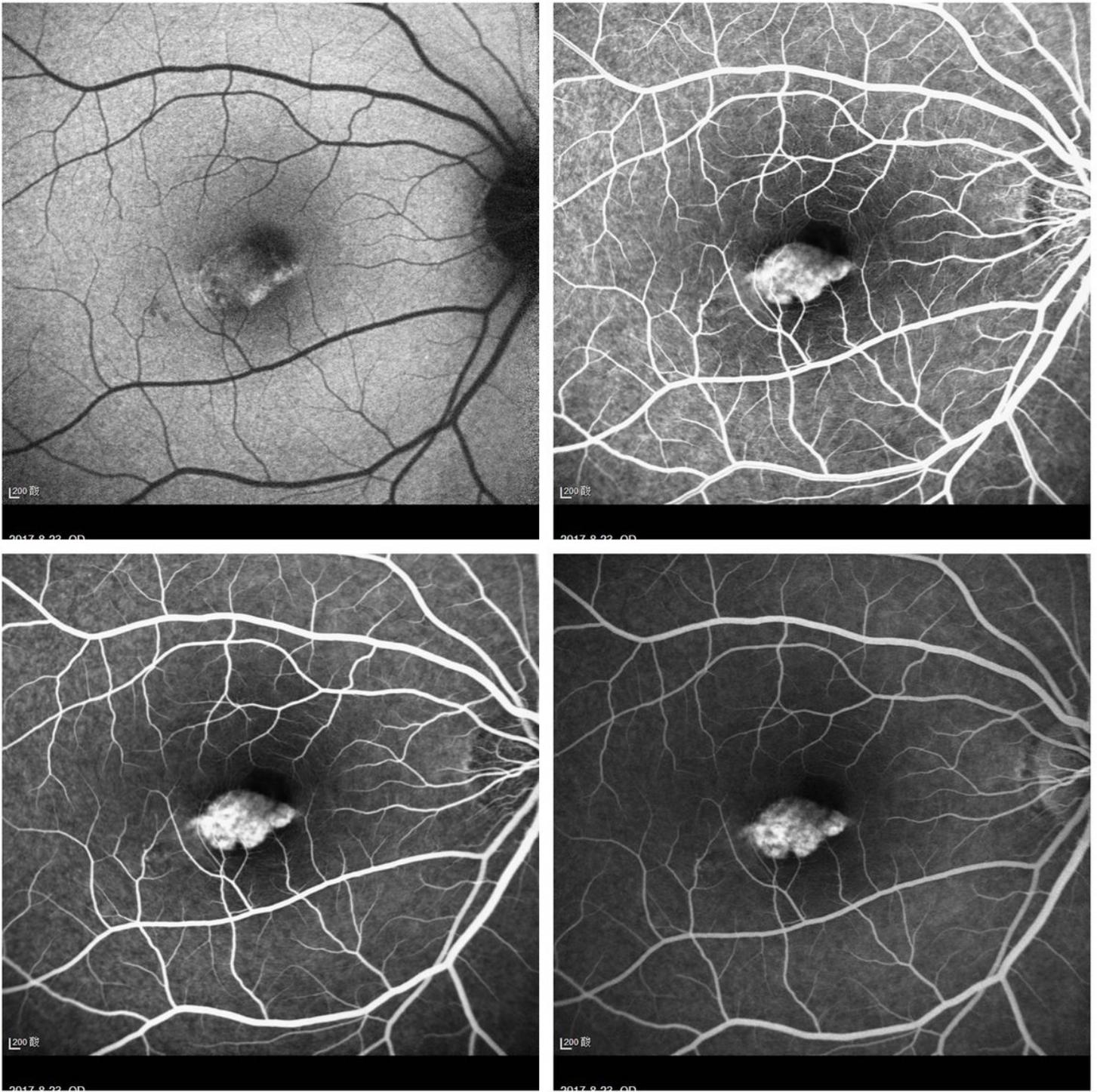
**Figure 3**

The SD-OCT showed a normal inner retina, mildly thinned outer retina and RPE in the temporal macular area, with correspondingly increased choroidal reflectivity.



**Figure 4**

OCT angiography (OCTA) of choroid capillary segment revealed increased density of the choroidal vasculature, corresponding to the area of the lesion.



**Figure 5**

With FAF, the lesion showed slight hypoautofluorescence with marginal mild hyperautofluorescence (Figure 5, A). FFA of the lesion showed variegated fluorescence and no leakage and change in the morphology during the whole imaging process (Figure 5, B,C,D).