CASE REPORT

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A Case Series of Variations in Visual Field Defects in Chiasmal Syndrome: Insight from Sellar Tumor-Associated Cases

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ABSTRACT

Introduction: Visual field examination is important in evaluating patients with visual loss, intended to assess defects and locate lesions that occur along afferent visual pathways. The optic chiasm is an important structure in Neuro-ophthalmology because of the arrangement of visual pathways and visual field defects caused by various pathological conditions such as compression, inflammation, demyelination, ischemia, and infiltration. This case series aimed to report the variation of visual field defects that can occur in patients with chiasmal syndrome.

Case Report: First cases, 54 years 54-year-old male presenting with blurry vision in both eyes for two months before, there were missing letters when reading. Presenting Humphrey Visual Field (HVF) 30-2 examination shows bitemporal hemianopia. Second case, 49 years 49-year-old female presenting with vision loss in the left eye for three months before with HVF examination showing temporal hemianopia in the left eye and unspecified defect on the right eye. The third case, 40 years 40-year-old, presented with vision loss in the left eye for two months. The HVF shows temporal hemianopia in the right eye and is difficult to evaluate due to visual loss in the left eye. The last case, 62 years 62-year-old male, presented with blurry vision in the left eye for two months before. Presenting HVF examination shows the right homonymous hemianopia defect.

Conclusion: There are many variations of visual field defects in patients with chiasmal syndrome. It is essential to know the anatomical of the optic chiasm and the technique of visual field examination.

Keywords: Chiasmal Syndrome, Optic Chiasm, Bitemporal hemianopia, Visual Field defect.
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INTRODUCTION

A visual field examination is one of the neuro-ophthalmology examinations that is important in evaluating patients with decreased central and peripheral visual acuity that can occur due to various medical conditions such as glaucoma, stroke, intracranial tumors, or other neurological deficits. Visual field examination is used to localize the lesion along the visual afferent pathway, determine the pattern of the visual field defect, and determine the extension of the visual field defect.[1,2]

Optic chiasm is one of the essential parts of the afferent visual pathway. The arrangement of nerve fibers in the optic chiasm provides a specific visual field defect that can be caused by various pathological processes such as compression, inflammation, demyelination, ischemia, and infiltration. Bitemporal hemianopia is the most common type of visual field defect in the optic chiasm lesion. Other visual field defects may result from compression of adjacent structures in the optic chiasm.[1,2]

This case series reported four cases of sellar tumor-associated chiasmal syndrome with a variety of visual field defects.

CASE HISTORY

Case 1

A 54-year-old male patient came with blurry vision in both eyes for two months, described as being missing parts of letters, especially at the beginning and the end of sentences. The patient also said that he often hit objects around him when he was walking.

Visual acuity on the right eye is 6/18 with pinhole 6/12, and the left eye is 6/45 with pinhole 6/20. The anterior segment examination is within standard limits on both eyes. Funduscopic examination showed well well-defined, round Optic Nerve Head (ONH) with slightly pallor on the temporal side, flat retina, and good macular reflexes in both eyes. Ocular computed tomography (OCT) examination showed the mean retinal nerve fiber layer (RNFL) thickness in the right eye is 97 µm and 90 µm in the left eye. Humphrey Visual Field (HVF) 30-2 showed bitemporal hemianopia (Figure 1). CT-Scan examination showed there is a well-defined hypodense lesion with fluid density in the sellar region, contrast enhancement was found on the side of the lesion, with the lesion pressing the optic chiasm. The cavernous sinus was well visible around the lesion. Suggests a benign sellar subarachnoid cyst (Figure 2).

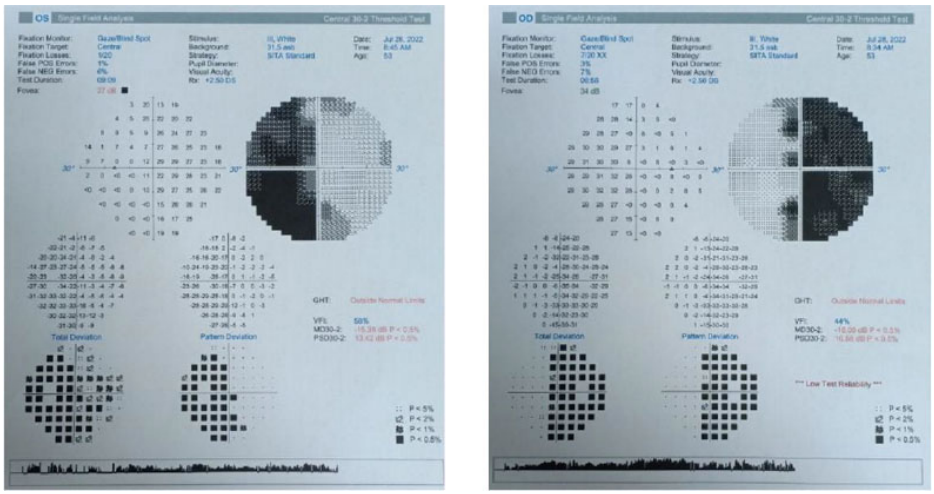

Figure 1. HVF 30-2 examination showed the bitemporal hemianopia.

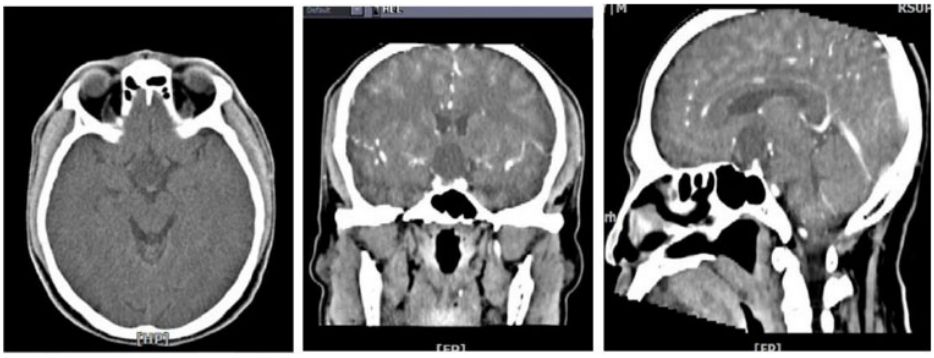

Figure 2. Hypodense lesion with fluid density in the sellar region compressing the optic chiasm, shown on CT Scan Examination, suggests a benign sellar subarachnoid cyst.

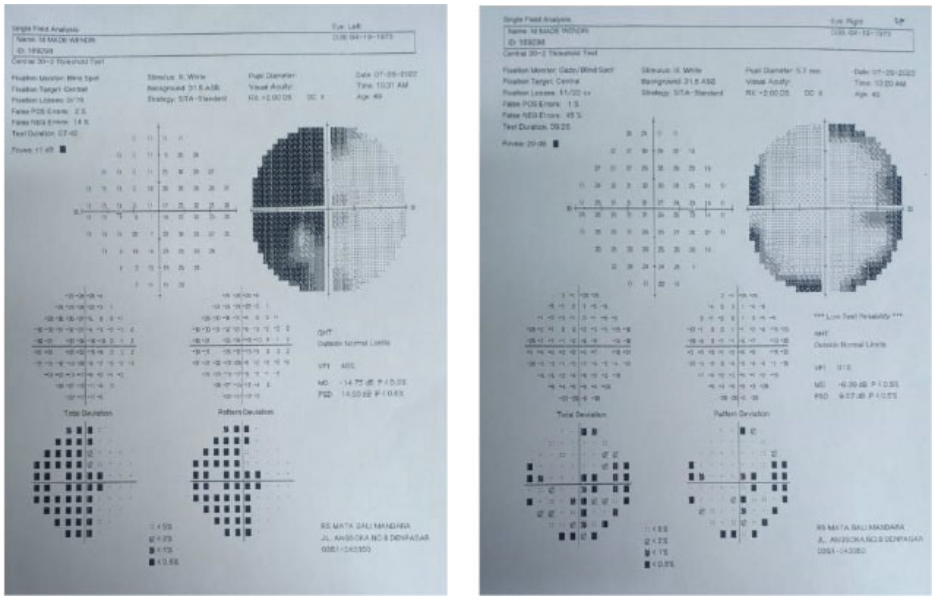

Figure 3. HVF 30-2 examination showed temporal hemianopia on the left eye.

Case 2

A 49-year-old female patient came with blurry vision in the left eye for three months before the examination. There is a history of headache, and the other systemic condition was denied. Patients with a history of contraceptive used for more than 10 years.

Visual acuity in the right eye is 6/9 with pinhole 6/7.5, and in the left eye is 6/30 with pinhole 6/12. There is a grade 2 Relative Afferent Pupillary Defect (RAPD) in the left eye. The rest examination on both eyes was within the standard limit. Funduscopic examination revealed a well-defined, round optic nerve with a flat retina and good macular reflexes in the right eye and slightly optic atrophy in the temporal area of the left eye. Mean RNFL thickness in the right eye is 102 µm, and in the left eye is 81 µm, with slightly thinning on the temporal side. A temporal hemianopia in the left eye was shown on the HVF 30-2 examination (Figure 3). A CT scan was performed on this patient. It showed a round-shaped heterogeneous density extra-axial supratentorial mass with clear boundaries in the sellar to suprasellar region, which appeared to be pressing the optic chiasma superiorly. Leads to an image of a pituitary macroadenoma (Figure 4).

Case 3

A 40-year-old female patient came with complaints of blurry vision in the left eye for two months before the examination. There is a history of headache prior to blurry vision, but systemic symptoms or other neurological symptoms were denied. There is a history of contraceptives used for more than 10 years.

Visual acuity in the right eye is 6/60 with pinhole 6/15and No Light Perceptions (NLP) in the left eye. The pupils showed round shaped on both eyes with grade IV RAPD on the left eye, the rest examination was within normal limits. Funduscopic examination of the right eye was within normal limits, and there was an optic atrophy in the left eye. Temporal hemianopia on the right eye was shown on HVF examination, whereas on the left eye could not be evaluated due to decreased visual acuity (Figure 5). CT-Scan examination revealed a solid mass with inhomogeneous contrast enhancement. The mass appears obliterating the bilateral optic chiasma, especially on the left, suggesting a macroadenoma (Figure 6).

Case 4

A 62-year-old male came with blurry vision in his left eye for 2 months before that, accompanied by headache. History of fever, eye pain, double vision,

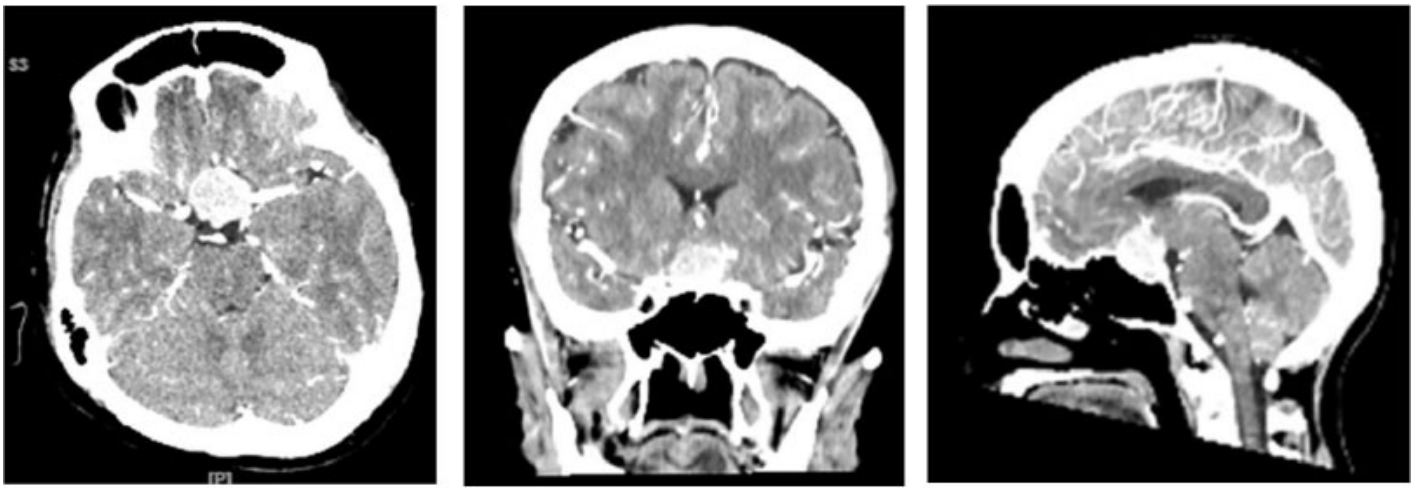

Figure 4. CT Scan examination shows that there is an extra-axial supratentorial mass with heterogeny density that is pressing the optic chiasm superiorly, suggesting the pituitary macroadenoma (red arrow).

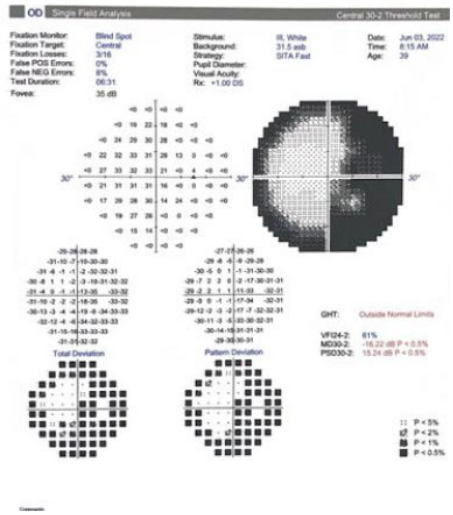

Figure 5. HVF 30-2 examination in the right eye showed temporal hemianopia.

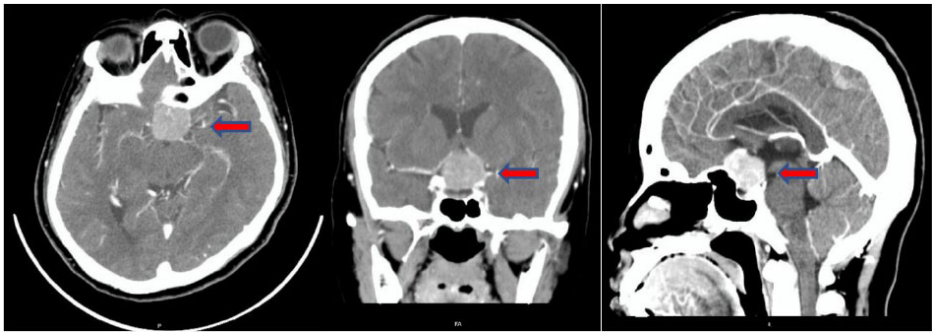

Figure 6. CT-Scan examination showed solid mass with inhomogeneous contrast enhancement measure in sella-suprasella appeared obliterating bilateral optic chiasm mostly in the left side, suggest macroadenoma.

tinnitus, weight loss, sweating, and other neurological deficits and systemic diseases was denied.

Visual acuity in the right eye is 6/60, did not improve with pinhole, and 2/60 did not improve with pinhole in the left eye. Pupil examination showed round-shaped shaped in both eyes with grade 3 RAPD in the left eye. Funduscopic examination revealed well-defined, round ONH slightly atrophic in both eyes with flat retina and good macular reflexes. The other anterior segment is within normal limits. The OCT-RNFL examination showed a mean thickness in the right eye is 82 µm and 72 µm in the left eye. The HVF 30-2 examination showed the presence of right homonymous hemianopia (Figure 7). CT-Scan examination showed there is a solid mass with perifocal edema and strong contrast enhancement in the left parasella that obliterated the cavernous sinus and optic chiasma, which extended to the left parietal, causing obstructive hydrocephalus and midline shift, suggesting a meningioma (Figure 8).

DISCUSSION

The optic chiasma is a part of the visual afferent pathway, which is the commissure formed by the optic nerve fibers. It is located 10 mm above the sella tursica, which consists of the pituitary gland, and its diameter is 12–18 mm. As the optic nerve fibers travel through the optic chiasm, about 47% of the optic nerve fibers on the temporal side will continue to the ipsilateral side, and about 53% of the nerve fibers on the nasal side will cross to the contralateral side. The nasal fibers are divided into two parts: the superior nasal nerve fibers will form the posterior Wilbrand's knee before heading to the contralateral side, and the inferonasal section will form a convex loop known as the anterior Wilbrand's knee before heading to the contralateral side. The anatomical position of the optic chiasma was defined as prefixed, normal, or postfixed based on its relative position to the sella. In most cases (80%), the position of the optic chiasm was directly above the sella diaphragm. The variations in anatomical positions of the optic chiasm in Sella tursica can cause various visual field defects when a pathological condition occurs.[1,2,3,4,5]

The pathological conditions that occur in the optic chiasm produce a neuro-ophthalmological symptom known as chiasmal syndrome, characterized by decreased visual acuity, a visual field defect, and optic atrophy on funduscopic examination. Other symptoms include

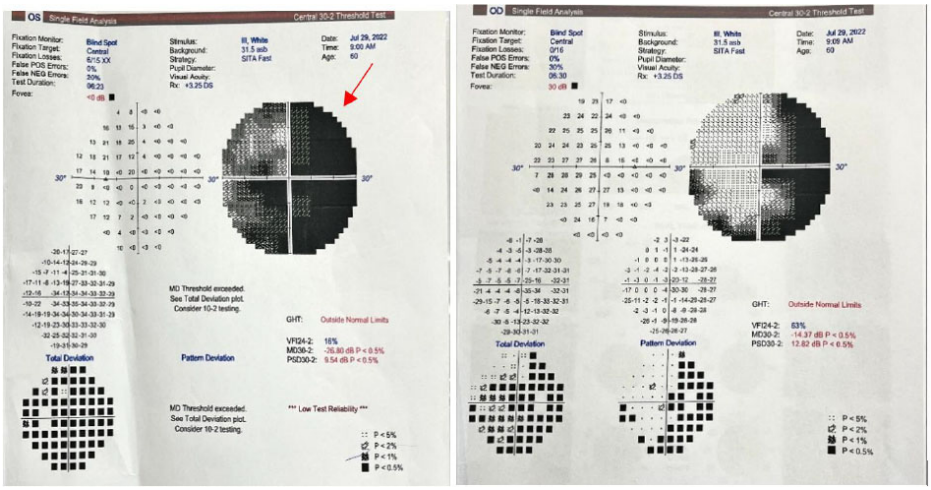

Figure 7. HVF 30-2 examination showed right homonymous hemianopia.

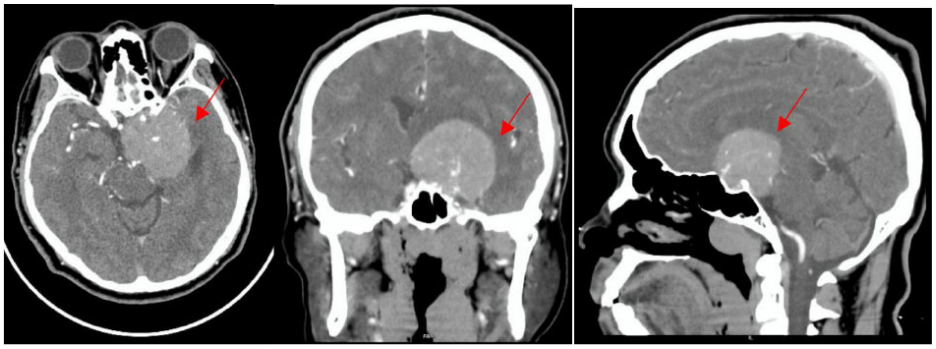

Figure 8. CT-Scan examination axial-coronal-sagittal section, solid mass in the left parasella that obliterated the cavernous sinus and optic chiasma, which extended to the left parietal, causing obstructive hydrocephalus and midline shift, suggesting a meningioma (red arrow).

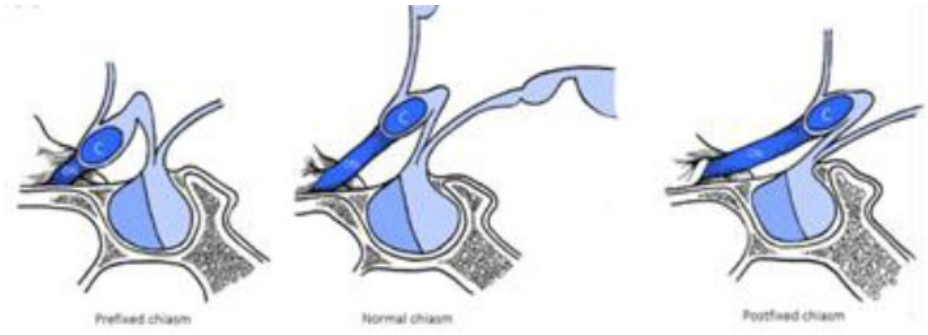

Figure 9. Variations of the anatomical position of the optic chiasm in relation to sella tursica and pituitary gland.[5]

diplopia, dyschromatopsia, defects in stereoscopic function, and systemic symptoms such as headaches and hormonal disturbances that can occur in chiasmal syndrome.[1,6] Astorga-Carballo in 2017 reported, the most common presenting symptoms found in patients with chiasmal syndrome are decreased visual acuity (54.8%), peripheral visual field defect (19.2%), systemic symptoms secondary to hormonal disturbance (17.3%), headache (9.6%), and diplopia (0.9%).[7] All of the patients in these cases reported having impaired visual acuity in one eye, with the exception of the first case, who had visual field problems. Also, headaches were discovered in three cases.

Visual field defects may occur depending on the location of the pathological condition and the anatomical position of the optic chiasm. Compression of the anterior segment of the optic chiasm, or post-fixed position, will result in a unilateral hemianopsia defect. In some cases, a scotoma may be found on the supero-temporal visual field of the contralateral eye, called a junctional scotoma, because of compression in the anterior Wilbrand's knee. Compression of the body of the optic chiasm causes a bitemporal hemianopia visual field defect, which is the most common visual field defect in lesions of the optic chiasm. This occurs due to compression of the nasal side nerve fibers that cross the optic chiasm. A defect in the posterior segment of the optic chiasm, or prefixed position, will result in a mixed homonymous hemianopia visual field defect, with a defect that has occurred in the previous optic chiasm.[2,5] In this case series, the visual field defects on the anterior segment of the chiasm were found in the second and third cases, defects in the body of the optic chiasm were found in the first case, and the fourth case shows a defect posterior to the optic chiasm.

Pituitary adenomas are the most common lesions that produce chiasmal syndrome, accounting for 12–15% of cases. Pituitary adenomas are divided into non-functioning and functioning adenomas. Neurological symptoms due to mass effect are commonly found in non-functioning adenoma, whereas hormonal disturbances are commonly found in functioning adenoma.[8] Based on their size, pituitary adenomas are divided into microadenomas when the size is less than 1 cm and macroadenomas when the size is more than 1 cm. Visual loss occurs if there is an expansion of the tumor beyond the sella tursica. Expansion to the suprasellar area and intracranial part of the optic nerve will produce chiasmal syndrome.[6,8] Meningioma in the Sella-Tursica area is a rare condition and is estimated to account for about 1% of tumors in the Sella area. Meningiomas originate from the subchiasm area, which has a tendency to compress the optic chiasm superiorly and/or posteriorly, causing visual field defects and decreased visual acuity.[8,9] Intracranial arachnoid cysts are benign intracranial cystic lesions found in 1.4% to 2.3% of intracranial lesions. Arachnoid cysts in the intrasellar region are a very rare condition,

estimated to occur in 0.6% to 0.8% of all lesions in the sellar region. Intrasellar arachnoid cysts can cause compressive symptoms mimicking those of non-secreting pituitary adenomas.[9,11] As found in this case, meningioma was present in almost all of the cases. An arachnoid cyst was only found in one case.

The visual prognosis in patients with chiasmal syndrome is related to the degree of loss of RNFL thickness caused by compression of the visual pathways, as measured by an OCT scan. The prognosis is poor if the mean RNFL thickness is less than 75 µm.[1,5] The first and second cases have a good visual prognosis because the mean RNFL thickness is more than 75 µm, as shown on the OCT scan. Poor visual prognosis was found in the third and fourth cases because there was an optic atrophy on funduscopic examination and NLP of visual acuity in the third case, and the mean RNFL thickness was less than 75 µm on the OCT scan in the fourth case.

CONCLUSION

A visual field examination is one of the most essential neuro-ophthalmologic examinations to determine the involvement of visual pathways in patients with visual impairment. Understanding the anatomy of the optic chiasm and its anatomical position and relation to the sella tursica is very important in estimating the location of the lesion that compresses the visual pathway and the resulting visual field defects. The role of ophthalmologists in the management of patients with chiasmal syndrome is vital because early detection will improve the visual prognosis of patients.

DISCLOSURES

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Ethics Approval

Written inform consent had been collected from patients regarding the use of medical data for scientific purposes. The permission was also obtained from the head of the Ophthalmology Department, Bali Mandara Eye Hospital, Bali-Indonesia.

Conflict of Interest

None to state.

Author Contribution

All authors contributed equally in the data collection and writing of this manuscript.

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