DEPARTMENT OF OPHTHALMOLOGY

FACULTY OF MEDICINE UNIVERSITAS PADJAJARAN

NATIONAL EYE CENTER CICENDO EYE HOSPITAL

BANDUNG

Case Report : Chiasmal Syndrome with Mass in Sella Turcica
Presenter : Magdalena Purnama Soeprajogo

Supervisor : Antonia Kartika, MD

Has been reviewed and approved by
Supervisor of Neuro-Ophthalmology Unit

Antonia Kartika, MD

Monday, September 21th 2020

08.15 AM

Chiasmal Syndrome with Mass in Sella Turcica

Abstract

Introduction

Undiagnosed chiasmal syndrome come to neurosurgeons only after being misdiagnosed leads to decreased visual acuity, progression of the pathological process, and irreversible loss of sight.

Purpose

to describe the clinical characteristics of patients with the chiasmal syndrome .

Case Report

a 26 years old male with a chief complaint of blurred vision on the right eye since 2 months. Visual acuity on the right eye was 0.63 and on the left eye was 1.0. The visual field testing showed bitemporal hemianopia. OCT RNFL showed marked thinning on right eye. The brain CT scan showed homogenous enhancing mass at sellar tursica. Neurosurgeon referral was advised to patient.

Result

The brain CT scan showed homogenous enhancing mass at sellar tursica. Neurosurgeon referral was advised to patient.

Conclusion

Bitemporal hemianopia are common visual field defect in chiasmal syndrome. Neuroimaging examination is mandatory to confirm the topical diagnosis. Chiasmal syndrome early detection of by ophthalmologists and timely referral to relevant specialists will facilitate improved visual function outcomes and quality of life for patients

Keywords

Chiasmal syndrome, bitemporal hemianopia, sellar tursica.

I. Introduction

Chiasmal syndrome is the name given to the group of symptoms and signs that occur together as a result of lesions affecting the optic chiasm. Pathology in any of the key anatomical structures that lie adjacent to the optic chiasm has characteristic effects on the optic chiasm, giving rise to a unique constellation of visual signs and symptoms, collectively known as the chiasmal syndrome. Depending on the particular pathology involving these adjacent structures, and the consequent relative location of damage to the chiasm. There are a variety of clinical manifestations in the chiasmal syndrome, with decrease vision as the initial presentation of many of the optic chiasm injuries. It usually presents as a temporal visual field defect by the central compression of the optic chiasm.[1,2,3]

Bitemporal hemianopia describes the ocular defect that leads to impaired peripheral vision in the outer temporal halves of the visual field of each eye. This

condition commonly results from a tumor or lesion impinging on the optic chiasm, the decussation point of the optic nerve conveying visual information from the nasal retina in each eye.[3-5]

The etiology varies from congenital, traumatic, iatrogenic causes to extrinsic or intrinsic lesions. The most common extrinsic lesion is the pituitary adenoma which may promote compression of the optic chiasm causing visual disturbances. Intrinsic cystic lesions in the optic chiasm are an common cause of bitemporal hemianopia compared with compressive lesions extrinsic to the chiasm. Therefore, a suspicion starting with the clinical history, a correct diagnosis, prompt management and rehabilitation is essential for these patients. The aim of this case report is to describe to describe the clinical characteristics of patients with the diagnosis of chiasmal syndrome. [1,4,5]

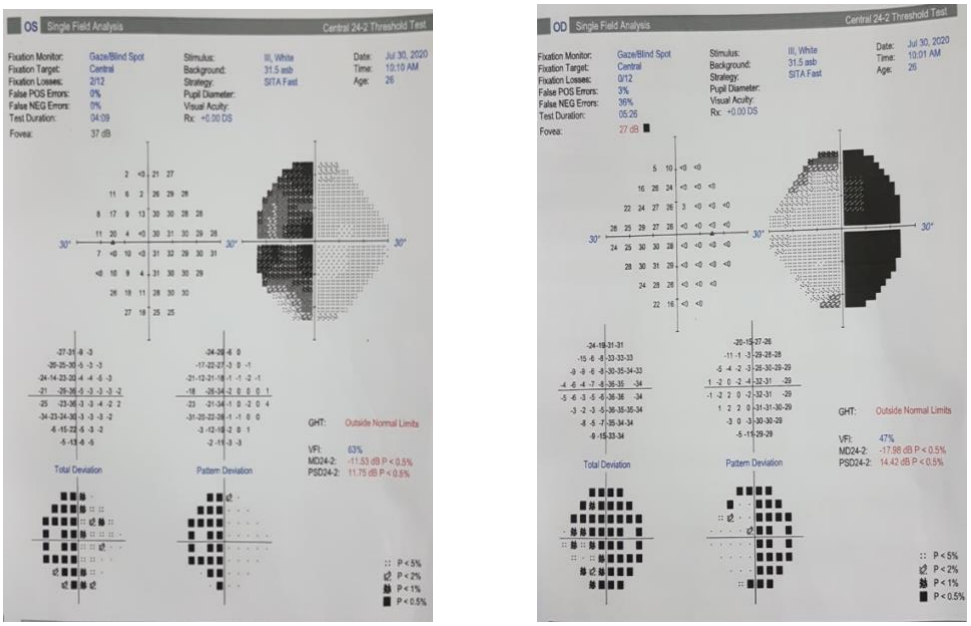
II. Case Report

A 26-year-old man came to Neuro-ophthalmology Clinic of Cicendo National Eye Hospital on July 30th, 2020 with a chief complaint of gradual blurred vision on right eyes since 3 months ago. The patient also experienced non specific headache. He did not experience red eye, nausea and vomiting, dizziness, weakness on extremity, double vision, fever, or pain with eye movement. There was no history of trauma, hypertension, diabetes mellitus, or other systemic diseases. There were no history of stroke, autoimmune disease, tumor, and alcohol or drugs consumption. The family history of stroke, autoimmune disease, tumor were denied. General examination was within normal limit, blood pressure 110/70 mmHg, pulse rate 80x/minute, respiration rate 16x/minute, and body temperature 36,5°C.

Ophthalmic examination showed visual acuity on the right eye was 0.63 difficult to pinhole and 1.0 on the left eye. Intraocular pressure on both eyes were within normal limit. Primary eye position was orthotropia with normal eye movement. Pupil on both eyes were round with direct pupillary light reflex on the right eye decreased with positive RAPD grade I. Anterior segment examination on both eye were within normal limits. Posterior segment examination on the right eye showed

round optic disc, defined margin with pallor at temporal side and on the left eye showed round optic disc, defined margin and normal cup-disk ratio. On the right eye there was temporal scotoma on amsler grid, color vision 38/38 (ishihara) and contrast sensitivity (1.25%). On the left eye there was temporal scotoma on amsler grid, color vision 38/38 (ishihara) and contrast sensitivity (1,25%). Confrontation examination showed visual field disturbance in the temporal part of both eyes. Other cranial nerve, motoric, and sensoric examination were normal. Stereopsis testing was not undertaken.

Visual field examination was performed with Humphrey Visual Field Analyzer version 3.1. with central 30.2 treshold test by Zeiss. It revealed visual field defect on both eyes bitemporal hemianopia. Peripapillary retinal nerve fiber layer thickness (pRNFL) was assessed by Cirrus HD-OCT model 5000 by Zeiss. It revealed thinning on the right eyes.



(a) (b)

Figure 2.1 HVF central 30.2 treshold test on on the left eye (a) right eye (b) showed bitemporal hemianopia.

Brain CT Scan with contrast showed homogenous mass that enhancing with cyctic multiple component at intrasellar to suprasellar and the size 50 mm x 30mm suggestive of pituitary tumor. The patient was diagnosed with bitemporal hemianopia caused by chiasm opticum lesion caused by pituitary tumor and early papil atrophy OD caused by compressive lesion. The patient was referred to the neurosurgery department to perform next treatment.

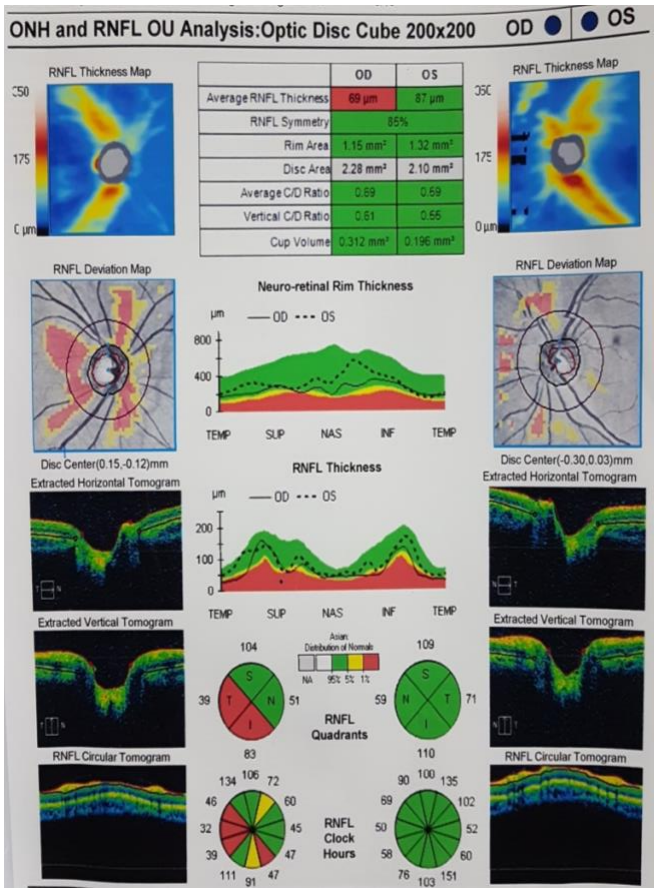
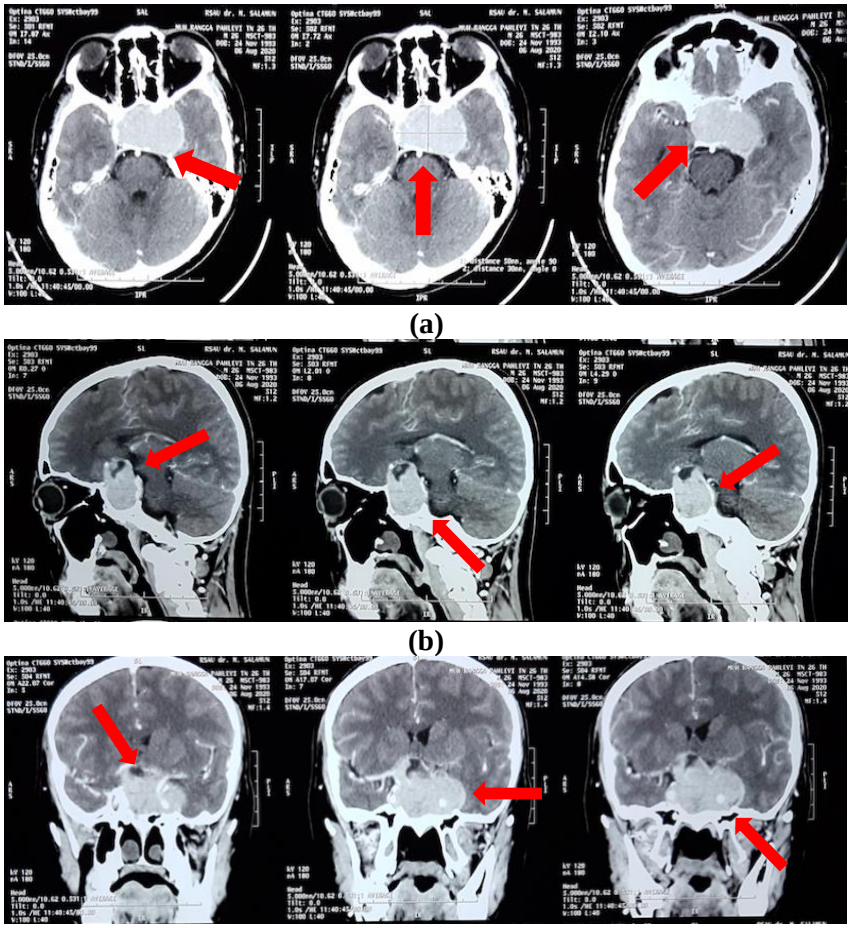


Figure 2.2 Optic disc OCT showed thinning of RNFL on right eyes



(a)

(b)

(c)

Figure 2.3 Brain CT-Scan with contrast showed enhancing mass. (a) axial plane showed homogen mass at sella tursica with size 50mmn x 30mm (red arrow). (b) sagittal plane showed homogem mass located at intra sellar to suprasellar (red arrow) and cyctic multiple component at cranial side. (c) coronal plane showed homogen mass (red arrow) at intra sellar to suprasella and cyctic multiple component at cranial side.

III. Discussion

The optic chiasm is a flattened ring composed of a tuft of fibers located at the junction of the anterior wall of the third ventricle with its floor. It contains approximately 2.4 million afferent nerve fibers that anteriorly reach both eyes and then continue along the optic tract. It measures approximately 8 mm in anterior to posterior axis, 15 mm wide and 4 mm high. The anatomic position of the chiasm is described as pre fixed, normal, or post fixed based on the relative position of the chiasm (anterior, above, or posterior) to the sella. In the majority of cases (80%) the

chiasm is situated overlying the diaphragma sellae. The optic chiasm is a structure located along the visual pathway at the point where the optic nerves of the right eye and left eye join each other. The nasal fibres of the retina decussate at the optic chiasma. It is the arrangement of the fibres at the chiasma that accounts for the typical visual field defects seen when a lesion occurs at the site. The most common field defect observed in patients with optic chiasmal injury is bitemporal hemianopia, although the field loss can be of varying degrees. [4,6,7]

The chiasmal syndrome is a constellation of signs and symptoms that include changes in the visual field, decreased visual acuity and atrophy of the optic nerves, which are associated with lesions in the optic chiasm. Other symptoms such as diplopia, alterations in chromatic sensitivity, changes in the appearance of the head of the optic nerve, headache and systemic manifestations secondary to variations in the pituitary hormones may also arise. A visual field defect may be the earliest sign of a lesion at the optic chiasm and the classical visual field defect seen in chiasmal syndrome is the loss of the temporal hemi-fields of vision in both eyes (so-called bitemporal hemianopia). In this case, bitemporal hemianopia found in the confrontation test and Humprey examination which indicates a lesion in the body of the chiasm which damages the crossing nasal retinal fibers. Any lesion of the chiasm (such as a tumor, vascular lesion, traumatic lesion, etc.) may cause this, and there are subtle variations in the extent of the bitemporal hemianopia that can help give clues as to the causative lesion. The exact field defect will vary according to the type of lesion and the precise individual anatomical relationship of the chiasm to the structures that surround it. Loss of the temporal visual fields can lead to postfixation blindness and the inability to keep the eyes both facing in the same direction, a phenomenon known as hemi-field slide. [2,5,7]

Visual impairment that is the most common reason for the initial visit of patients with undiagnosed chiasmal syndrome to the ophthalmologist's office, although visual impairment is not an early symptom of the disease. The visual acuity may also be reduced, as may the color vision on testing with pseudoisochromatic plates, with a tendency to have more severe red-green deficits and milder blue-yellow losses. This patient reported 3 moths duration of gradual blurred vision on right eye.

Another frequent complaint is headache which is usually referred as retro orbita. In some cases, patients may consult for diplopia due to the injury of the oculomotor nerves. This patient reported non specific headache which has been attributed to some causes including mechanical compression of the mass. [6-8]

The compressive optic neuropathy is associated with pallor of the optic nerve and visual field defects that obey the vertical meridian. Compressive optic neuropathy is associated with significantly thinner nasal and temporal sectors. This patient showed optic disc atropy at temporal side on right eye in posterior segmen examination. Ophthalmic symptoms have a prognostic value since the disease may lead to a total loss of visual function. The primary pathogenetic mechanism of chiasmal syndrome is local compression of the chiasm proper from pathology focus which, in turn, leads to optic nerve fiber atrophy. The optic chiasm also lies close to a venous space (the cavernous sinus) where the nerves that control eye movement pass – lesions in this area can damage these nerves and cause further problems with the eyes moving together, resulting in diplopia. Anyone presenting with the above features of chiasmal syndrome should be investigated with urgent neurological imaging, in order to promptly diagnose and, if necessary, treat a potentially life-threatening cause. Neuroimaging was performed to identified the exact location of the lesion. This patient undergone brain-CT Scan with contrast showed homogenous mass that enhancing with cyctic multiple component at intrasellar to suprasellar suggestive of pituitary tumor and referral to neurosurgery unit. [5,6,9]

The unique configuration of the optic chiasm, causes that a lesion typically produces changes in visual function, particularly visual field defects that are in some cases diagnostic, the pattern of visual field loss reflects the location. Typically, the pituitary adenoma compresses the chiasm from its caudal aspect, causing a bitemporal hemianopsia that starts affecting the lower fibers and then the upper fibers. Aline et all repoted the bitemporal hemianopsia may be reported as frequent as 41% and the most common diagnosis was pituitary macroadenoma. The most common lesions producing the chiasmal syndrome include pituitary adenoma, parasellar meningioma, craniopharyngioma, and parasellar internal carotid artery aneurysm. Other CNS mass lesions can produce third-ventricle

dilation and secondary posterior chiasmal compression. Pituitary adenomas are the most common cause of chiasmal compression and may occur at any age, although they are rare in childhood. Patients with nonsecreting tumors typically present with vision loss, their tumors having reached a relatively large size without causing other symptoms. Tumors that actively secrete hormones (such as prolactin or growth hormone) are often detected before vision loss occurs because of their systemic endocrine symptoms. In this case the patient was 26 year with no systemic endocrine symptoms. [8,10,11]

Tumors smaller than 10 millimeters are called microadenomas; they are located within the sella. Ophthalmological abnormalities arise when there is tumor expansion beyond the sella turcica. Ophthalmological symptoms depend on the direction of growth of the tumor. Suprasellar extension of the pituitary adenoma and intracranial portions of the optic nerves produce a chiasmal syndrome. The tumor initially compresses the central chiasm containing decussating nasal retinal fibers from below the optic chiasm. This is followed by the development of a bitemporal hemianopia. Decreased visual acuity is found when the tumor affects the intracranial portion of the optic nerve. [9,12,13]

Chiasmal syndrome as a manifestation of a chiasmal- and- sellar disorder requires surgical treatment in most cases. A study of Danesh-Meyer et al has established a clinical marker that correlates strongly with the degree of visual recovery after surgical intervention in patients with significant visual loss as a result of chiasmal compression. The degree of reversibility of visual dysfunction with compression of the anterior visual pathway is related to the loss of RNFL thickness, as measured by the OCT. The study demonstrated that there is an increasing probability of improvement to near normal visual function with increasing RNFL thickness up to approximately 85 μm , after which there is no further improvement in visual function. Ganglion cell complex (GCC) thinning can occur before visual function loss. OCT analysis of the ganglion cell layer may be useful for the prognosis of visual function recovery from surgical or medical decompression of the chiasm. Early surgical decompression before significant RNFL and GCC thinning may lead to better postoperative visual function. In the presence of

bitemporal hemianopia, optic nerve pallor is a predicting factor for postoperative visual function. This patient the RNFL thickness, as measured by the OCT showed 69 μm suggest the prognosis visual function after surgical intervention was dubia ad malam. The prognosis of quo ad vitam and sanationam is depending on the etiology. Therefore the prognosis of quo ad vitam and sanationam in this case was dubia. [11,12,14]

IV. Conclusion

Detection of the focus of the disease in the retrobulbar region is of great practical importance since in such cases atrophy is frequently caused by neurosurgical disease and thus requires not ophthalmological, but neurosurgical care. Patients with supposed chiasmal syndrome should undergo a comprehensive eye examination including visual acuity, pupil responses, ocular motility, intraocular pressure measurement, anterior biomicroscopy, direct ophthalmoscopy, color vision testing and careful visual field testing. The main ophthalmologist's task in these patients is to detect early signs of compressive optic neuropathy before descending optic nerve atrophy develops. Conducting careful visual field studies in the early disease is critically important. Color vision studies may reveal the earliest manifestations.

Reference

1. Cantor LB, Rapuano CJ, Cioffi GA. The Patient With Decreased Vision: Classification and Management. In: Neuro-Ophthalmology. San Fransisco. American Academy Ophthalmology. 2016. 109, 150-154p.
2. Miller NR, Subramanian PS, Patel VR. Topical Diagnosis of Optic Chiasmal and Retrochiasmal Lesions. In: Walsh and Hoyt's Clinical Neuro-Ophthalmology The Essentials. Third Edition. Philadelphia. Wolters Kluwer. 2016. 12, 380-387p.
3. Trobe JD. The neurology of vision. Oxford ; New York: Oxford University Press; 2001. 451 p.
4. Salmon JF. Neuro-ophthalmology. In: Kanski's Clinical Ophthalmology : A Systematic Approach. Ninth Edition. United Kingdom. Elsevier. 2020. 786-795p.
5. Rubin RM, Sadun AA, Piva AP. Lesion of the Optic Chiasm, Parasellar Region, and Pituitary Fossa. In: Ophthalmology Fifth Edition. Philadelphia. Elsevier Inc; 2019. 909-917p.
6. Haque S, Reid K, O'Neil R, Lueck C. Cystic Optic Chiasm Lesion: Atypical Magnetic Resonance Imaging Findings. Neuroophthalmology. 2017 Aug; 41(4): 211–214.
7. Carbalo AA, Ojeda JC, Suarez MF. Chiasmal syndrome: Clinical characteristics in patients attending an ophthalmological center. Saudi J Ophthalmol. 2017; 31(4): 229–233.
8. Amir AJ, Ang L. Case Series on Chiasmal Lesions with Ocular Manifestations seen at the Eye Center of a Tertiary Government Hospital in Philippines. Jos. 2020;2(3):2436-470.
9. Moonkan LV, Thomas PA, Harwani AA. Traumatic chiasmal syndrome: A meta-analysis. Am J Opthalmol Case Rep.2018 Mar; 9: 119–123.
10. Martin JT, Corbett JJ. Practical Neuroophthalmology. Chapter 5: Disorders of the Chiasm and Retrochiasmal Visual Pathways. 2013 by McGraw-Hill Education LLC
11. Azmeh A. Neuro-Ophthalmology Findings in Pituitary Disease. 2018. 83-98.
12. Yazici B, Kivanc SA. Isolated bitemporal hemianopsia due to traumatic chiasmal syndrome. Ulus Travma Acil Cerrahi Derg 2016;22(1):97–99
13. Carballo AA, Ojeda JC, Camargo MF. Chiasmal syndrome: clinical features in mexican patients, a 5 year review. *Invest. Ophthalmol. Vis. Sci.* 2015;56(7):5559.
14. Vasyuta VA, Pedachenko YE. Neuro-opthalmological aspect of chiasmal-and-sellar disorders. Journal of Ophthalmology (Ukraine). 2019;1 (486):72-77.