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ORIGINAL ARTICLE

VISUAL FIELD DEFECTS IN NEUROLOGICAL DISEASES

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K.J.N. Sivacharan, C.H. Madhavi, Hanumantharao G. "Visual Field Defects in Neurological Diseases". Journal of Evolution of Medical and Dental Sciences 2014; Vol. 3, Issue 05, February 03; Page: 1134-1145, DOI: 10.14260/jemds/2014/1957

BACKGROUND: Visual field defects in neurological diseases have diverse presentations and provide a clue in localizing the site of lesion, help us in knowing the progression of a disease. They also guide us in assessing the post operative recovery in surgical removal of a tumor. Visual field defects also help in modifying the life style of patients. standard automated perimetry is procedure of choice and is easy, reproducible, accurate and has replaced most of the conventional ways of assessing visual fields **AIMS:** The aim of present study is to evaluate and study the defects in visual pathway cause by lesions involving various parts of the visual pathway. **SETTINGS:** The study was conducted in a teritiary eye care center on all patients who complained of visual field defects attending the outpatient department of ophthalmology and who have been referred from other specialities with complaints of visual field defects. **DESIGN**: This was a retrospective analysis of all cases who have been evaluated with visual field defects between 2003 – 2004. **MATERIALS AND METHODS**: This was a retrospective study on 100 patients who have been evaluated with visual field defects between 2003 – 2004. **RESULTS**: Bitemporal hemianopia was seen in 19 patients. Homonymous hemianopia was seen in 64 patients. Quadrantanopia was seen in 4 cases. Enlarged blind spot was seen in 10 cases. **CONCLUSION**: Visual field assessment is important in evaluating the lesions involving visual pathway standard automated perimetry is the procedure of choice for majority of lesions involving the visual pathway. Visual field defects are use to monitor the progression and recurrence of disease and as a guide for treatment. Visual field testing should be performed in all patients with lesions of visual pathway.

KEYWORDS: Pituitary adenoma, hemianopia, visual fields, quadrantanopia, homonymous hemianopia, bitemporal hemianopia

INTRODUCTION: The ocular signs of Central Nervous system disorders often appear complicated and confusing, but in most cases they are explained by anatomy of that part of Nervous system. There are several potentially serious diseases of nervous system which may first present with ocular manifestations. Ocular examination should specifically include visual acuity, visual fields, color perception, extra ocular movements and fundoscopy. [1] Visual field testing forms an important part of examination and helps in localizing the site of lesion. Visual field defects are very important because they measure the functional visual loss. The perimetry is and objective method of assessing visual damage. Automated perimeters are objective accurate and supported by useful software packages to assist in assessment of visual fields with statistical level of confidence.

AIMS AND OBJECTIVES: The aim of present study is to present different types of visual field defects in various neurological diseases detected by Humphrey's automated perimeter.

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MATERIALS AND METHODS: Hundred patients who attended the outpatient Department of Ophthalmology with complaints of defective vision or patients who have been referred from other medical specialties with complaints pertaining to visual fields defects were included in the present study. In all the above cases, the other major cause of visual field defect, glaucoma has been ruled out by measuring intra ocular pressure and optic disc evaluation.

INCLUSION CRITERIA: Patients who attended the outpatient department of ophthalmology with complaints of defective vision or patients who have been referred from other medical specialties with complaints pertaining to visual fields defects

EXCLUSION CRITERIA:

1. Co-existing cataract and glaucoma
2. Patients too ill to allow and adequate visual field assessment on automated perimetry

FIELDS PROTOCOL: The 30-2 programme on Humphrey's Field Analyzer with a white on white Goldman size 3 target was use for visual field examination. All patients underwent full threshold strategy for visual field examination. The reliability criteria used were fixation losses < 20%, false positive and false negative errors < 33%. Only fields reliably performed were including the analysis.

Any field defect was diagnosed by following criteria:

1. Depression of thresholds 5 DB or more in 3 or more contiguous points adjacent to the vertical mid line.
2. Pattern deviation plot showing 3 or more contiguous points adjacent to the vertical mid line in the involved quadrant depressed to 1% probability level with normal mirror image points across midline.

All cases were assessed radiologically by CT / MRI with or without contrast.

RESULTS: In our present study mean age of presentation of pituitary adenoma was 33 years. Typical bitemporal hemianopia was seen in 19 patients, atypical visual field defects were seen in 9 cases. Pituitary adenoma was the predominant cause of bitemporal hemianopia. Craniopharyngioma was the cause in 3 cases. Pituitary tumors which were only more than 10mm in height could only cause chiasmal compression. The precise visual field defect caused by a lesion compressing the optic chiasma depends on location of optic chiasma with respect to sella tursica. The direction in which growth of lesion occurs and whether lesion directly damages the nerve fibers or it damages by compressing them against contiguous structures.

Homonymous hemianopia was seen in 64 cases. Complete Homonymous hemianopia was seen in 42 cases. Incomplete Homonymous hemianopia with less involvement in one or two quadrants was seen in 22 cases. Congruous Homonymous hemianopia was seen in 22 cases. Congruous Homonymous hemianopia is defined as when the visual field defects in each eye are completely symmetric in each and every aspect to the other eye. Cerebrovascular accident was the common cause of homonymous hemianopia and was more common in males. Male predominance was probably due to patients being chronic smokers, alcoholics, hypertensives which were implicated in cerebrovascular accident. Congruous homonymous hemianopia is common with lesions involving

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the posterior part of visual field. Field defects produced by lesions in temporal lobe were denser superiorly while field defects produced by lesion of parietal lobe were denser inferiorly occipital lobe hemianopias were associated with macular sparring.

4 cases of quandrantanopia were observed and all cases were seen in age group of less than 50 years of age. All the 4 cases showed pie in the floor (Inferior-quadrantanopia). Other defects that can be seen are superior quadrantanopia.

10 cases presented with enlarged blind spot secondary to papilledema or idiopathic intracranial hypertension.

4 cases showed diffuse field loss secondary to demyelinating lesions secondary to multiple sclerosis.

DISCUSSION: Visual field assessment is important in evaluation of lesions involving visual pathway and should be done at initial presentation and periodically in follow up. Standard automated perimetry helps in following ways [2]

1. Diagnostic-Helps in localizing site of lesion
2. Follow up-provides excellent tool to monitor the resolution or recurrence of disease processes affecting visual pathway
3. Daily lifestyle of patient.

Visual field testing can be performed by a number of different techniques including confrontation (at the bedside), tangent screen, Goldman kinetic perimetry, and automated static perimetry. [3]The sensitivity of confrontation techniques is about 20% for detection of arcuate field defects and about 70% for detection of hemianopia when compared to Goldman. [4] Standard automated perimetry provides adequate testing of the visual field in a majority of neuro-ophthalmic patients. Goldman perimetry is useful in patients with severe visual and neurological deficits or patients with “isolated peripheral visual field defects”. [5] However, lack of trained technicians limits the use of Goldman perimetry to a few centers.

To understand the lesions involving the visual pathway it is necessary to understand the anatomy of visual pathway.

The course taken by the visual fibres from the retina through to visual cortex is known as primary visual pathway

Visual pathway from the retina:

1. Optic nerve
2. Optic chiasma
3. Optic tract
4. Lateral geniculate body
5. Optic Radiation
6. Striate Cortex
7. Peristriate Cortex

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Optic nerve: Optic nerve in essence is a tract of brain, outgrowth of cerebral vesicle, whose fibers possess no neurilemma unlike any peripheral nerve. Optic nerve can again be divided into

1. Optic nerve head
2. Intraorbital portion
3. Intracanalicular portion
4. Intracranial portion

Optic chiasma: Partial decussation of human retinal axons takes place in optic chiasma. Optic chiasma is a flattened band embedded in anterior third of ventricle between two thalami and projecting into the chiasmatic cistern. It lies 10mm above the sphenoid.

Retinotopic distribution of axons continues in optic chiasma

1. Uncrossed fibres-form 47% of total fibres and lie in temporal part of chiasma and carry axons from ipsilateral temporal hemiretina
2. Crossed fibres-form 53% of total fibres and carry fibres from nasal half of retina, macular fibers of retina send both crossed and uncrossed projections

Optic tracts: Optic tracts extend posteriorly from angle of chiasma between tubercinerium and the anterior perforated substance. In the optic tract fibres from optic chiasma are rearranged to correspond with the position of lateral geniculate body. Macular fibres (crossed and uncrossed) occupy an area dorsolaterally. Lower retinal quadrants are lateral and fibres from upper are medial, fibres from peripheral part of retina lie more anteriorly.

Lateral geniculate body: Lateral geniculate body is an oval structure located at termination of optic tracts. Each geniculate body consists of 6 layers of neurons. Crossed fibers end in layer 1, 4, 6 and uncrossed fibers in layers 2, 3, 5.

Optic radiation: Optic radiation or geniculo calcarine pathway arises in the lateral geniculate nucleus and is the relay station of fibres carrying visual impulses to the occipital lobe. In the optic radiation, upper retinal fibres occupy upper part of optic radiation and lower retinal fibres occupy lower part of optic radiation. Macular fibres are in central part of optic radiation.

Striate cortex: Striate cortex is largely on medial aspect of occipital lobe around and in calcarine sulcus with extensions into uncus and lingual gyrus. There is point to point localization of retina in cortex, each area of retina being precisely represented in a corresponding area of striate cortex. Upper and lower quadrants of retina are represented respectively above and below the calcarine sulcus. Periphery of retina is represented anteriorly and macula towards occipital pole. The most anterior part of striate area represents the extreme nasal periphery of retina, corresponding to monocular temporal crescent in visual field.

Visual field defects due to optic pathway lesions are caused by variety of causes such as

1. Tumors
2. Neuro vascular lesions
3. Infections

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Because of diverse pathological mechanisms of these defects most of them demand the combined attention of ophthalmologists, neurologists, neurosurgeons and radiologists. However ophthalmologists and neurophthalmologists assume an important role in their early diagnosis.

1. Cerebro vascular accidents are one of the common causes of the visual field defects.
2. Rupture of intra cranial aneurysm – since the circle of Willis is closely related to optic chiasma, optic tracts, these rupture giving rise to characteristic visual field defects
3. Pituitary tumors represent 10-15% of intra cranial neoplasms which also cause characteristic visual field defects

Visual field defects can also be used in follow up of patients who undergone neurosurgery for tumor removal and to assess the recovery of visual function.

Lesions caused by diseases of optic nerve

Centrocaecal scotomas-lesions involving the papillomacular bundle

1. **Unilateral**-
   1) Optic neuritis (sudden visual loss)
   2) Compressive lesion-slowly progressive visual loss
2. **Bilateral**
   1) Some cases of optic neuritis
   2) Chronic papilledema

Papilledema refers to the swelling of the optic nerve head secondary to raised cerebrospinal fluid (CSF) pressure. Idiopathic intracranial hypertension is defined as raised intracranial pressure (ICP) in the absence of radiologic and laboratory abnormalities reflecting any other known cause of raised ICP. Vision loss is the most feared complication of IIH with at least 10% of patients progressing to blindness from IIH. The visual field defects that result from papilledema in IIH are “disc-related defects” and are similar to those found in glaucoma. Visual field losses may be identified in as many as 96% of patients using disease-specific perimetric strategies on automated perimetry. The most common defects seen in IIH are blind spot enlargement, generalized constriction, and loss of the nasal visual fields, especially inferonasal [6] Other common field defects described include inferior altitudinal loss, superonasal and superotemporal, loss, arcuate defects, and scotomas (central, cecocentral, and paracentral. Patients with IIH should be followed with sequential quantitative perimetry to aid rational decision making and prevent vision loss. The same visual field testing strategy should be used at each visit to obtain comparable data in follow-up.

Visual field defects in optic neuritis at presentation included diffuse visual field loss (48%), altitudinal defects (15%), central or cecocentral scotoma (8.3%), arcuate or double arcuate (4.5%), hemianopic defects (4.2%), and others. [4] Patients with hemianopic field defects (13% during the first year) were more likely to show abnormalities on the brain magnetic resonance imaging (MRI) at baseline as compared to patients without these field defects, indicating the presence of demyelinating lesions and multiple sclerosis. [7] Visual fields help quantify the depth of visual field loss, identify atypical cases of optic neuritis, and aid in counseling patients about prognosis.

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Visual field defects in nonarteritic anterior ischemic optic neuropathy (NA-AION) include altitudinal field defect (classically occurring in the inferior hemi field), central scotoma, arcuate scotoma, and quadrantic defects. [8]

Lesions caused by diseases affecting the chiasma

Binocular vision begins at chiasma and it is the first portion where single lesion produces simultaneous defects in visual field. Lesions of the optic chiasm can produce a variety of visual field defects including bitemporal hemianopia, junctional scotoma (anterior chiasmal defect), quadrantanopia and bitemporal, or unilateral temporal scotoma depending on the site and extent of the lesion.

1. Bitemporal hemianopia-typically seen in of pituitary adenoma most commonly. Loss of visual field in temporal hemianopia is clockwise in right eye and anticlockwise in left eye. Restoration of visual field defect after surgical removal of tumor is in opposite direction of field loss
2. Binasal hemianopia in optic chiasma lesions occurs in
   a. Multiple sclerosis
   b. Aneurysm of internal carotid artery with displacement of chiasma to opposite side
   c. Arachnoiditis of optic chiasma, tumors with ventricular dilatation and by hydrostatic compression of chiasma.
   d. Infratentorial tumors with ventricular dilatation and hydrostatic compression of chiasma
   e. Intraventricular tumors of third ventricle
   f. Meningiomas of lesser wing of sphenoid
3. Altitudinal hemianopia-inflammatory lesions of optic chiasma
4. Homonymous hemianopia-posterior portion of tumor involving the beginning of optic tracts.

Visual field improvement following resection of the pituitary tumor occurs in three stages.

Stage one - Is the early fast phase of recovery seen within few days to a week of the surgery. In a few individuals, there can be complete normalization of the visual fields.

Stage two - Is the phase of slow recovery which is seen within a few weeks of the surgery to a few months. During this stage, the visual fields show significant and presumably slow and sustained improvement.

Stage three - Is the late phase starting a few months after decompression to a few years. During this stage, there is minimal improvement of the visual fields.

Poor prognostic signs for improvement of visual fields include dense and extensive preoperative visual field deficit, pituitary tumor volume greater than 5 cc and the postoperative development of a surgically “empty sella” (which is associated with inflammatory scarring and descent of the chiasm into the empty sella). [9]

Visual fields should be obtained periodically (1–3 months or more frequently) based on the clinical presentation, lesion characteristics, type of intervention (surgical, medical, or radiation), and patient’s visual complaints. SAP seems to be more sensitive in detecting early field defects. Goldman fields may be used to assess peripheral fields in patients with advanced field defects. Visual fields are crucial in guiding ongoing treatment and judging treatment success in a number of patients with sellar lesions.

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Post chiasmal visual pathway: Post chiasmal interference in visual pathway produces homonymous hemianopia in visual field from a single lesion. Homonymous hemianopia is bilateral and consists of loss of vision in two right halves / left halves of visual field. Lesions in anterior part of optic tract produce incongruous visual field defects. Post chiasmal visual pathway comprises of 2/3 of entire visual system. Neurological, radiological, laboratory evidence are essential in localizing site of lesion but visual fields produce a corroborative evidence in localizing site of lesion. Vision loss in a patient with retrochiasmal pathway lesion indicates macular splitting and usually indicates that more than half of macula is involved since 20/20 vision is possible when half of macula is functioning.

Lesions of lateral geniculate body: They are extremely rare. Lesions of lateral geniculate body produce relatively congruous hemianopia.

Lesions involving optic radiation: Optic radiation is most commonly involved in lesions affecting internal capsule such as vascular, traumatic, brain abscess of otogenic origin. Visual defects vary from sector shaped defects, quadrantanopias to total hemianopia. [10]

Lesions caused due to involvement of occipital striate cortex: Characteristic visual field defects are congruous homonymous hemianopia with macular sparring since macula area due to double blood supply of macular area. [11] Trobe et al provided a practical analysis of patients presenting with isolated Homonymous Hemianopia. Out of 104 cases studied 89% of the disorders were due to vascular occlusion, 3.5% subsequent to trauma, 3% associated with neoplasms and 4.5% were due to miscellaneous causes. [12]

In a recent study on the natural history of homonymous hemianopia, spontaneous improvement of the visual field defect has been observed in 38% of patients. Visual field improvement was defined as an improvement of the field defect by at least 10° horizontally and 15° vertically using similar isopters on the Goldman visual fields and significant changes in mean and pattern deviations in Humphrey visual fields. [13] The probability of finding an improvement in the visual field defects was 50–60% if the initial visual fields were obtained within a month of the neurological injury and this dropped to about 20% if the initial fields were obtained after 6 months. No other factor including age seemed to affect significantly the chances of visual field improvement. The natural history of visual field improvement is especially important when evaluating claims of improvement by potential rehabilitation treatments for homonymous hemianopia. Studies have shown that in patients with hemi paresis from a unilateral hemispheric stroke, the probability of achieving relative independence in ambulation and self-care function diminished significantly when accompanied by a visual field defect. [14]

Visual fields should be assessed in all patients who present with lesions that are close to or involve the visual pathway. Follow-up visual fields may be obtained at 6- to 8-week intervals (earlier for more aggressive lesions such as tumors) until the fields have stabilized. [15] Patients unable to meet the rehabilitation goals in a timely manner should be suspected to have additional deficits such as visual field defects and evaluated by perimetry.

Visual field deficits arising from neuro-ophthalmic conditions can adversely affect the quality of life and activities of daily living. Abnormalities on the subscales of the National Eye Institute Visual

Function Questionnaire (NEI-VFQ 25) are seen in different neuro-ophthalmic conditions including optic neuropathy from multiple sclerosis, chiasmal defects from pituitary adenoma, and homonymous hemianopia from retrochiasmal lesions (infarct, trauma, tumor, and hemorrhage). Deficits on the VFQ subscales often do not correlate with objective parameters of visual function and an understanding of the different subscales and how it affects certain visual tasks may aid in planning better assessment strategies and rehabilitation.

Homonymous hemianopia causes patients to have impairment of daily activities such as personal hygiene, meal preparation, driving, shopping, and telephone usage. Patients with homonymous hemianopia involving the central 5° complain of difficulty in reading, and are classified as "hemianopic dyslexia" Patients with homonymous paracentral scotoma may be impaired while driving despite having a relatively large visual field intact. The scotomatous area often overlies the side-view mirror on one side and impairs the ability to change lanes safely. The subject of minimum visual field requirements for driving has been one of much debate. Hemianopic patients have demonstrated poor blind side hazard detection for pedestrians that were not compatible with driving in driving simulators.[16] Patients with hemianopia and quadrantanopia (especially inferior quadrantic defects) were noted to have difficulty with lane position/lane change, steering steadiness, and gap judgment compared to normal controls. Other studies dispute the importance of visual fields for driving safety and standards and suggest that assessment of on-road driving performance under the supervision of a trained specialist may be the best option. The International Council of Ophthalmology (ICO) recommends a binocular continuous field of 120° in the horizontal meridian split approximately between the right and left halves as the minimum visual field requirement.

Visual restoration therapy: In an article British Journal of Ophthalmology Sebal et al described a visual restoration therapy in patients of Homonymous Hemianopia by patients practicing perimetry for one hour every day six days a week for six months.

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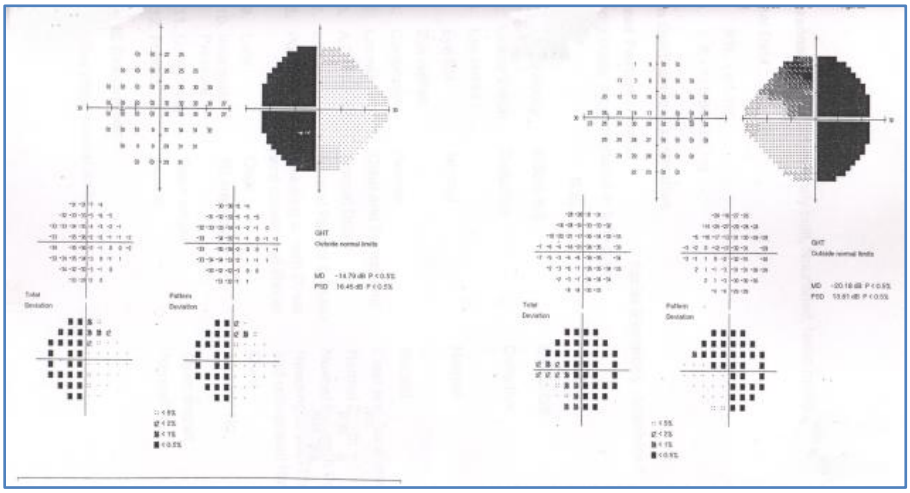

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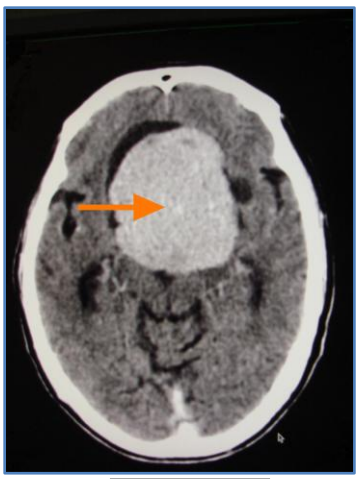

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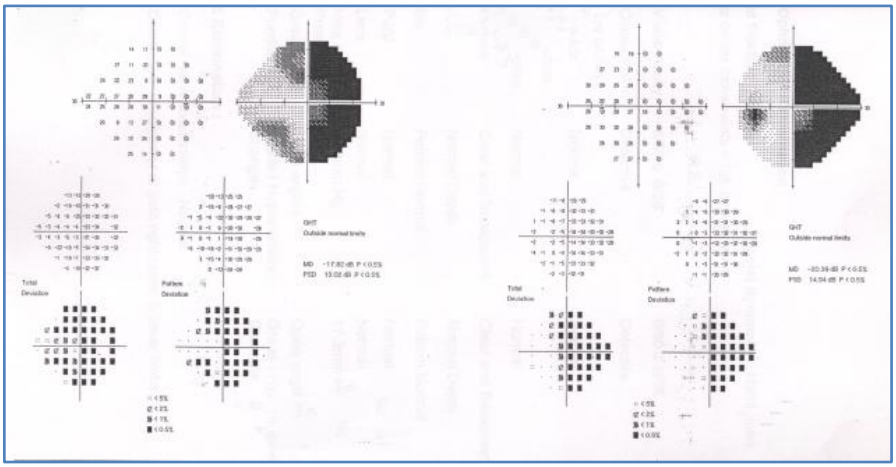

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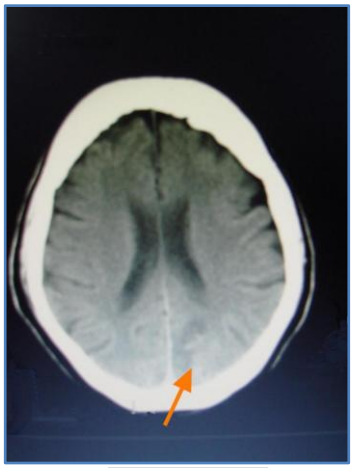

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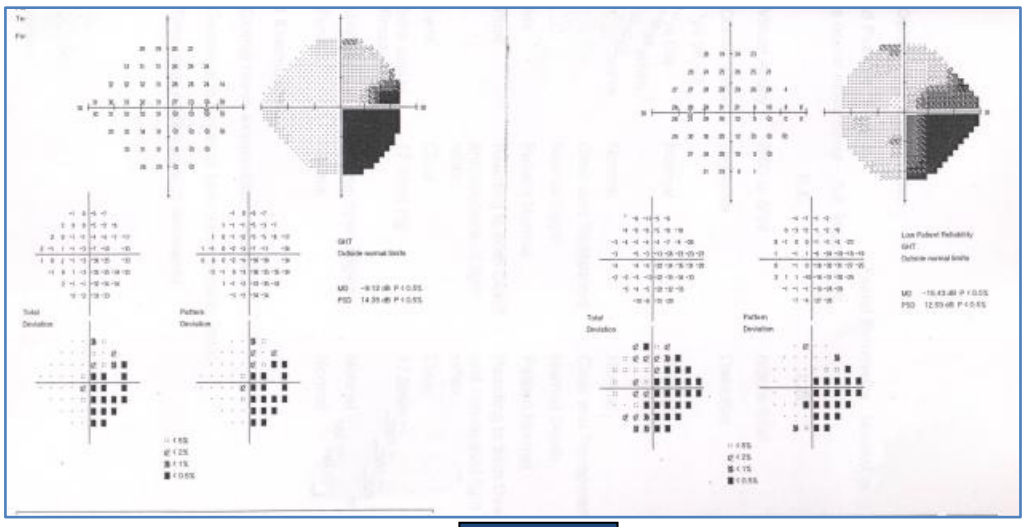

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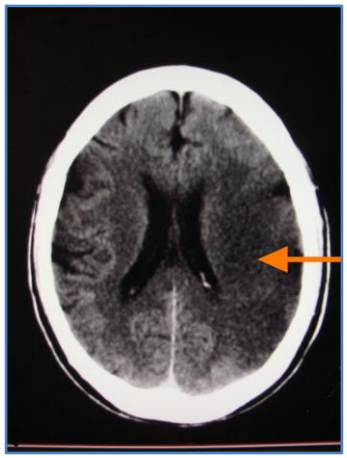

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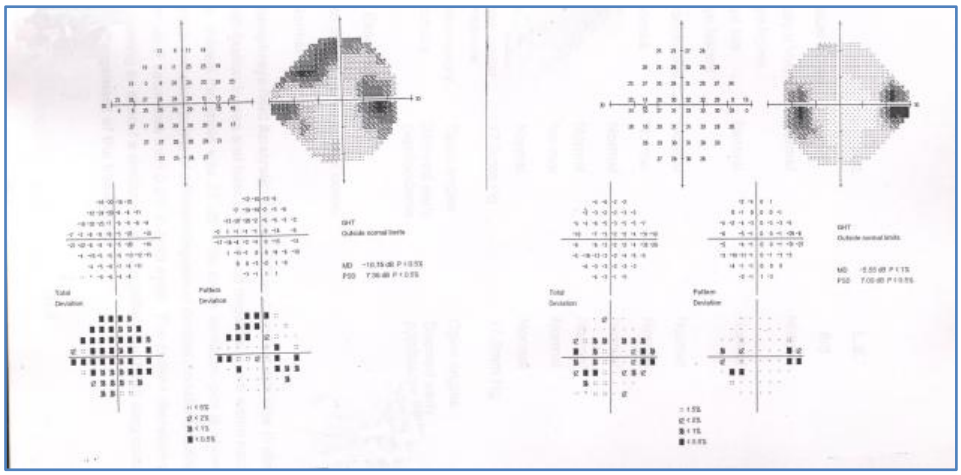

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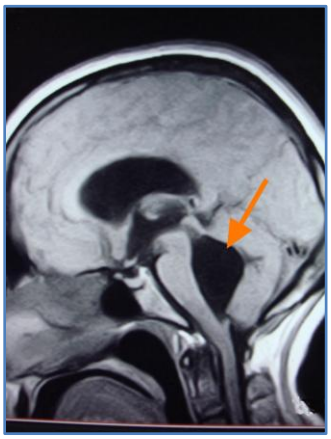

Photo 8

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