Chapter

1

Loss of Vision

INTRODUCTION

Loss of vision is a common and often alarming neurological presentation. Accurate localisation of the lesion—whether ocular, prechiasmatic, chiasmatic, postchiasmatic, or cortical—is critical for timely diagnosis and management. This chapter provides a structured, bedside-oriented approach to vision loss, emphasizing clinical reasoning, anatomical localization, and neurologically relevant differentials.

Case 1: A 60-year-old male presented with history of episodic loss of vision in left eye lasting for 1–2 minutes with complete recovery in-between. What is the diagnosis?

Amaurosis fugax or transient monocular blindness (TMOB) is the classical symptom of carotid artery disease. Severe stenosis of ipsilateral internal carotid artery (ICA) can result in reduced blood flow in ophthalmic artery (first intradural branch of ICA) with resultant retinal ischemia and TMOB. These patients complain of constriction of visual fields starting from periphery and moving towards centre. Similarly, embolization from carotid artery or cardiac chambers may also result in TMOB. TMOB in such a situation is not associated with headache or pain in the eye, unless patient has suffered from carotid artery dissection. You will get history of vascular risk factors such as diabetes (DM), hypertension (HT), coronary artery disease (CAD), smoking, chronic alcohol intake in such patients. TMOB may be a warning sign of impending ICA stroke. Carotid Doppler or preferably CT angiogram (CTA) should be done immediately to rule out carotid artery stenosis and managed on emergency basis.

Occasionally angle closure glaucoma, optic disc drusen (calcific deposit on optic nerve head), retinal detachment or retinal migraine may present as TMOB. Transient worsening of vision may be seen on exposure to heat or after exercise in patients suffering from demyelinating optic neuropathy (Uhthoff's sign).

Carotid doppler was found to be normal in our patient.

✍ Key Learning Point
Urgent carotid doppler or CTA should be done in case of TMOB.

Our patient comes back after 2 weeks with complains of headache, more severe on left fronto-temporal region for last 3–4 days associated with blurring of vision left eye. There is no pain on eye movements. He is not a known case of diabetes, hypertension or any other chronic ailment.

How to approach monocular or binocular blindness?

First step in a patient who presents with loss of vision is to confirm whether it is monocular or binocular. Even if patient gives history of monocular symptom, always check vision in contralateral eye, as patient may not perceive defect in both eyes. You should also rule out refractory error at this point by doing pinhole test—vision improves remarkably if patient looks through a small hole, confirming refractory error is the cause of blurred vision. Visual acuity is affected only when lesion is in chiasm or anterior to it. Retrochiasmatic lesions do not affect visual acuity unless they are bilateral.

Key Learning Point

Pinhole test helps in ruling out refractory error.

Before proceeding further, let us understand the anatomy of visual pathways. Nasal half of retina carries information from temporal field while temporal part of retina carries visual information from nasal half of visual field (Fig. 1.1).

Visual information from retina is then carried in optic nerve which is seen as optic disc on fundus examination with the help of ophthalmoscope. This optic nerve head is not myelinated and branches of central retinal artery are seen emerging through this disc which is called optic cup. This optic disc corresponds to physiological blind spot. Macula lies temporal to optic nerve head and is responsible for central vision (Fig. 1.2).

Nasal fibres carrying visual information from temporal fields cross at optic chiasm, while temporal fibres responsible for nasal field pass uncrossed and both continue as optic tract. Optic tracts terminate at lateral geniculate body from where 10% of

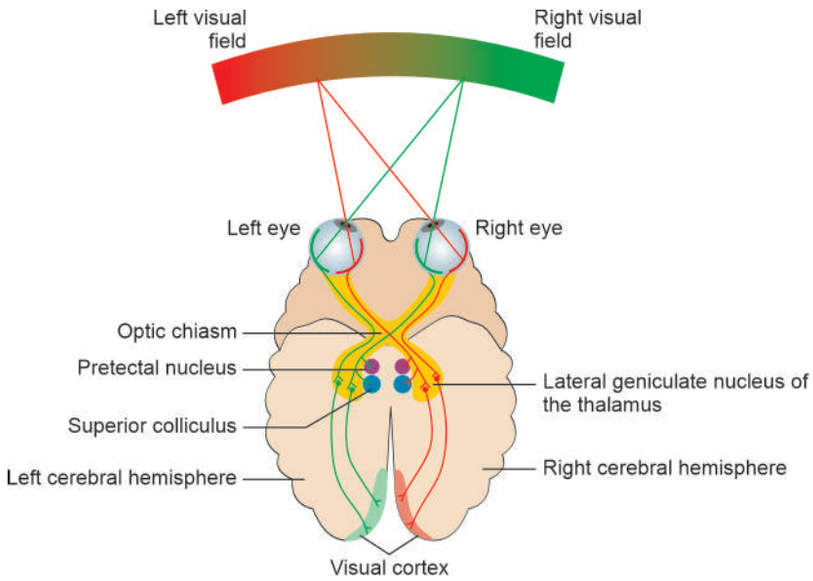


Fig. 1:1: Visual pathway

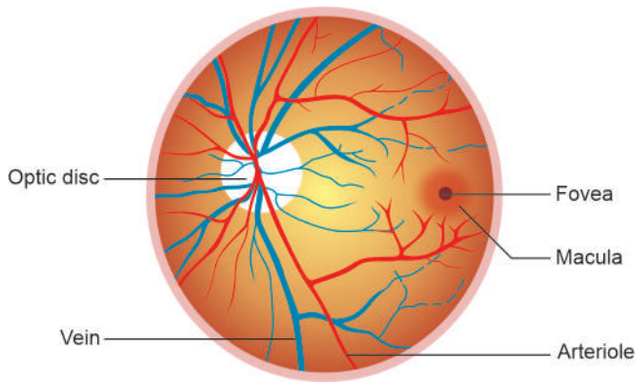


Fig. 1.2: Optic disc and macula (temporal to the disc)

fibres carrying information for light reflex continue to midbrain and terminate at bilateral pretectal nucleus (Fig. 1.1) Both pretectal nucleus in turn are connected to single Edinger-Westphal nucleus. From here, parasympathetic pupillo-constrictor fibres travel in 3rd nerve bilaterally and supply iris. This is why both pupils constrict in response to light thrown on one eye (Fig. 1.3).

Remaining 90% of the fibres continue as optic radiation. Inferior part of optic radiation, which carries visual information from ipsilateral upper nasal and contralateral upper temporal field, travel in temporal lobe. These fibres travel anteriorly in temporal lobe to form Meyer's loop before they continue posteriorly to occipital lobe. Superior part of optic radiation travels in parietal lobe and carries information from ipsilateral lower nasal and contralateral temporal visual field. These fibres terminate in occipital lobe around calcarine fissure in striate cortex. Arrangement of fibres in occipital lobe is such that macula is represented in occipital pole, upper bank of striate cortex carries information from contralateral inferior field while inferior lip controls information from contralateral superior visual field. Blood supply of visual cortex is derived from branches of middle and posterior cerebral arteries. Arrangement of this supply is such that macula gets blood supply from both the arteries.

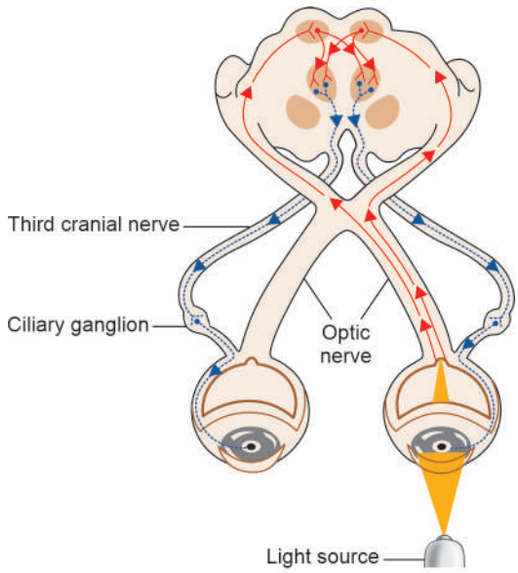


Fig. 1.3: Pathway for light reflex

Confrontation test is the simplest bedside test by which you can check whether problem is monocular or binocular. While doing confrontation test, patient should be at one arm distance from you. In this way, any visual stimulus presented by you will be at equal distance from you as well as from patient (Fig. 1.4).

When patient is suffering from central loss of vision (central or centrocecal scotoma), his optic nerve (or macula) is affected. While all causes of monocular blindness are restricted to prechiasmatic lesions, exception to this rule is "Junctional scotoma". Always check contralateral field because if lesion is at the junction of optic nerve and chiasm, you will find associated contralateral superior quadrantanopia.

The explanation that a junctional scotoma is caused by damage to a specialized "Wilbrand's knee" is considered an anatomical misconception in modern neuro-ophthalmology. In fact, the junctional scotoma of Traquair occurs because a tumor in the anterior sellar region simultaneously compresses the ipsilateral optic nerve (causing the scotoma) and the anterior part of the optic chiasm (causing the contralateral field defect (Figs 1.5A and B).

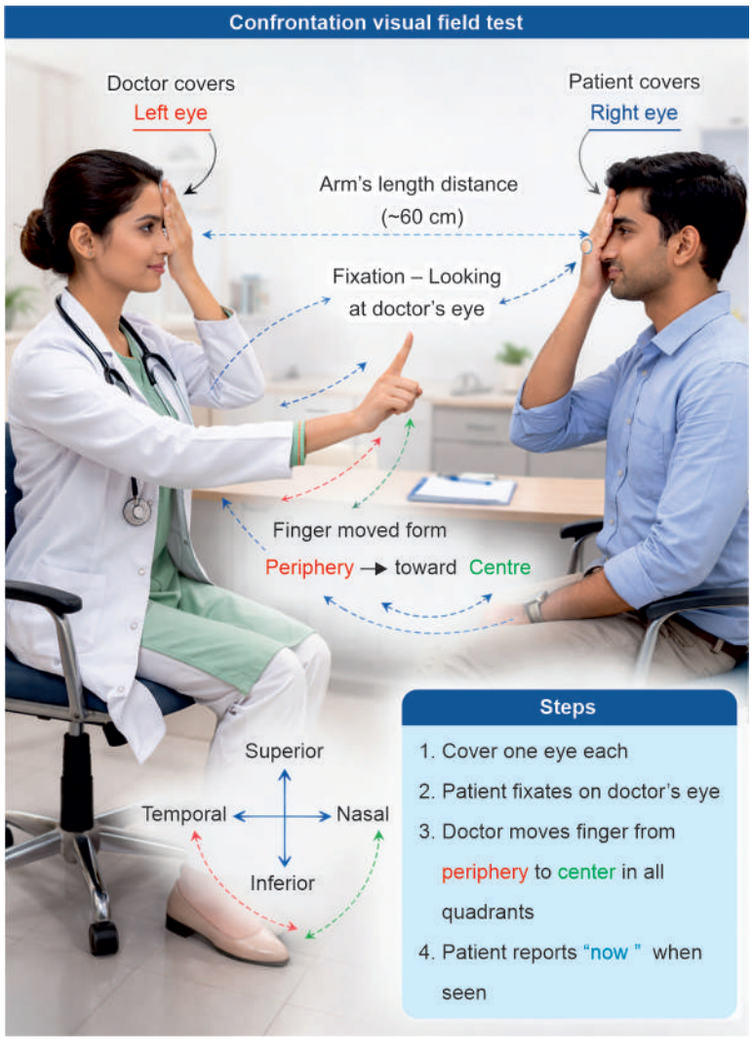
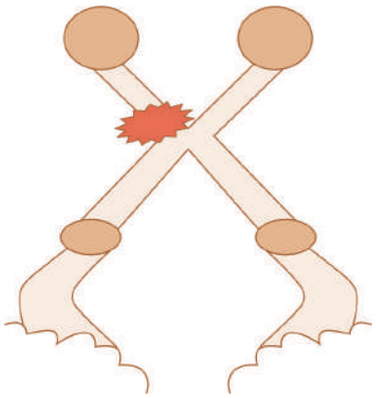


Fig. 1.4: Confrontation test



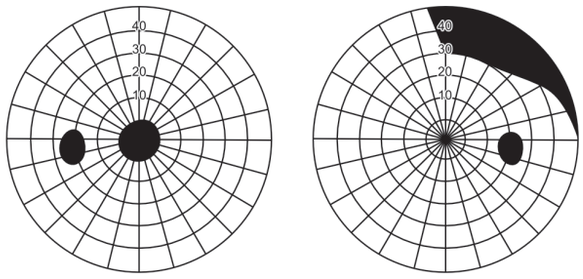


Fig. 1.5B: Junctional scotoma

Fig. 1.5A: Site of lesion in junctional scotoma

If patient can see only upper or lower half of field (defect in horizontal meridian), then lesion is prechiasmatic. This horizontal altitudinal defect is characteristic of anterior ischemic optic neuropathy (AION, Fig. 1.6).

If patient can see only one half of visual field (defect in vertical meridian) then lesion is postchiasmatic in contralateral optic tract or cortex involving optic radiations causing homonymous hemianopia (Fig. 1.7).

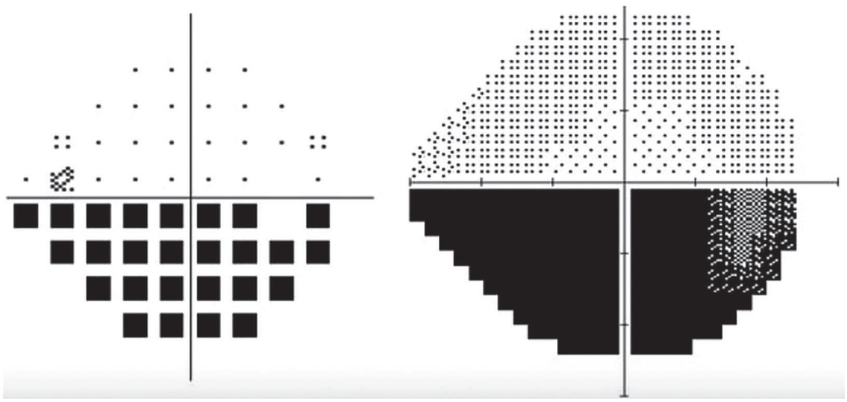


Fig. 1.6: Horizontal altitudinal field defect seen in AION

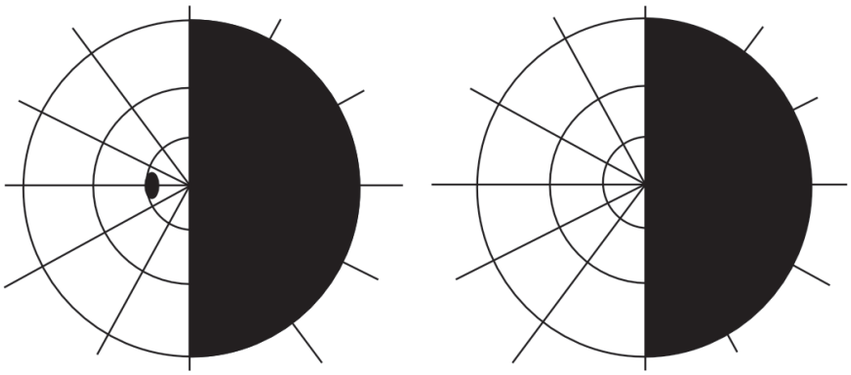


Fig. 1.7: Homonymous hemianopia because of lesion at contralateral optic tract, optic radiation or visual cortex

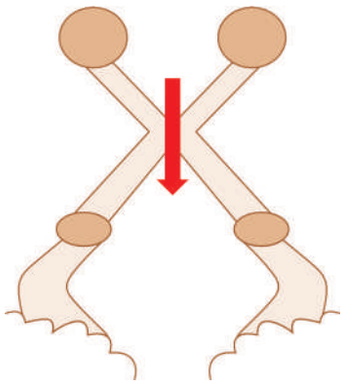


Fig. 1.8: Lesion at optic chiasm

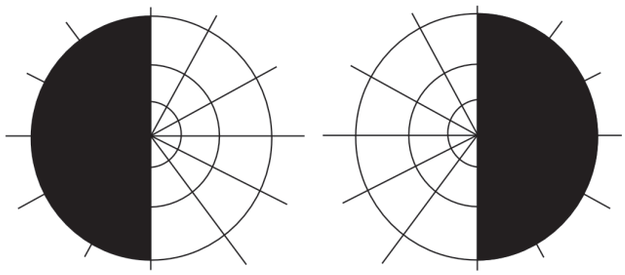


Fig. 1.9: Bitemporal hemianopia because of lesion at optic chiasm

With both eyes open, if patient cannot see both temporal fields but can see central field, then he is suffering from bitemporal hemianopia and lesion is at optic chiasm (pituitary adenoma or suprasellar mass causing compression of chiasm) as crossing nasal fibres carrying information from bilateral temporal fields are affected (Figs 1.8 and 1.9).

Key Learning Points

- Prechiasmatic pathology causes monocular loss of vision.
- Always check vision in both eyes even if patient gives history of monocular loss of vision, otherwise you can miss junctional scotoma.
- In case of horizontal altitudinal defect always think of AION.
- Postchiasmatic lesion causes vertical meridian defect.

Confrontation test confirms that he is suffering from monocular blindness. What are the ocular causes of sudden monocular blindness?

Once monocular blindness is confirmed, next step is to confirm whether problem is with optic nerve and its central connections or it is localised to eye itself. When patient cannot see out and when you cannot see inside patient's eye, then possibly patient is suffering from local eye pathology like corneal opacity, cataract, vitreous hemorrhage or retinal detachment, etc.

Any ocular pathology obstructing the visual pathways can result in monocular blindness. Besides trauma, vitreous hemorrhage and retinal detachment are two local pathologies, which can cause sudden loss of vision in one eye. In case of vitreous hemorrhage, you cannot see the disc and media appears greenish. Such condition may be seen in association with subarachnoid hemorrhage (Terson's syndrome). When retina is detached from retinal pigment epithelium, one can complain of floaters, flashes of lights or shower of dots followed by a grey curtain that comes over from the periphery causing loss of vision. Central serous retinopathy (CSR) is another situation where central vision is affected acutely. Because pathology is at macula in case of CSR, optic disc is normal on fundoscopy. Acute angle closure glaucoma also presents with monocular diminution of vision. In this situation patient complains of sudden onset headache, vomiting, mild congestion of one eye, mid dilated but reacting pupil, raised intraocular pressure and normal fundus examination.

Key Learning Point

Ocular causes of acute onset monocular blindness are vitreous hemorrhage, retinal detachment, CSR, angle closure glaucoma.

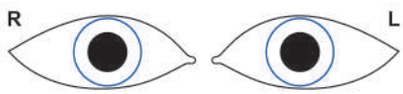
On fundoscopic examination media is clear. Is he suffering from Optic Neuropathy (ON)?

Now check for pupillary light reflex—both direct and consensual. For checking light reflex, patient should look at distance to avoid near reflex. Room light should be dull, not very bright so that it does not cause pupillary constriction and at the same time allow you to appreciate pupillary reaction. Check for speed and amplitude of constriction of pupil on both the sides one by one. Then check consensual reflex-constriction of contralateral pupil when light is thrown to ipsilateral eye. Next step is to do swinging light test- light is thrown to one eye and pupillary constriction is checked in both the eyes (better if a thin film is placed between the two eyes so that light does not reach directly to contralateral eye). Wait for 3 seconds before you move light to opposite eye and check direct as well as consensual reflex.

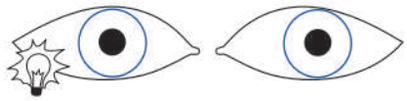
When light is thrown on one eye and if optic nerve is affected (left eye in our case), pupil in affected (left) eye will start dilating instead of constriction. Similarly, no consensual pupillary constriction will be seen in contralateral (right) eye and it will also be seen as dilating (Fig. 1.10).

This happens because afferent limb of light reflex (left optic nerve) is unable to carry the information to pretectal nucleus and Edinger-Westphal nucleus. Hence the efferent information for pupillary constriction via third nerve to ipsilateral and contralateral eyes is not generated. This is called "relative afferent pupillary defect"

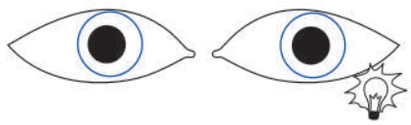
(1) Begin with darkroom, bright pen light, and patient fixated at distant object



(2) **Shine light into right (R) eye**. Both pupils constrict



(3) **Swing light to left (L) affected eye**. Instead of pupil constriction, both pupils will dilate.



(4) **Swing light back to right (normal) eye**. Both pupils constriction

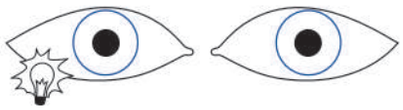


Fig. 1:10: Relative afferent pupillary defect (RAPD)

(RAPD). If pupil is affected and RAPD is present (as confirmed in this case), lesion is anterior to optic chiasm (left optic nerve in this case). As nerve fibres arise in the retinal layer, major retinal damage like central retinal artery occlusion (CRAO) and retinal detachment also cause RAPD. But pupillary reflex is normal and RAPD is not seen in patients with central serous retinopathy (differential diagnosis retrobulbar optic neuritis—see later). If patient cannot see with one eye because of local ocular pathology and pupil is distorted because of previous cataract surgery, you can still confirm integrity of ipsilateral optic nerve by checking consensual light reflex.

Since nasal fibres cross to opposite side in optic chiasm and travel in contralateral optic tract, lesion in optic tract (before light reflex fibres leave the tract) can rarely cause loss of light reflex in contralateral eye, but visual field defect will be bilateral (homonymous hemianopia). Because light fibres leave visual pathways and join lateral geniculate body, pupil is not affected in lesions posterior to lateral geniculate body (cortical lesions).

Key Learning Point

RAPD localises lesion anterior to optic chiasm.

Examination reveals vision restricted to hand movements in left eye involving inferior half of visual field (with sparing of superior half) and left pupillary light reflex is lost with RAPD. Can this be CRAO or BRAO?

Sudden painless loss of vision in one eye which develops over seconds or minutes and does not recover suggests possibility of CRAO. Ophthalmic artery is the branch of ICA. Its main branch is central retinal artery (CRA) which traverses within the optic nerve, emerges at the nerve head and divides in superior and inferior branches. These branches further divide in nasal and temporal branches and supply inner two third of retina. Central retinal artery does not supply blood to optic nerve (Fig. 1.11).

When only one branch of retinal artery is involved, this is called branch retinal artery occlusion (BRAO). It can cause loss of vision in affected visual field-superior, inferior, temporal or nasal (Figs 1.12A to C). AION and CRAO/BRAO both cause painless loss of vision with loss of light reflex and RAPD. Clinical differences among AION, CRAO, and BRAO are presented in Table 1.1.

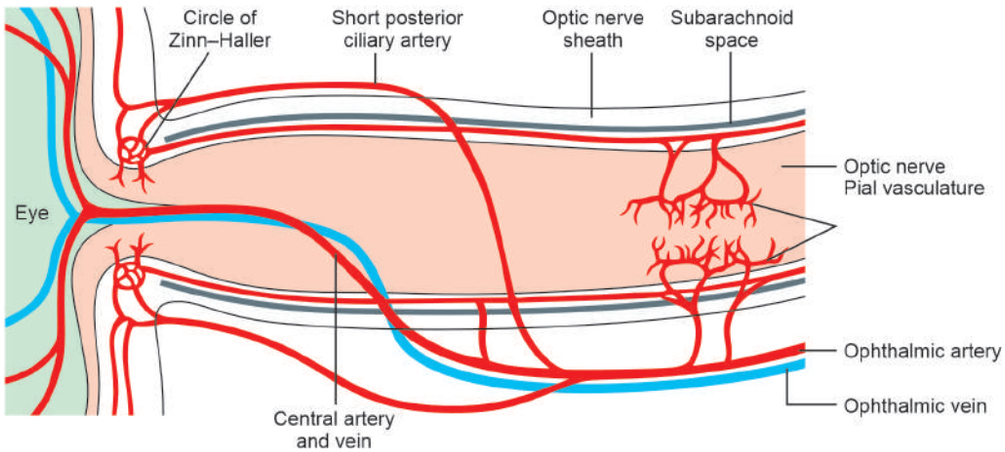


Fig. 1.11: Ophthalmic artery and its branches

Table 1.1: Differences in AION, CRAO and BRAO			
	AION	***CRAO***	***BRAO***
Onset	Slowly over 3–4 days	Over seconds to minutes	Over seconds to minutes
Field defect	Horizontal altitudinal defect (inferior most common)	Complete monocular loss of vision	Loss of vision in superior or inferior quadrant

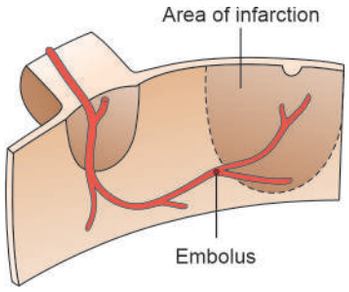


Fig. 1.12A: Embolus at bifurcation of blood vessel

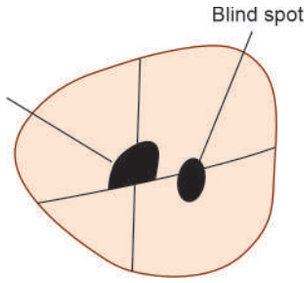


Fig. 1.12B: Inferior branch occlusion causes upper field defect as these vessels supply up to horizontal meridian

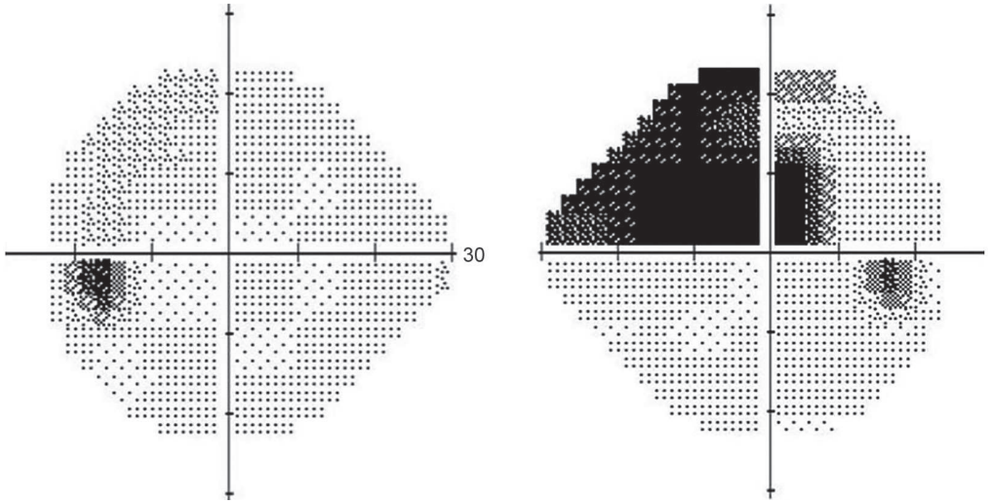


Fig. 1.12C: BRAO causing superior field defect

Let us see Fundus with Ophthalmoscope

If central vision is affected, there is pain on eye movements, pupillary light reflex is lost, RAPD is seen and optic nerve head appears normal on fundus examination, it can be because of retrobulbar neuritis (RON). More than 70% of the patients with demyelinating optic neuropathy have normal disc because pathology is behind the nerve head (patient do not see anything outside and you do not see anything abnormal inside). It takes around 4–6 weeks for optic atrophy to set in, it is the time when you will see pale optic nerve head on fundus examination.

Central vision is also affected in central serous retinopathy (CSR), but optic disc is normal, pupillary reflex is normal and RAPD is not seen.

In case of CRAO or BRAO, retina appears pale or white, thread like non-perfused capillaries are seen which are reduced in number and cherry red spot is seen at fovea.

Cherry red spot represents underlying perfused macula by choroidal circulation which is supplied by posterior ciliary arteries. Pale yellow bright cholesterol emboli may be seen in one of the vessels, "the Hollenhorst plaque" (Fig. 1.13).

Central retinal vein occlusion (CRVO) is another cause of sudden moncular blindness. On fundoscopy you will see dilated tortuous veins, cotton-wool spots and retinal haemorrhages (Fig. 1.14).

Our patient is 60 years of age, not suffering from any vascular risk factors and is complaining of headache along with TMOB followed by loss of vision in one eye. Because inferior half of visual field is affected, suggesting altitudinal defect, diagnosis is anterior ischemic optic neuropathy (AION). Since it is associated with headache and patient is not suffering from any atherosclerotic vascular risk factors then possibly he is suffering from arteritic AION (A-AION). Fundus examination confirmed disc oedema on left side. It is seen in elderly patients (>60 years of age) suffering from temporal arteritis, which is also known as giant cell arteritis. Inflammation of vessel wall results in reduced blood supply to optic nerve head and retina leading to loss

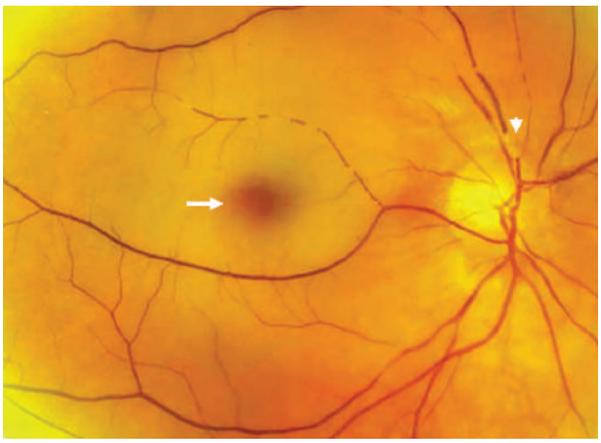


Fig. 1.13: Hollenhorst plaques (arrow head). White arrow showing cherry red spot

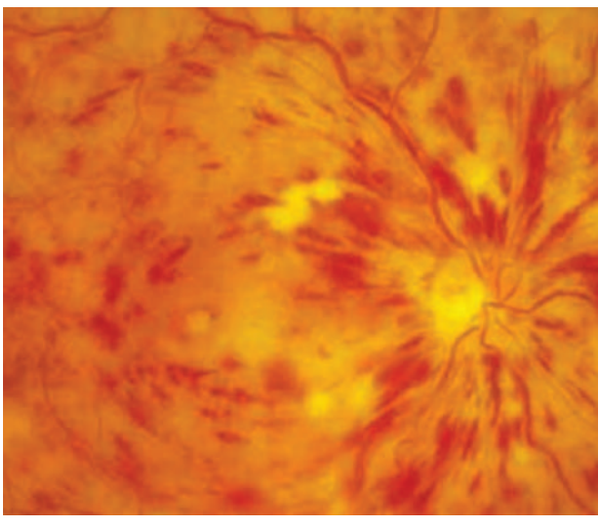


Fig. 1.14: Central retinal vein occlusion

of vision. Patient may give history of jaw claudication (pain in jaw while chewing) or body pains (polymyalgia rheumatica). On examination ipsilateral superficial temporal artery (STA) pulsations are absent and on palpation it is felt like a cord or thread.

Temporal artery ultrasound may confirm hypoechoic vessel wall thickening (halo sign), non-compressibility of vessel wall or occlusion. ESR, CRP and platelet counts are raised in these patients. If ESR is more than half of age in males and more than half of age plus 10 in female patients, it is considered abnormal. Temporal artery biopsy can confirm the diagnosis. Because of skip lesions, sensitivity of biopsy is low in these cases. Hence, it is proposed that biopsy should include at least 2 cm (preferably 3–6 cm) long segment of the vessel and sample should be taken from both the sides. If suspicion is strong, patient should be put on parenteral high dose steroids even without confirmation. Failure of which may result in permanent irreversible loss of vision. Histopathological changes can be seen even for long after putting patient on steroids, hence biopsy can be done even after treatment is started.

Non-arteritic AION (NA-AION)

It is important to differentiate A-AION from NA-AION. Classical feature of this condition is "disc at risk". Those with a crowded optic disc (cup-to-disc ratio <0.3) are at risk of developing NA-AION. This type of disc is also seen in fellow eye, a finding which raises suspicion against A-AION. These patients are suffering from one or more atherosclerotic vascular risk factors. If an elderly patient who is suffering from risk factors like DM, HT, etc., presents with loss of vision in one eye and he does not complain of headache or eye pain, then diagnosis is possibly NA-AION. STA is not affected in these patients and prognosis for recovery is poor.

Any elderly patient who presents with altitudinal field defect involving horizontal meridian and disc appears normal, consider possibility of posterior ischemic optic neuropathy (PION). Like AION, it can also be arteritic or non-arteritic in origin. Another important etiological factor is prolonged intraoperative hypotension resulting in perioperative PION.

Biopsy confirmed the diagnosis of temporal arteritis and he responded to steroid treatment. Oral steroids should be continued at least for 12–18 months in tapering doses. In refractory cases, tocilizumab can be considered.

How to differentiate AION from demyelinating ON?

Since optic disc is swollen in both the conditions one should know how to differentiate between the two. Disc oedema in AION is associated with surrounding hemorrhage while in demyelinating ON, disc edema is seen without hemorrhage. Demyelinating ON is disease of young adults, females are affected more than males. It is usually associated with painful eye movements. Other findings like absent pupillary reflex and presence of RAPD are also seen in demyelinating ON. Hence if a young person (<50 years) who is not suffering from vascular risk factors, presents with monocular painful loss of vision over 3–4 days, diagnosis is probably demyelinating ON. On MRI optic nerve appears hyperintense on T2 weighted images and may show contrast enhancement, which is not true for AION.

Other Causes of Optic Neuropathy

Occasionally monocular blindness is detected incidentally on routine examination or otherwise and mis-interpreted as sudden in onset. When you see optic nerve

head is swollen, vision in one eye is severely affected, light reflex is absent and RAPD is present; a diagnosis of optic neuritis is made, but MRI brain fails to reveal any pathology and patient does not show any response to high dose intravenous steroids. Now what is the diagnosis? Please do remember, visual loss in slowly growing optic nerve glioma may be detected incidentally. Neuroimaging is normal in such a situation unless you do MRI brain with orbital cuts with contrast and fat suppression images. Similarly, optic sheath meningioma gives appearance of tram-track on contrast images.

Key Learning Points

- A-AION is seen in elderly patients (>60 years) while demyelinating ON is a disease of young individuals.
- Headache is the presenting feature of A-AION, pain on eye movements is a feature seen in demyelination ON.
- Systemic features like jaw claudication, body pains, raised ESR/CRP are seen in A-AION.
- Horizontal altitudinal defect is seen in A-AION while central and centrocaecal scotomas are seen in demyelinating ON.
- NA-AION is seen in elderly patients with atherosclerotic risk factors. "disc at risk" in both eyes is the characteristic finding in such cases.

Disc is swollen in papilledema also. How to differentiate it from optic neuritis?

Primary complaint in ON is loss of vision and field defects are either central scotoma, centrocecal scotoma or paracentral scotoma (Figs 1.15A to C).

On perimetry, enlargement of blind spot and concentric constriction of visual fields are the two major early findings in papilledema (Figs 1.16A and B).

Blindness is transient and bilateral in cases of papilledema (known as transient visual obscurations or TVOs). Rapidly progressive loss of vision in one eye over 3–4 days is the presenting complaint in cases of demyelinating ON. Eye movements are painful in demyelinating ON, while headache is usually the primary complaint in patients with papilledema. Exception to this rule is fulminant idiopathic intracranial hypertension (IIH), which is rarely seen in young obese patients who present with sudden bilateral diminution of vision associated with headache.

Loss of pupillary light reflex and presence of RAPD are seen in ON not in papilledema, unless optic atrophy has set in. Papilledema is usually bilateral, exception being Foster-Kennedy syndrome where basi-frontal tumor causes compression of ipsilateral optic nerve with resultant atrophy and contralateral papilledema because of raised intracranial pressure. When there is optic atrophy in one eye because of previous

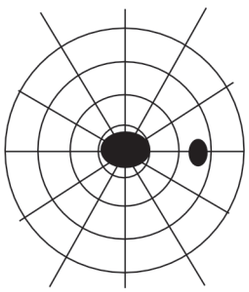


Fig. 1.15A: Central scotoma

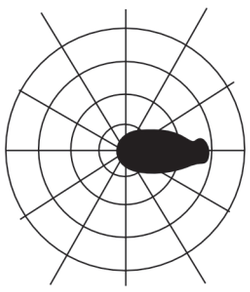


Fig. 1.15B: Centrocecal scotoma

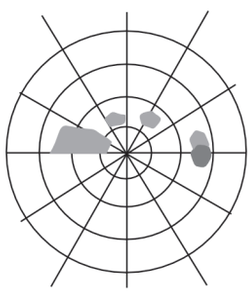


Fig. 1.15C: Paracentral scotoma

Fig. 1.16A: Enlargement of blind spot

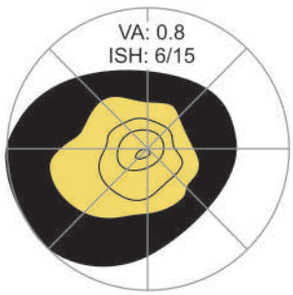


Fig. 1.16B: Concentric constriction of visual field

insult and recent disc edema in other eye because of fresh optic neuritis, it is called pseudo-Foster Kennedy syndrome. Visual evoked potential (VEP) settles the issue in any case where P100 latency is delayed in case of ON, while it is normal in case of papilledema. Optical coherence tomography and fluorescein angiography are other ways of confirming papilledema.

Key Learning Points

- Papilledema presents with headache and TVOs. ON presents with pain on eye movements and monocular loss of vision.
- Papilledema is bilateral and ON is usually unilateral.
- Concentric constriction of visual fields with enlarged blind spot is seen on field charting in papilledema. Central or centrocecal scotoma is seen in ON.
- P100 latency on VEP is normal in papilledema but it is prolonged in ON.

Binocular Loss of Vision

Bilateral simultaneous or sequential optic neuritis is one of the important causes of bilateral loss of vision and is seen in neuromyelitis optica (NMO) and myelin oligodendrocyte glycoprotein (MOG) related disorders. While NMO affects long segment of optic nerve posteriorly along with involvement of optic chiasma, MOG involves long segment anterior optic nerve along with enhancement of optic nerve sheath (optic peri neuritis). MOG optic neuritis can present as chronic relapsing inflammatory optic neuropathy (CRION) which responds to steroids and these patients are steroid dependent. There are specific diagnostic criteria, clinical and radiological pattern of these demyelinating disorders. We will not go into details but one should rule out these disorders if patient presents with bilateral simultaneous/sequential optic neuropathy.

Another rare condition which presents as acute or subacute bilateral sequential (or simultaneous) painless loss of vision is Leber hereditary optic neuropathy (LHON). It is maternally inherited mitochondrial disorder which typically affects males between 15–35 years of age. Any young male patient who presents with acute or subacute bilateral optic neuropathy and fails to respond to steroid therapy, consider possibility of LHON.

Compression of chiasm from above or down can cause bitemporal hemianopia. Lesion of optic tract result in contralateral hemianopia where defect is incongruous. Congruity means size, shape and slope of defect in both the eyes. If size, shape and slope of visual field defect is different in both the eyes, it is called incongruous. Incongruity is the characteristic of tract lesions because fibres affected are widely placed (Fig. 1.17).

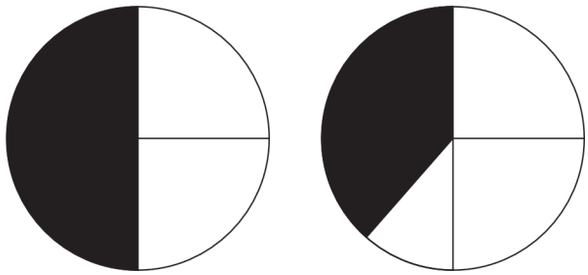


Fig. 1.17: Incongruous field defects seen in optic tract lesion

This is in contrast to cortical lesions where field defect is congruous (Fig. 1.18).

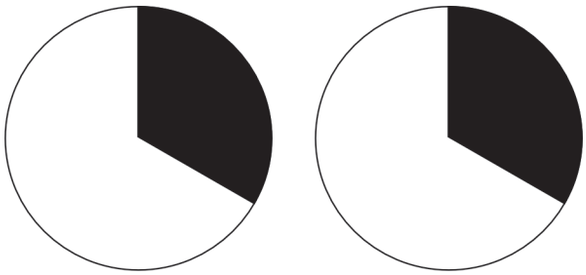


Fig. 1.18: Congruous field defects seen in optic radiation lesion

Cortical Visual Loss

Cortical visual loss is associated with certain features like visual inattention, hemineglect, prosopagnosia (inability to recognise faces), geographic disorientation (inability to recognise places and paths), dressing and constructional apraxia if defect is in right parietal lobe. It may be associated with Gerstman's syndrome [acalculia (inability to calculate), agraphia (inability to write), finger agnosia (inability to recognise fingers), right left confusion], if lesion is in left parietal lobe. Parietal lobe lesion may cause contralateral inferior homonymous quadrantanopia if lesion is restricted to superior part of optic radiations (Fig. 1.19).

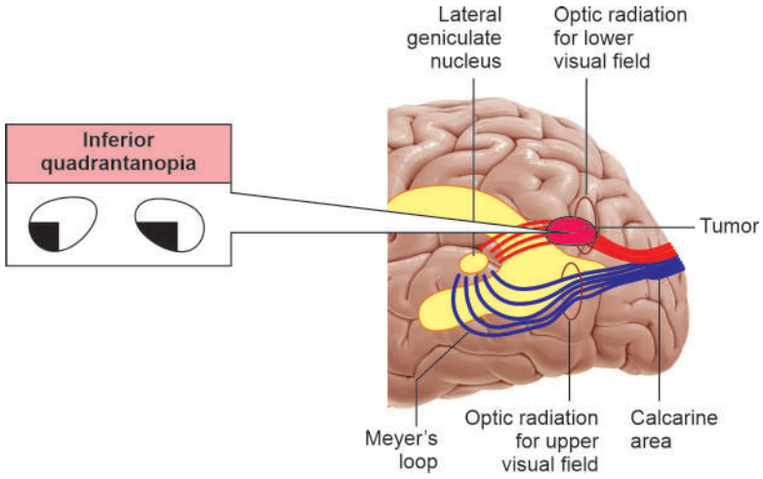


Fig. 1.19: Contralateral homonymous inferior quadrantanopia in parietal lesion

Anterior temporal lobe affection may cause involvement of Meyer's loop (inferior nasal fibres loop anteriorly) with resultant contralateral superior homonymous quadrantanopia (Fig. 1.20).

Riddoch's phenomena is another characteristic of cortical lesion where patient can appreciate moving object but can miss static object in hemianopic field. With left occipital lobe lesions when splenium of corpus callosum is involved, alexia without agraphia (patient cannot read what he has written) may be observed. It is a type of disconnection syndrome where patient suffers from right homonymous hemianopia and is unable to retrieve lexical visual information processed in the intact right occipital lobe (which is the only source of getting visual information from visual fields). Contralateral homonymous hemianopia in all these situations may be associated with hemiparesis contralateral to the side of lesion. If both occipital lobes are affected (basilar artery syndrome), there may be complete loss of vision with macular sparing (tubular vision). This happens because macular region is represented at occipital lobe tip, which has got dual blood supply both from middle and posterior cerebral arteries (Fig. 1.21).

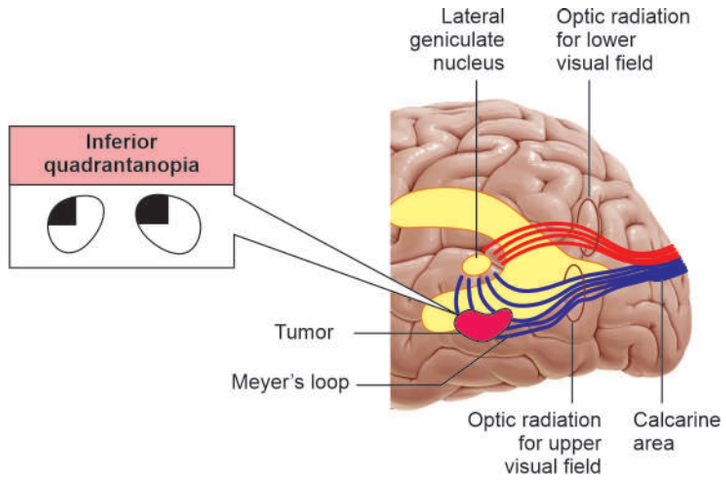


Fig. 1.20: Contralateral homonymous superior quadrantanopia in temporal lesion affecting Meyer's loop

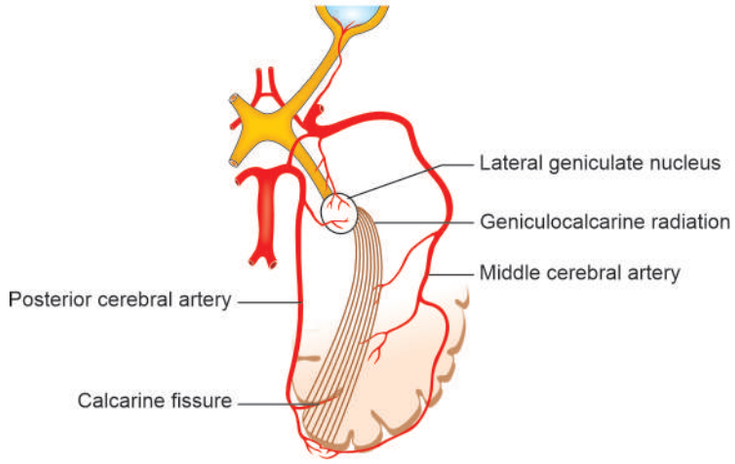


Fig. 1.21: Occipital pole is supplied by both middle and posterior cerebral arteries

Similarly, PCA strokes can spare anterior striate cortex, the area that controls vision in peripheral temporal crescent, hence it can cause contralateral homonymous hemianopia with sparing of temporal crescent. If only anterior striate cortex is affected (occlusion of branch of MCA), contralateral monocular temporal crescent syndrome ('half moon syndrome') results (exception to the rule that monocular defect is caused by prechiasmatic pathology). Few other features of cortical blindness are Anton syndrome (denial of blindness) and Balint syndrome (optic ataxia—inability to reach for object under visual guidance, oculomotor apraxia—a defect in voluntary horizontal eye movements results in inability to initiate saccades and simultanagnosia—inability to recognise whole picture despite ability to perceive its parts). Palinopsia (persistence of image even after removal of object), polyopia (multiple images of single object) and optic allesthesia (abnormal orientation of object) can also result from cortical lesions.

Posterior reversible encephalopathy syndrome and reversible vasoconstriction syndrome can also cause cortical blindness in specific clinical situations.

How to Confirm that it is Functional Blindness

Besides menace reflex (sudden unexpected visual threat) which should cause reflex blinking, you can check vision by number of bedside tests.

- Simplest test is to ask the patient to give his signature on a piece of paper. If he fails to do so, possibility is that he is not suffering from true blindness. Because signature requires proprioception, not vision.
- Similarly ask the patient to touch tip of both index fingers. This again requires proprioception, not vision. Hysterically blind patient will fail to do so.
- Optokinetic nystagmus is seen if a striped paper is moved in front of patient's eye, if patient is not blind.
- If you rock a big mirror to and fro in front of patient's eyes, eye will move up and down with it. In blind patients this phenomena will not be appreciated.
- If visual field is restricted to same area even with increasing the distance (tunnel vision), there is strong possibility that patient is malingering because with increasing distance, visual field area also widens (funnel vision, Fig. 1.22).

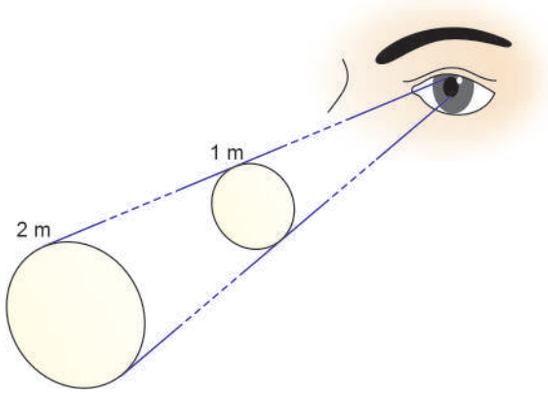


Fig. 1.22: Funnel vision, with increasing distance visual field area widens in case of true blindness

Schematic Approach to Loss of Vision