Case Report

A rare case of cortical blindness - A case report

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Abstract
Cortical blindness refers to loss of vision caused by bilateral occipital lobe lesions with presence of intact anterior visual pathway. Neurological visual impairment encompasses a broad spectrum of conditions. These include cerebral visual impairment, visual neglect, visual agnosia, various visual perceptual disorders, homonymous hemianopia, lack of facial recognition, delayed visual development and cortical blindness. Cortical blindness refers to loss of vision produced by lesions affecting geniculocalcarine visual pathways. Complete Cortical blindness is much less common than incomplete blindness. Cortical blindness is mostly due to cerebrovascular accident but also reported in pregnancy induced hypertensive encephalopathy, trauma, as result of MS exacerbation, post cardiac surgery, obstetric hemorrhage, MELAS, cerebral angiography, adrenoleukodystrophy, CNS angiitis and HIV-related progressive multifocal leukoencephalopathy. In this case report, we present a case of patient with cortical blindness due to bilateral occipital Infarcts.

Key words
Cortical blindness, Bilateral occipital infarcts.

Introduction
The term “Cortical Blindness” describes the partial or complete loss of vision resulting from a brain lesion. In this disorder there are no ophthalmological causes and with normal pupillary light reflexes. With this type of visual impairment the eyes can be fully intact, but the visual information cannot be transmitted to the brain regions in which these are processed into meaningful visual input such as brightness-contrasts, colors, objects, faces. They are “blind” to the visual information that is not transmitted. Cortical Blindness can however also affect vision in total; this requires damage to both sides of the brain, usually due to bilateral lesions of the striate cortex in the occipital lobes [1]. Cortical
blindness is a part of cerebral blindness which is defined as loss of vision secondary to damage to the visual pathways posterior to the lateral geniculate nuclei [2]. With cortical blindness in both halves of the visual field a person is really completely blind, he/she cannot consciously process visual input any longer, cannot identify or describe objects, cannot recognize faces, cannot read a text or reach for an item.

In the year 1895, Austrian neuropsychiatrist Gabriel Anton described patients with bilateral occipital lobe lesions, who were completely blind but were unaware of their blindness leading to confabulation [3].

Etiology
Cortical blindness can affect both children and adults. In children, common causes include; Congenital abnormalities of the occipital lobe, cardiac arrest, status epilepticus, hypoxia or perinatal asphyxia, cerebral infarction, meningitis, encephalitis, subacute sclerosing leukoencephalitis, hypoglycemia, uremia, hydrocephalus, shunt malfunction, head trauma, cardiac surgery, cerebral or vertebral angiography , drugs (i.e., cyclosporin A and steroids), acute carbon monoxide poisoning, and the ictal state in occipital lobe epilepsy or postictal phenomenon [4].

In adults, it is seen in lesions of the primary visual cortex of the occipital lobes secondary to multiple disorders including; Stroke, Cardiac embolism, Head trauma, Occipital lobe epilepsy, Hyponatremia, Severe hypoglycaemia, Creutzfeldt-Jacob disease, Infection e.g. HIV, Eclampsia, MELAS (mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes), cardiac surgery, cerebral or vertebral angiography, IV contrast medium. Rarely, transient cortical blindness can be caused by Infective endocarditis or Hypertensive encephalopathy, Posterior reversible encephalopathy syndrome (PRES) [5, 6, 7].

Epidemiology
Exact epidemiological data is not available. Studies have shown a high incidence of cortical blindness in patients of cerebral stroke in the range of 20 to 57% [8]. Cerebrovascular disease was the most common cause according to various studies, followed by surgery, particularly cardiac surgery, and cerebral angiography [9].

Pathophysiology
Knowledge of the visual pathway is important for the localization of lesions in cortical blindness. In cortical blindness, lesion lies in the striate cortex, and often the nearby areas of the brain are also involved.

Visual pathway
The optic disc is the starting point of the optic nerve. The optic disc does not contain any visual receptors. It is the physiological blind spot. Fibers from the temporal Half of retina are situated in the lateral half i.e. temporal half of the optic nerve while fibers from the nasal half of retina are situated in the medial half i.e. nasal half. Similarly, upper retinal fibers are present superiorly and lower fibers are present inferiorly in the optic nerve. The optic nerve extends from the retina to the optic chiasm and the length is about 5 cm. It is conventionally divided into four portions- intraocular, intraorbital, intracanalicular, and intracranial.

The optic nerve is covered by the layers of meninges. The intraocular part is the optic disc wherefrom the intraorbital portion starts leading to the intracanalicular portion as the nerve traverses the optic canal. Then the two optic nerve exit from the optic canals and form the optic chiasm where temporal hemiretinal fibers continue ipsilaterally and nasal hemiretinal fibers decussate and join the opposite optic tract. The tracts extend from the chiasm to the lateral geniculate body (LGB). Afferent fibers from the pupil leave the tract just anterior to the LGB. The visual afferents synapse in the LGB and second-order neuron starts as geniculocalcarine pathways (optic radiation) and terminate in the calcarine cortex of the occipital lobe. The primary visual cortex (V1) lies in the
Brodmann’s Area 17. Area 18 (V2) also known as the parastriate or parareceptive area, receives and interprets impulses from Area 17. The peristratiae or perireceptive cortex, Area 19 (V3, V4, V5) has connections with Areas 17, 18, and with other portions of cortex.

The anterior choroidal artery and thalamoperforators from the posterior cerebral artery (PCA) supply the optic tract. Optic radiation is supplied by the middle cerebral artery (MCA) and the occipital lobe is primarily supplied by the PCA. The occipital lobe also receives supply from the MCA.

History and physical examination
To make a proper diagnosis and to determine the aetiology of Cortical blindness, proper history should be elicited, like birth history, antenatal history, addictions, drug history, hypertension, diabetes, palpitation, fever and any history of head trauma. A complete neurological and ophthalmological examination should be done. An important thing to remember is that pupillary light reflex remains intact in cortical blindness, and extraocular movements will be normal. There is no relative afferent pupil defect (RAPD) in cortical blindness.

There may be clinical features depending on the location of the lesion. Lesion in temporal lobe causes acute disturbance in memory, particularly if it affects the dominant lobe. Posterior cerebral artery occlusion may produce visual hallucinations of brightly colored scenes and objects (peduncular hallucinosis) due to damage to the thalamus, brainstem [10]. Left-sided large PCA stroke can produce visual agnosia due to disconnection between language and visual systems whereas right-sided stroke can produce prosopagnosia due to involvement of the inferior occipital areas, fusiform gyrus, and the anterior temporal cortex [11].

Depending on the extent of the damage to V1, the loss of vision varies between a small scotoma about the size of the blind spot, a quadrantanopia, and full hemianopia. In the majority of cases, however, central vision, including the foveal representation, remains intact because of the blood supply of the occipital pole. The fovea is generally represented in the occipital pole and it receives its blood supply from the middle cerebral artery along with the branches of the posterior cerebral artery. Because of this dual blood supply, stroke causing the complete destruction of V1 is extremely rare. This preservation of central vision is very important for rehabilitation. Another peculiarity that can be seen in Cortical blindness patients is 'blind sight' where an individual can perceive coarse flickering movement in the blind field. This is due to preserved unconscious visual processing abilities in the individuals because of the heterogeneity of damage to V1. Other features of visual cortex lesion include Anton syndrome, Riddoch phenomenon, and formed visual hallucinations.

Anton syndrome: It is also known as visual anosognosia when a person cannot see but always denies the blindness even with clear evidence of blindness. These individuals often try to walk through the closed door or wall and in the process of denial they take help of confabulation. It occurs due to lesions in the V1.

Riddoch phenomenon: This is also known as statokinetic dissociation. Here patients can only perceive moving objects in the blind field, not the static ones [12]. Patients may not perceive color or details of the moving objects except the movement. This syndrome is often seen in lesions of the occipital lobe. The pathophysiology of Riddoch syndrome due to occipital lobe disease is thought to involve visual inputs reaching the V5 (motion processing cortex) bypassing the V1 area, leading to a conscious awareness of motion within a blind field.

Benson syndrome: Also known as posterior cortical atrophy, is a form of atypical Alzheimer’s disease [13]. It was first described by Dr. DF Benson in 1988 [14]. Here patient shows a decrease in visuospatial and
visuoperceptual capabilities but language, learning and cognition remain intact in the early stages [15]. Here the damage lies in the occipital cortex and associated areas of temporal and parietal lobe which can be viewed in MRI (magnetic resonance imaging) brain.

Posterior reversible encephalopathy syndrome (PRES): PRES presents with acute onset headache, seizures, altered consciousness, and visual disturbance [16]. It is usually seen in association with malignant hypertension, eclampsia. Typical MRI brain findings are of bilateral white-matter abnormalities in vascular watershed areas affecting mostly the occipital and parietal lobes [16, 17, 18]. With proper management of hypertension and other associated risk factors, PRES can be completely reversed including the MRI findings.

Balint syndrome: Austro-Hungarian neurologist and psychiatrist R Balint first described it [19]. It is characterized by simultanagnosia, oculomotor apraxia, and optic ataxia. Lesion lies in the bilateral parietal lobes and in some cases occipital lobe [20].

**Treatment and management**

Apart from standard management of the cause which in most cases is stroke, the major part of treatment is visual training and rehabilitation.

Three common modes of interventions are 1) restitution therapy, 2) compensation therapy, and 3) substitution therapy.

1) Restitution therapy is done to recover visual field deficits. It is like the perimetry [21]. Here, the patient detects multiple light spots on a black screen across blind and normal visual hemifield.

2) Compensation therapy acts by compensating for visual loss by saccadic eye movements [22]. It helps to capture visual stimuli that would otherwise fall onto the blind part of the visual field.

3) On the other hand, substitution therapy uses prism or other devices to project the visual stimulus from the blind side of the visual field to the normal one [22].

**Differential diagnoses**

Differential diagnoses of cortical blindness are: Hemineglect, Prosopagnosia, Simultagnosia, Malingering

**Prognosis**

Prognosis depends on the severity of the damage to the visual cortex. Extensive bilateral occipital lesions have a worse prognosis than transient ischemic attacks. Sometimes with extensive training and tasks patients can achieve some aspects of visual performance matching the intact hemifield vision, but full recovery of vision in all aspects doesn't occur after damage to the V1 area [23]. Prognosis also depended on the cause of Cortical blindness and proved to be much better in patients with Cortical blindness following surgery or cerebral angiography, than following a stroke [24, 25]. Order in which vision was recovered: light, motion, form, color, central and, finally, peripheral vision [26].

**Complications**

Cortical blindness causes a great amount of morbidity to the patients. Their daily life gets hampered. Family members of the patient, are also get affected by this. Cortical blindness leads to a socioeconomic burden. The patients may be more prone to falls and fractures.

**Case report**

A 60 year old female admitted to the hospital, with complaints of painless loss of vision in both eyes and slurring of speech of 4 days duration. No history of head injury, headache, vomiting, giddiness, loss of consciousness, weakness of limbs, loss of sensations, double vision, difficulty in swallowing, difficulty in chewing, urinary and fecal incontinence.

**Past history:** Patient has history of left hemiparesis – 2 years ago, for which she took treatment and recovered. Patient was a known case of NIDDM since 3 years and is on treatment for the same. No history of Asthma, HTN, TB, thyroid disorder, epilepsy, CRHD, MI or bleeding tendency.
No significant family history. Menstrual history: attained menopause 5 years ago.


Vitals: Temp- Normal, PR =90/min, regular. BP =130/80 mm of Hg. RR=18/min

CNS examination: Patient was conscious, Incoherent, with Confused Conversation, Rt. Handed, not oriented in time & place, Memory – Decreased, Speech – Dysarthria +. Optic Nerve – No Light Perception In Both Eyes. Pupils – Both sides 3 mm, reacting To Light accomodation reflex absent - Facial Nerve-Lt. UMN Facial Palsy +. Other CN – Normal.


SENSORY SYSTEM EXAMINATION: Pain, Temperature normal in all limbs. Pressure, Vibration, Joint sensations normal in all the limbs. Fundus – Both Eyes Immature Cataract ++ media clear, Fundus - Pale & No Papilloedema. No signs of Meningeal Irritation. Skull & Spine – Normal. Examination of Cardiovascular and Respiratory Systems was normal.

Investigations - Hb-10.4 g%, RBC count 7.2 mill/cumm, WBC count 8600 cells/cumm with Neutrophils - 72%, Lymphocytes - 24%, Eosinophils - 2%, Basrophils - 0%, Monocytes - 2%, Platelet count – 2.4 lakhs/cu mm. ESR 1st hr - 20 mm, Blood Urea 24 mg%, Serum Creatinine - 0.6 mg. Complete Urine Examination (CUE) – Albumin-Nil, Sugar-Nil, Random Blood Sugar of 300 mg/dl, Serum Electrolytes: Na – 139 m mol/L, K – 4.3 m mol/L, CL – 105 m mol/L. LFT – Normal, Fasting Lipid Profile-WNL, USG Abdomen-NAD, HIV- Non reactive, HbsAg- Negative, HBC- Negative, X-ray Chest PA view- NAD, ECG- Q++ in L1 and L2. CT scan brain (Figure - 1) showed: Bil. occipital lobe infarcts, EEG and VEP was not done.

A diagnosis of CVA – C.Thrombosis in both PCA territories with bilateral cortical blindness & and old Rt.MCA territory thrombosis with Lt.hemiparesis was made.

Treatment given
1. Inj MANNITOL 10% 100 ml iv TID
2. Inj AMPICILLIN 500mg iv QID
3. Inj. AMIKACIN 500 mg iv BD
4. INJ. ACTRAPID 6 UNITS S/C TID
5. Diabetic Diet
6. TAB.ECOSPRIN 75 MG
7. Inj OPTINEURON 1 amp iv OD
8. I.V.Fluids
9. Physiotherapy

Discussion
Cortical blindness is rare; mostly described the condition in vascular pathologies. Visual anosognosia was most commonly associated with occipital infarcts. Other vascular cases of visual anosognosia were in the setting of post pregnancy reversible cerebrovascular syndrome, systemic angiitis, and hypovolemic shock. The syndrome has also been reported in association with 6 other conditions: progressive multifocal leukoencephalopathy in the setting of HIV, adrenoleukodystrophy, bisynchronous occipital seizures, mitochondrial encephalopathy and lactic acidosis. Visual evoked potentials (VEPs) may have been helpful to support this theory, but were not obtained [27].
Although stroke remains the most common cause of Cortical blindness, surgery, especially cardiac surgery, and cerebral angiography are now frequent causes. Cardiac surgery, which was rarely performed three decades ago, may cause cerebral dysfunction through a variety of mechanisms, including anoxia from hypoperfusion, cerebral hemorrhage, and blood, fat, and air embolism. Ischemic neuronal damage of the calcarine cortex is frequent in patients who do not survive heart surgery.

The most common cause of cortical blindness is occlusion of the posterior cerebral arteries (embolic or thrombus). Hypoxic-ischaemic encephalopathy; Progressive multifocal leucoencephalopathy. Other leucodystrophies and bilateral gliomas are other causes. Bilateral occipital lobe infarction though extremely rare can occur due to thrombosis or embolism affecting the vertebro basilar system of vessels usually triggered off by fall in the blood pressure. When the terminal bifurcation of basilar artery is involved the symptoms may be bilateral. Melamed and Abraham reported three similar cases. In all the cases clinical and angiographic evidence of basilar artery occlusion was seen. These patients are generally in their sixties with a history of general atherosclerosis or history of vertebro-basilar transient ischaemic attacks. The onset of blindness is sudden; characteristically patients exhibit no other neurological signs. The eyes are still able to move through a full range, but optokinetic nystagmus cannot be elicited. Visual imagination and visual imaginary dreams are preserved. With very rare exception no cortical potential can be evoked in the occipital lobes with light flashes or pattern changes. The alpha rhythm is lost in EEG. A striking feature of patients with cortical blindness is subjective unawareness of ones disability (anosognosia). The other less common symptoms include visual hallucinations due to cortical irritation, associated apathy and mental disturbances. Rarely, oval pupils have been reported with bilateral cerebral infarctions. A large embolus in the basilar artery is usually fatal [28].

**Conclusion**

A diagnosis of CVA – C. Thrombosis in both PCA territories with bilateral cortical blindness & and old Rt.MCA territory thrombosis with Lt. hemiparesis was made. EEG and VEP was not done. Cortical blindness is rare; mostly described the condition in vascular pathologies. Hence this case report. The most common cause of cortical
blindness is occlusion of the posterior cerebral arteries (embolic or thrombus). Bilateral occipital lobe infarction though extremely rare can occur due to thrombosis or embolism affecting the vertebro basilar system of vessels.

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