A Review of Neuro-ophthalmologic Emergencies

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ABSTRACT

Background
Neuro-ophthalmic emergencies are relatively uncommon, however their outcome cause severe morbidity and even mortality. The ocular manifestations of these disorders are pointers to a more dangerous central nervous system or systemic pathology. The review aims to highlight the major ocular disorders that constitute neuro-ophthalmologic emergencies with a view to increasing the index of suspicion of these visual/life threatening disorders among primary care physicians, neurologists and ophthalmologists.

Method
The available literature on neuro-ophthalmologic emergencies was reviewed, using available journals and internet based search engines and resources. Keywords employed were Neuro-ophthalmology Emergency and Ocular Morbidity.

Results
The incidence of this group of emergencies is lower than that of other ophthalmic emergencies such as ruptured globe and retinal detachment; however they are associated with higher morbidity and even mortality. These emergencies can be grouped into four major categories for ease of diagnosis. Nigerian literature on neuro-ophthalmologic emergencies was unavailable.

Conclusion
The key to prevention of morbidity and mortality from neuro-ophthalmologic disorders is to have a good knowledge of the manifestations of these disorders and a high index of suspicion.

Keywords: Neuro-ophthalmology; Emergency; Ocular Morbidity

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INTRODUCTION
Neuro-ophthalmologic emergencies are those ocular disorders of neurologic origin which are vision threatening and/or life threatening if not promptly diagnosed and treated.1 In some of these conditions delay of even a few hours have dire consequences.

The incidence of this group of emergencies is lower than that of other ophthalmic emergencies such as ruptured globe and retinal detachment; however they are associated with higher morbidity and even mortality.

The emphasis of this review are the emergencies where failure to recognize the early stages of these diseases result in adverse prognosis2,3,4. These emergencies can be grouped into four major categories for ease of diagnosis (Table 1).1

The first 3 groups are based on initial symptoms and the last group accommodates other disease conditions that constitute a neuro-ophthalmic emergency but do not strictly fall under the other 3 groups. The review aims to highlight the major ocular disorders that constitute neuro-ophthalmologic emergencies with a view to increasing the index of suspicion of these visual/life threatening disorders among primary care physicians, neurologists and ophthalmologists by providing relevant information on the clinical presentation and management of these emergencies.

Table 1. Categories of Neuro-ophthalmologic emergencies based on initial symptom

<table>
<thead>
<tr>
<th>INITIAL SYMPTOM</th>
<th>OCULAR DISORDER</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visual Loss</td>
<td>Giant cell arteritis, transient monocular visual loss, occipital infarcts, methanol optic neuropathy</td>
</tr>
<tr>
<td>Ophthalmoplegia</td>
<td>Carvenous sinus thrombosis, intracranial aneurysms, Wernicke encephalopathy</td>
</tr>
<tr>
<td>Visual Loss and Ophthalmoplegia</td>
<td>Pituitary apoplexy, mucormycosis</td>
</tr>
<tr>
<td>Others</td>
<td>Sixth cranial nerve palsy, traumatic optic neuropathy, carotid arterial dissection</td>
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Giant Cell Arteritis (GCA)
This condition is also known as temporal arteritis, granulomatous arteritis and cranial arteritis5. Patients with this condition are at an increased risk of developing irreversible visual loss in one or both eyes, hence maintaining a high index of suspicion is crucial in preventing visual loss2. Visual manifestations of GCA are the common mode of presentation making the ophthalmologists critically responsible for early diagnosis and treatment6.

The disease has a slight predilection for women over men (2-4 times higher). Caucasians are affected much more often than African-Americans and Hispanics5. GCA is a systemic inflammatory vasculitis of unknown aetiology that affects medium and large caliber arteries. Most patients with GCA are over 60 years of age. Sudden loss of vision is the most common manifestation of GCA, occurring in approximately 50% of patients5. The most common cause of visual loss in GCA is arteritic anterior ischaemic optic neuropathy2,5. Other ophthalmic vascular presentations are central retinal artery occlusion, branch retinal artery occlusion, choroidal ischaemia and the ocular ischaemic syndrome. The neuroophthalmic manifestations of GCA include diplopia, ptosis and pupillary abnormalities2,5.
Systemic manifestations of the disease include, new onset headache which is localized, scalp tenderness, jaw claudication, myalgia and constitutional symptoms like fever, malaise, weight loss and anorexia. Diagnosis of GCA is based on clinical signs and symptoms. Positive findings in three or more systems in the elderly should raise a high level of suspicion. Features predictive of permanent visual loss include, jaw claudication and temporal artery abnormality on physical examination. Laboratory findings are helpful. Elevated erythrocyte sedimentation rate (ESR), C-reactive protein level and thrombocytosis are commonly present. For a more definitive diagnosis a superficial temporal artery biopsy should be done.

The universally accepted treatment of GCA is high dose corticosteroid. Initial intravenous steroid therapy is advocated by followed oral therapy. However there are no prospective trials that show that intravenous therapy is superior to oral therapy. The recommended dosage is 1-2mg/kg/day and reach about 20mg/day at 6months and 10mg/day at 1year.

Other classes of drugs used in the treatment of GCA include, antiplatelets, tumour necrosis factor(TNF)-inhibitors, cyclosporine and methotrexate.

**Transient monocular visual loss (TMVL)**

TMVL and amaurosis fugax are used interchangeably to describe painless vision loss in one eye, resulting from ischaemia or vascular insufficiency and lasting for several seconds to a few minutes. Compromised perfusion of the occipital lobe, visual pathway or the eyes may be secondary to thromboembolism, hypoperfusion, or angiopasm. Embolic occlusions of the arteries supplying the eye are a common cause of transient visual loss in adults. Emboli causing circulatory disturbances may originate in the heart or the carotid arteries. Hypoperfusion may be due to hypotension, cardiac arrhythmias, anemia, heart failure, or atherosclerotic and arteritic cerebrovascular disease. Arteritic and nonarteritic anterior ischemic optic neuropathy may also present with transient visual loss. Angiopasm may cause a temporary reduction in blood flow to the visual system and transient visual disturbance.

TMVL usually presents in people over 50 years of age who have risk factors such as diabetes mellitus, hypertension and hyperlipidemia. Patients usually give a history of similar previous episodes, other cerebral transient ischaemic attacks. Ocular presentation includes temporary loss of vision, jaw claudication, branch retinal artery occlusion. Basic work up for a diagnosis includes, blood pressure measurement, urinalysis, neuroimaging studies (magnetic resonance angiography, computerized tomographic angiography or transcranial Doppler). Treatment depends on the cause. The modification of risk factors is important in the management and a multidisciplinary approach is usually required.

**Occipital Infarcts**

Patients with occipital infarcts are often misdiagnosed as having an intracocular or functional problem. In this condition the fundus, ocular motility and pupillary light reaction are normal, however there is visual loss. An isolated homonymous visual field defect (homonymous hemianopia) of sudden onset is the hallmark of occipital stroke. Embolism of the posterior cerebral artery often with a cardiac source is a common cause. Occasionally an occipital stroke may be the first sign of a myocardial infarction or atrial fibrillation. Headaches are not usually reported by these patients.

An emergent electrocardiogram is pertinent in ruling out a cardiac etiology. Neuromaging would be useful in making a diagnosis; however a hyper acute infarcts (occurring within the first 6months) may be difficult to detect.

Early recognition of these infarcts will aid early referral to a stroke specialist.

**Methanol Optic Neuropathy**

Methanol(methyl alchohol, wood alchohol) is a colourless flammable liquid with close resemblance and taste to conventional alchohol(ethanol,ethyl alchohol). Toxicity occurs from accidental ingestion or intentional overdose. It is mainly consumed by those in the lower socioeconomic class in developing counties due to its low cost. Methanol is metabolized to formic acid by alchohol dehydrogenase which causes metabolic acidosis leading to blindness, cardiovascular instability and death.

Presentation is initially with nausea and vomiting. Patients then develop abdominal cramps. In the later stages drowsiness may progress to obtundation and coma.

Visual loss is believed to result from interruption of mitochondria function in the retrolaminar optic nerve. This causes hyperemia, oedema and optic nerve atrophy. Scotomas which are seen maybe central or centrocecal.

Traditionally ethanol is used as an antidote therapy. Fomepizole however, inhibits alchohol dehydrogenase and prevents the formation of formic acid. Correction of metabolic acidosis and hemodialysis may be necessary.

**Carvenous Sinus Thrombosis(CST)**

Septic thrombosis of the carvenous sinus is another potentially life threatening condition which may present with diplopia and orbital signs. CST is usually a late complication of a facial infections, sinusitis, dental infections, otitis media and orbital infection. Midfacial infections(most commonly a furuncle) are the most frequent source of infection. The usual pathogens of acute infection are gram positive bacteria, while chronic infections are more associated with gram negative bacteria and fungi.

Clinical presentation is due to venous obstruction and impairment of cranial nerves related to the sinus such as the 3rd, 4th, 6th cranial nerves and the first and second branch of the 5th cranial nerve. Most patients present acutely with pain around the eye, orbital congestion, proptosis, ophthalmoparesis and chemosis. They also have fever and headaches. Without effective therapy, infection spreads...
within 24 to 48 hours to the contralateral eye via the intercavaneous sinuses. The patient rapidly develops mental status changes such as drowsiness, coma and eventually death.

Treatment consists of early and aggressive appropriate antimicrobial therapy. Mortality rate for septic CST is 30% regardless of therapy and less than 40% of survivors have full recovery as they end up with neurological deficits.

INTRACRANIAL ANEURYSMS (ICA)
An aneurysm is an abnormal local dilatation of the blood vessel wall due to a defect, disease or injury. The 3 major types of ICA are saccular, fusiform and dissecting. The common causes of ICA include hemodynamically induced or degenerative vascular disease, atherosclerosis, underlying vascular pathology (fibromuscular dysplasia) and high flow states as seen in arteriovenous malformations and fistula. ICAs are more common in women and the incidence of rupture increases with age peaking at the 6th and 7th decades. Majority of ICAs arise from the carotid’s main trunk at the level of the posterior communicating artery (PCOM), the ophthalmic artery, and the carvenous sinus. The PCOM is by far the most frequent location to cause a third nerve palsy prior to rupture.

Differentiating between aneurysmal 3rd nerve palsy and ischaemic 3rd nerve palsy is crucial in managing the patient. An aneurysmal compression of the 3rd nerve is a medical emergency. Frontal head pain is seen in both ruptured and unruptured aneurysms. Ophthalmic manifestations range from vision loss due to ophthalmic artery aneurysms, cortical blindness resulting from basilar aneurysms or ophthalmoplegia due to aneurysms of the circle of Willis or the carvenous sinus. Patients will have ptosis, limitation of adduction, elevation and depression. The pupil may be spared or involved. If the pupil is spared it is most likely an ischaemic 3rd nerve palsy and this resolves spontaneously within weeks. If the pupil is involved, it will be dilated with poor response to light and accommodation. Ruptured ICA causes subarachnoid hemorrhages which lead to sudden severe headaches, meningal irritation and even changes in sensorium. Unruptured ICAs cause pain, seizures and transient ischaemic attacks.

The 3 major modalities used to study ICAs are magnetic resonance angiography, computed tomography angiography and catheter angiography. Conventional arteriography is the gold standard however.

Treatment involves application of surgical clips and endovascular coils.

WERNICKE’S ENCEPHALOPATHY (WE)
This is an acute neurological disorder caused by deficiency of Thiamine(Vitamin B1). Predisposing states for thiamine depletion include; chronic alcoholism, malnutrition, chronic renal dialysis, conditions causing protracted vomiting such as hyperemesis gravidarum, gastric resection, and AIDS. Thiamine is a cofactor for several enzymes in the Kreb’s cycle and pentose phosphate pathway. When there is a deficiency this result in failure of the thiamine dependent systems which play a vital role in cerebral energy utilization thus propagating brain tissue injury.

The classic clinical triad of WE, are ophthalmoplegia, mental confusion and ataxia. Ocular abnormalities are the hallmark of WE. These include nystagmus, internuclear ophthalmoplegia, conjugate gaze palsies (reflecting cranial nerve involvement of the oculomotor, abducens and vestibular nuclei). WE is associated with high neurologic morbidity (Korsacoff amnesia). These patients may later develop hypothermia, coma and death.

Treatment involves the thiamine repletion with 100 -200mg of Thiamine intravenously or intramuscularly.

PITUITARY APOPLEXY (PA)
This refers to a hemorrhage or infarction of a pituitary tumour. It is a life threatening condition which is rare and usually stems from a sudden expansion of a pituitary adenoma (less than 10% of cases of adenomas) or less commonly a non adenomatous gland. This then causes a mass effect on adjacent structures such as the optic chiasm and hypothalamus. Precipitating factors include reduced blood flow (as in hypotension and valsalva maneuvers), stimulation of the gland in increased estrogen states such as pregnancy, anticoagulation, bromocriptine treatment, head trauma, pituitary irradiation and increased blood flow (as in malignant hypertension).

Clinical presentation is with sudden severe headache, neck stiffness, nausea, vomiting, altered level of consciousness, thermoregulatory and cardiorespiratory dysfunction due to hypofunction of the gland patients may exhibit irregular menses, decreased libido, hyponatraemia, hypothyroidism or hypocortisolism. Ocular presentation includes a painful ophthalmoplegia, vision loss which may be unilateral or bilateral with variable severity. Visual defects are common and are usually of the bitemporal pattern.

Diagnosis of this condition requires a high index of suspicion, because in a large majority of cases a tumour was not suspected prior to haemor rhage. Therefore the occurrence of headaches and visual symptoms in the setting of any predisposing factor should heighten suspicion.

MUCORMYCOSIS (PHYCOMYCOSIS)
This is a rare opportunistic, aggressive and fatal infection caused by the fungus mucoraca. It occurs in debilitating and immunompromised patients. Common predisposing conditions include diabetes mellitus, neutropaenia, haematologic malignancies, patients on chemotherapy and steroid therapy, burns and organ transplantation. The most common form of mucormycosis is the rhino-orbital cerebral form (ROC).
The mode of transmission is by inhalation. ROC mucormycosis develops via direct invasion of the orbit via the lamina papryacea of the paranasal sinuses. It invades blood vessels forming an occlusive vasculitis causing ischaemia, tissue necrosis with the formation of an eschar which is characteristic.

The clinical presentation is usually with headache, fever and periorbital pain. Ocular presentation is with visual loss, proptosis, ophthalmoplegia, conjunctival injection and chemosis. Acute orbital cellulitis is heralded by a blood tinged nasal discharge or epistaxis. Involvement of the brain could lead to brain abscess, intracranial hemorrhage, seizures and death. Making a diagnosis requires a high index of suspicion, with careful examination of the oral and nasal mucosa in at risk patients and biopsy of necrotic tissue. Treatment is with intravenous amphotericin B, posaconazole and hyperbaric oxygen. Surgical debridement of the necrotic tissue must be done early. ROC mucormycosis has a high morbidity and mortality rate ranging from 30 - 69%.

**SIXTH CRANIAL NERVE PALSY (ABDUCENS NERVE PALSY)**

This constitutes a neuro ophthalmic emergency in the setting of a raised intracranial pressure (ICP). This could be due to intracranial mass lesions or pseudotumour cerebri. ICP leads to downward displacement of the brainstem with stretching of the 6th nerve over the petrous bone.

Presentation is with headache, binocular horizontal diplopia, esotropia which is incomitant (the angle of deviation differs in various positions of gaze as a result of restriction) and papilloedema. Sixth nerve palsy associated with papilloedema is highly suggestive of increased intracranial pressure due to an intracranial mass lesion or pseudotumour cerebri. In this setting urgent neuroimaging is required and if imaging studies are negative, a lumbar puncture with specific attention to cerebrospinal fluid opening pressure is warranted.

**TRAUMATIC OPTIC NEUROPATHY (TON)**

TON is a devastating potential complication of blunt or penetrating cranio-orbital trauma. Damage to the optic nerve may occur from transection of the optic nerve fibres, haemorrhage, oedema or interruption of the blood supply. Common causes of TON include; motor vehicle accidents, bicycle accidents, falls, sports injuries and physical violence. TON could be direct or indirect. Direct TON is the term used when the optic nerve is impinged, crushed or transected during open craniofacial injuries such as penetrating wounds (e.g knives, pellets). Indirect TON occurs in the absence of direct optic nerve injury and is more common than the direct TON.

The diagnosis for TON is clinical. Patients with midfacial and cranial trauma should elicit a high index of suspicion for TON. It is also important to note that these patients may have no visible sign of injury. Clinically the patients present with loss of vision, relative afferent pupillary dilatation is the sine qua non. The treatment of TON is controversial as there are no uniform guidelines for its management. Corticosteroids have been recommended, however many patients with TON also have concomitant head injury and the CRASH trial (corticosteroid randomization after significant head injury) has shown that intravenous steroid treatment may be harmful.

**CAROTID ARTERIAL DISSECTION (CAD)**

CAD begins as a tear in the wall of the arteries which causes blood under arterial pressure to enter the wall and split the layers. This results in intramural haematoma and aneurysmal dilation which are both sources of microemboli and may also exert mass effects on surrounding structures.

CAD is a significant cause of stroke in all age groups, but occurs most frequently in the 5th decade of life. Internal carotid artery (ICA) dissection could be caused by trauma or could be spontaneous.

Maintaining a high index of suspicion anytime a patient presents with non specific focal neurologic complaints particularly involving the cranial nerves and after major or minor trauma is essential. The most common initial clinical manifestation of ICA dissection is pain which could be localized to the head, eye, jaw, face or neck. Patients may also present with oculosympathetic palsy, transient monocular visual loss, cranial nerve palsies, epistaxis, and hemiparesis.

CAD can be diagnosed with noninvasive methods such as magnetic resonance imaging, magnetic resonance angiography or computed tomographic angiography. The study of choice however is a T1 weighted MRI of the head. Treatment usually involves bed rest with anticoagulants to prevent thrombus formation and embolization. Most dissections heal spontaneously within 3 months with recovery of cranial nerve palsies.

**CONCLUSION**

Neuro-ophthalmologic emergencies though rare present a situation where failure to recognize the earliest signs of the disease process could adversely affect outcome. Clinical outcome is therefore dependent on accurate and timely diagnosis.

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