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Mini Review

Neuro-ophthalmological emergency disorders: A general view

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Abstract

Neuro-ophthalmological emergency disorders usually occur with symptoms of visual loss, diplopia, ocular motility impairment and anisocoria. In this mini-review, we aim to take look the common neuro-ophthalmological emergency disorders. The delayed diagnosis of the neuro-ophthalmological emergencies puts the patient at risk of death or blindness. If these are well-known, the discrimination and management of these emergency conditions will be easier.

Introduction

Neuro-ophthalmology is a subspecialty of both neurology and ophthalmology training the visual pathways including the optic nerve, chiasm, optic tract, lateral geniculate nucleus, optic radiation and cerebral visual cortex, nerves associated with ocular muscles and ocular muscles. Symptoms associated with neuro-ophthalmic disorders usually include afferent visual system disorders such as optic neuritis (especially Multiple Sclerosis (MS)), vision-related migraines, optic neuropathy, papilledema, pseudotumor cerebri, brain tumors or strokes, and the efferent visual system disorders such as anisocoria and other pupil abnormalities, diplopia and other visual disturbances (phospenes, etc.), un-explained vision loss, sudden temporary or permanent visual loss, ophthalmoplegia, ptosis, eyelid and facial spasms, eye movement disorders in paralitic or restrictive types (thyroid eye disease, Myasthenia gravis, nystagmus, blepharospasm, and acute visual perception disorders or high cortical visual dysfunctions [1-9]. However, the neuro-ophthalmological emergencies constitute vision or life-threatening conditions if diagnosis and treatments are not made promptly. The common neuro-ophthalmological emergency disorders are arteritic anterior ischemic optic neuropathy (AAION), pituitary apoplexy, cavernous sinus thrombosis, rino-orbital-cerebral mucormycosis (ROCM), isolated third nerve palsy, multiple unilateral or bilateral oculomotor palsies, acute methanol toxication, Tolosa-Hunt syndrome, Miller Fisher Syndrome and Horner syndrome [1-5, 13-15]. The main signs of the life or sight-threatening neuro-ophthalmological emergency conditions include diplopia, ptosis (acute painful or chronic painless), transient visual loss, anisocoria, acute painful or painless homonymous hemianopsia, acute bitemporal hemianopsia, severe pain in head/neck, painful ophthalmoplegia and ocular motility disorder [1-9].

A pupil-involving third cranial nerve (CN3) palsy is a true neuro-ophthalmological emergency. The ophthalmologist should be able to distinguish an aneurysm from other causes of CN3 palsy. The detection of early and marked pupillary involvement in CN3 palsy is critical. Because the compressive lesions of CN3 usually affect both the central

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somatomotor fibers and the peripheral superomedial pupillary fibers and they cause the pupil-involving CN3 palsy. However, ischemic lesions of the nerve involve only the central somatomotor fibers, so, ischemic CN3 palsy has no a pupillary involvement [1-9]. The main mechanisms of the aneurysmal CN3 palsy, including direct compression by an enlarged aneurysmal sac, subarachnoid blood irritation, or aneurysm pulsation. An aneurysmal CN3 palsy typically presents with pain, a mid-dilated pupil with poor or absent direct light reaction, ptosis and outward and downward shift of the eye. When an aneurysm compresses CN3, a sluggishly reactive or dilated pupil occurs. Because the pupil-involving CN3 palsy especially with an acute headache may be an indicator of an aneurysm, potentially fatal event, in case of its rupture or leak and subarachnoid hemorrhage. However, it should be remembered that a posterior communicating artery (PCOM) aneurysm may present initially with normal pupils [2,6-9]. Posterior communicating artery (PCA) aneurysms present with III nerve palsy 30% to 60% of the times, and approximately 40% of aneurysms are located at the level of PCA, the ophthalmic artery, and the cavernous sinus. Besides an aneurysm and vascular pathology, giant cell arteritis (GCA), pituitary apoplexy, demyelinating disease (MS), midbrain infarction, CN3 schwannoma/meningioma or cerebral metastasis, trauma and ophthalmoplegic migraine may also cause the CN3 palsy with pupil involvement. Although Myasthenia gravis (MG) may mimic a CN3 palsy, the pupil is never be involved in MG [1-12]. Myasthenia gravis is a neuromuscular disease leading to increasing muscle weakness and fatigue at the end of the day. Its initial ocular symptoms are ptosis and diplopia [5,7,9]. In case of the suspect for an aneurysm, an emergent brain magnetic resonance angiography or computed tomographic angiogram detecting aneurysms as small as 3 mm in size should be performed [4].

Giant cell arteritis (GCA)or Horton Disease is a severe inflammatory vascular disease that can lead to stroke, blindness, and heart attack. GCA is a systemic immune-mediated granulomatous vasculitis involving large- and medium-sized arteries. Although the death from GCA alone is rare, it usually results from the stroke or aortic dissection. Thus, AAION is another neuro-ophthalmological and medical emergency disease. Because AAION forms 10% of all patients with anterior ischemic optic neuropathy, and it has about a 75% risk of the blindness in both eyes within days or weeks when AAION is not diagnosed recognized and treated promptly [1-11].

Horner Syndrome is presented with ptosis, miosis, and anhidrosis without the visual loss. It should be considered as a neuro-ophthalmologic emergency because it may indicate underlying serious pathology such as malignancy (neuroblastoma, lung tumors, thyroid carcinoma, and metastatic tumors), stroke (in the lateral medulla oblongata), or carotid artery aneurysm or dissection even if the underlying cause is more commonly benign [1-7,12]. Carotid artery dissection (CAD) presents with pain on a side of the face (ear or jaw) and anisocoria. However, CAD may be revealed with less frequent ophthalmological features of dissection include transient monocular vision loss, Non-arteritic AION, posterior ischemic optic neuropathy, central retinal artery occlusion, ocular ischemic syndrome, and ocular motor nerve palsies. Carotid artery aneurysm (CAA) may present with diplopia (due to a sixth nerve palsy) and clinical signs of Horner Syndrome [1-5,12]. The pituitary adenoma may be diagnosed with a headache, hormonal changes, or visual disturbances and bitemporal hemianopsia. Fortunately, it has usually benign nature and does not grow fastly, though, it can cause severe visual loss and even blindness if it compresses on the chiasm or the visual pathways. Additionally, it should be considered that in some pituitary adenoma cases,nonclassical/uncharacteristic/untypical or unilateral/asymmetrical temporal visual field defect or junctional scotoma may be observed if the compression region is the distal optic nerve or anterior to the chiasm [1-8].

Pituitary apoplexy or pituitary tumor apoplexy is characterized by a sudden severe headache and neck stiffness due to the meningeal irritation, and hormonal dysfunction

and visual acuity and visual field defect. It is caused by the sudden enlargement of the pituitary gland from acute hemorrhage or ischemia in an existing pituitary adenoma. Ocular signs are derived from the compression on surrounding structures such as optic nerve, optic chiasm and/or cavernous sinus or chiasm and the impairment of the ocular motility from the involvement of the cranial nerves crossing the cavernous sinus. Visual field loss is characteristically a bitemporal visual field loss or a junctional scotoma. Ocular motility impairment is unilateral or bilateral ophthalmoplegia (most commonly third nerve palsy, followed by sixth and fourth nerve palsies) can be seen in about 50% of the cases. Pituitary apoplexy is a life-threatening condition [1-8,13]. Miller Fisher Syndrome is a rare acute immune-mediated neuropathy and a variant of Guillain-Barré Syndrome. Its cardinal clinical features are ataxia, areflexia, and ophthalmoplegia. MFS commonly presents with diplopia following the acute onset of bilateral complete external ophthalmoplegia [14]. Tolosa-Hunt syndrome (THS) is a painful ophthalmoplegia caused by nonspecific inflammation with unknown etiology in the anterior cavernous sinuses or superior orbital fissure and orbital apex. Its clinical findings include unilateral orbital pain associated with paralysis of one or more of the third, fourth and sixth cranial nerves. In addition to the involvement of the ocular motor nerves, ophthalmic and maxillary branches of the fifth cranial nerve may be affected [15].

Transient visual loss (TVL) is a sudden, reversible visual function impairment lasting for less than 24 hours. Visual loss is usually associated with positive visual phenomena such as flashes, flickers, colorful or geometric shapes. It may be bilateral or unilateral. It has been considered that the main causes of TVL are ischemia, migraine or seizure [1-9]. Posterior reversible encephalopathy syndrome (PRES) is a rare syndrome characterized by a bilateral visual loss, together with a headache, confusion, seizures, and vomiting. The cause may be malignant hypertension, eclampsia or certain drugs (immunosuppressive or antineoplastic) [1-9]. Transient ischemic attack (TIA) is currently defined as “a transient episode of neurologic dysfunction caused by focal brain, spinal cord or retinal ischemia without acute infarction.” In the past, it was defined based on symptom duration lasting less than 24 hours, with typical episodes lasting less than 1 hour. In a one-third of the cases, TIA is associated with visual symptoms including isolated homonymous hemianopia, bilateral blindness and diplopia [1-9,12].

Rhino-orbital-cerebral mucormycosis (ROCM) is a rare (1.7 cases/million), acute, and aggressive opportunistic fungal infection. It is typically fatal if RCOM cannot urgency treated. It especially occurs in several immunocompromised individuals with diabetes, chronic immunosuppression, hematological malignancies, hematopoietic stem cell transplantation, and solid organ transplantation. Diabetic ketoacidosis is the most common predisposing factor. ROCM results from the invasion to palate, mouth, face, paranasal sinuses, orbit, and sometimes cerebrum following the inhalation of fungal sporangiospores. The ethmoid sinus is a critical region because the mucormycosis in this sinus may extend through the lamina papyracea into the orbit, extraocular muscles, and optic nerve [1-9,16-20]. ROCM has a very high residual morbidity and mortality ratios because of the fungal angioinvasion, vascular occlusion, and extensive tissue necrosis. Additionally, the reaching of the antifungal drugs to the infection site is inadequate due to vascular thrombosis and the aggressive surgery is limited due to the complex rhino-orbital-cerebral anatomy. As the extension to cavernous sinus of the infection might be fatal due to the promotion of intracranial thrombosis or the formation of a mycotic aneurysm, ischemia or subarachnoid hemorrhage, early diagnosis and aggressive management of RCOM is essential and critical [16-20]. ROCM is often presented as a painful orbital apex syndrome including pain at the head/facial, fever, nasal mucosal ulceration/necrosis, sinusitis, unilateral orbital cellulitis and periorbital-facial edema, ptosis, complete ophthalmoplegia, early vision loss due to

optic neuropathy, facial anhidrosis, and decreased corneal sensation due to cavernous sinus involvement. A painful black necrotic ulceration and scar on the skin, nasal mucosa, or palate may occur in the one to five of the patients. ROCM treatment includes emergency antifungal therapy with amphotericin B, the elimination of the risk factors, and surgical debridement of the infected and devitalized tissue [16-20].

Cavernous sinus thrombosis (CST) is a rare vision- and life-threatening disorder. It often occurs in the third decade with a mean age of 22 years old. The septic type of CST has high mortality and morbidity rates. Septic type usually occurs as a late complication of an infection of the central face due to the infections spreading through the venous system, from the face, paranasal sinuses, teeth, ear, or orbit. Staphylococcus aureus is the most common causative agent. The aseptic type of CST is usually caused by the events promoting the thrombosis such as polycythemia, vasculitis, pregnancy, the use of oral contraceptive drugs, trauma, surgery or cranial tumors. The main signs of septic type are presented with abrupt fewer, proptosis, chemosis, ptosis, cranial nerve palsies (third, fourth, fifth nerves) in a vast majority. Additionally, headache, lethargy and optic disc edema are seen over a half of the cases. In less a half of the cases, ION, central retinal vein or artery occlusion, hemiparesis and the signs of meningismus may be observed. Treatment of septic CST consists of prompt therapy directed at the primary infection source, including a third-generation cephalosporin, nafcillin, and metronidazole in accord with infectious disease consultation and consideration of early anticoagulation with low-molecular-weight heparin, which may decrease mortality and morbidity. Aseptic CST is managed with primary attention to coagulation status and any underlying disease. However, initial antibiotics treatment is recommended, until a septic etiology is ruled out [21-25].

Methanol poisoning is presented in almost a half of the patients with ocular symptoms including partial/total visual loss or blurred vision, photophobia, visual hallucinations after six hours or more following methanol ingestion. Ocular examination usually shows normal to constricted visual field, sluggish or non-reactive pupils, nystagmus, hyperemic optic discs, papilloedema, retinal edema and hemorrhages and decreased vision to total vision loss. The most common acute field defect is a dense central scotoma. As methanol poisoning is a life-threatening neuro-ophthalmological disease and it presents with ocular symptoms in a half of the patients, its early diagnosis is very critical for the management [26-27].

Orbital compartment syndrome (OCS) is one of a few true ophthalmologic emergencies. Its cardinal findings are the resistance to retropulsion of the globe, a tight orbit and tense eyelids, acute onset vision loss, diplopia, ocular tenderness or pain, periorbital edema, proptosis, impaired eye motility, fixed dilated pupil or an afferent pupillary defect, the elevated intraocular pressure, ecchymosis and subconjunctival hemorrhage. It maybe indirectly included into the class of neuro-ophthalmologic emergencies because, in OCS, the vision loss arises from some ischemic or occlusive optic nerve diseases such as central retinal artery occlusion, traumatic or compressive optic neuropathy and ischemic optic neuropathy [28-29]. OCS is mainly caused by the hemorrhage, infection/abscess, tumor, edema/emphysema in the orbit, retrobulbar injection and preexisting medical disorders. OCS develops due to acute intra-orbital pressure rising and if not immediately treated, irreversible visual loss occurs. However, in this emergency neuro-ophthalmologic disorder, the first intervention should perform by emergency physician or ophthalmologist, and the neurologic intervention and consultation are usually not needed for the prevention of the vision [28-29]. In conclusion, the ophthalmologist or emergency physician should be aware of the neuro-ophthalmic emergency disorders such as GCA, ROCM, CST, pupil-involved CN3 palsy, and Horner syndrome.

Authorship Contributions

Concept and Design, Data Collection, Analysis, Interpretation: Burak Turgut, Feyza Çaliş Karanfil, Fatoş Altun Turgut; Literature Search: Fatoş Altun Turgut; Writing: Burak Turgut, Feyza Çaliş Karanfil.

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