Painful Ophthalmoplegia during the Early Lockdown of Novel COVID-19 Pandemic

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INTRODUCTION

Painful ophthalmoplegia can present with various aetiologies which can be very harmful and needs immediate intervention. It is a very stressful and painful condition for the patient. An appropriate diagnosis can be made through proper neuroimaging. Only a few cases need extensive workup like an orbital pseudotumor and Tolosa-Hunt syndrome. We describe a set of five different cases who presented with painful ophthalmoplegia, out of which three patients came with associated proptosis. An early diagnosis was made based on both clinical and imaging i.e., magnetic resonance imaging (MRI) or computed tomography (CT) findings. An early diagnosis can help in preventing harmful effects and save from sight-threatening situations.

Ophthalmoplegia indicates paralysis, weakness or restriction of eye muscles. There are various classifications like internal / external; painful / painless; total / partial; pupil involving / sparing. Periorbital or hemicranial pain with ipsilateral ocular motor nerve palsies can be grouped under painful ophthalmplegia.[1] Broadly, the causes are neuropathy, myopathy and disorders effecting the neuromuscular junction. Here we report five cases of painful ophthalmoplegia. One interesting fact was that all the five patients presented to us during the Novel Covid 19 pandemic. Covid test was done for two patients which turned out to be negative. The remaining 3 patients were treated on OPD basis.

PRESENTATION OF CASE

Case 1. Painful Ophthalmoplegia with Multiple Cranial Nerve Palsy Due to Retrobulbar Lesion (Orbital Cavernous Haemangioma)

A 48-year-old female was admitted with a one-day history of protrusion of left eye associated with drooping of the upper eyelid, pain, diplopia, headache and defective vision. There was no history of any thyroid disorder or trauma to the left eye. On examination, the left eye showed axial proptosis with moderate ptosis and restriction of extraocular movements in all the gazes (Figure 1). Corneal sensation was decreased in the left eye. The proptosis was non-pulsatile without ocular bruit. Examination of the right eye was normal. The best corrected visual acuity (BCVA) was 6 / 9 in the right eye and 6 / 36 in the left eye. Colour vision of both eyes was normal. Pupils were reacting to both direct and indirect reflexes. On palpation, resistance was felt on retropulsion. Fundus examination was normal. Except II, III, IV, V and VI, all other cranial nerves function appeared normal. On clinical examination, her vital signs and systemic examination were normal. There was no organomegaly or lymph node enlargement. Her thyroid profile, ESR and C-reactive protein were normal. MRI of the orbit reported a well-circumscribed intraconal retrobulbar lesion possibility includes Orbital Cavernous Haemangioma (Figure 1).

The patient was treated with six doses of intravenous steroids following which proptosis and pain improved. The patient was discharged with oral steroids. At present, she is on follow up.

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Case 2. Painful Ophthalmoplegia Due to a Mass in the Superomedial Aspect of the Eyeball (Orbital Pseudotumor)

A 26-year-old female presented with swelling of the left eye associated with pain for 5 days. On examination, there was left eye periorbital oedema, mechanical ptosis and conjunctival chemosis. The best corrected visual acuity (BCVA) was 6 / 9 in both eyes. The pupillary reaction was normal to both direct and indirect reflexes. Fundus examination of both the eyes was normal. She was treated as left eye preseptal cellulitis with oral and topical antibiotics. Ear, nose and throat (ENT) opinion was taken. Patient reviewed after 4 days. On examination, the eyeball was slightly displaced downwards (Figure 2). A 2 × 1 × 1 cm mass was felt on the medial aspect of the eyeball with tenderness, irregular margins, firm in consistency, not fixed to the underlying structures with mechanical ptosis and conjunctival congestion. There was a restriction of elevation and abduction. There was no lymph node enlargement. CT orbit reported a poorly marginated soft tissue density lesion involving intra- and extraconal compartments of the left orbit in superior quadrants with soft tissue thickening of left eyelid possibility includes pseudotumor (Figure 2). Investigations were sent and a diagnosis of pseudotumor was made. Patient was started on systemic steroids. We lost the follow up of the patient due to the Covid pandemic situation.

Case 3. Painful Ophthalmoplegia Due to Frontal Paranasal Sinus Mass Lesion (Frontal Mucocele).

A 50-year-old male presented with protrusion of left eye for 6 months. It was gradually progressive in nature. On examination, there was eccentric proptosis in which the eyeball was protruding inferiorly. There was a restriction of extraocular movements in all the gazes. BCVA was 6 / 6 in both eyes. Colour vision was normal. Both eyes fundus examination was normal. He was a known diabetic for 5 years and was uncontrolled. MRI Orbit with contrast reported left frontal paranasal sinus mass lesion with left extraconal extension causing compression of the left eyeball and superior rectus muscle, lesion also causing cortical breach of the inner table of left frontal bone without any intracranial extension, most probably, a mucocoele and had suggested for biopsy. But unfortunately, we lost the follow up of the patient due to Covid pandemic situation.

Case 4. Painful Ophthalmoplegia Due to Third Nerve Palsy Resulting from Posterior Communicating Artery Aneurysm

A 75-year-old female presented with sudden onset of left eye drooping of the upper eyelid since one week associated with a severe headache and left eye pain. There was no history of trauma or fever or weakness of the face or limb or tremors. No systemic illness. On examination, there was complete ptosis of the left eye. BCVA was 6 / 12 in the right eye and 6 / 60 in the left eye. The left eyeball was down and out (Figure 3). There was a restriction of elevation, depression and adduction in the left eye. Right eye pupil was 3 mm in size and reacting to light while left eye pupil was 6 mm in size, mid-dilated, fixed and not reacting to direct and indirect reflexes. On lifting the upper eyelid, the patient complained diplopia hence diplopia charting was done which showed a crossed diplopia towards left side on dextro-elevation, dextro-version and dextro-depression. Fundus examination of the right showed normal fundus and left eye showed pallor of the disc with peripapillary atrophy and arteriolar attenuation. MRI of brain reported Posterior Communicating Artery Aneurysm, Subarachnoid Haemorrhage. The patient was referred to a neurosurgeon for further management.

Case 5. Painful Ophthalmoplegia Due to Bilateral Isolated Sixth Nerve Palsy "False Localizing Sign" Due to Subdural Haemorrhage

A 45-year-old female presented with sudden onset of double vision since 4 days associated with headache. She had a history of a road traffic accident and sustained multiple injuries on the right side of her face following which she developed double vision within 4 days. Diplopia was sudden in onset in horizontal gaze and worsens for distant vision. BCVA was 6 / 9 NIP in both eyes. There was, both eyes, esotropia of 15° in primary position. Abduction was restricted in both eyes. All other extraocular movements were normal. Both the pupils were reacting to both direct and indirect light reflexes. Fundus examination of both eyes showed blurred and elevated disc margins, obliteration of physiological cup, A:V was 2:4, engorged veins, streak haemorrhage was present adjacent to the disc margin which indicates bilateral established papilledema (Figure 4). Diplopia charting showed uncrossed diplopia in both eyes. Her systemic examination and vitals were normal. Automated perimetry showed enlargement of blind spot. CT showed subdural haemorrhage in the right temporoparietal region (Figure 4). She was treated with intravenous low molecular weight heparin and systemic steroids. She was reviewed after one month in which her extraocular muscles (EOM) became normal and also her fundus examination was normal with normal disc and vessels (Figure 5). The perimetry was repeated which was normal. The patient was asked for regular follow-ups for observation.

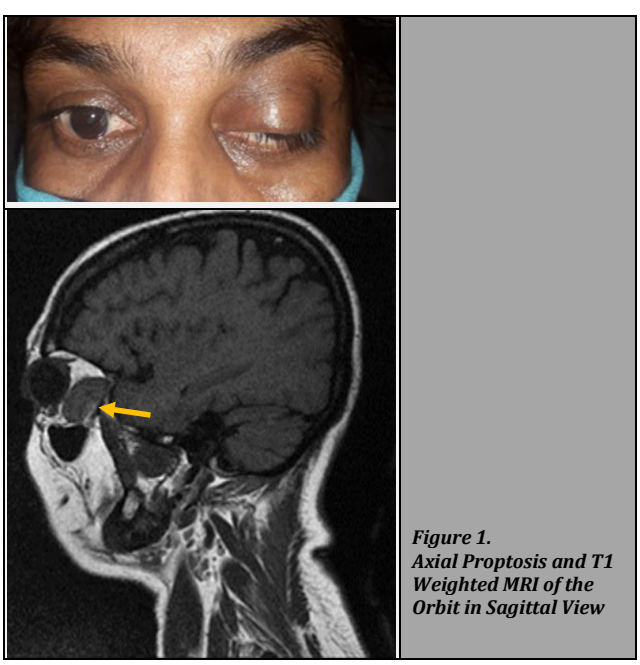


Figure 1.
Axial Proptosis and T1 Weighted MRI of the Orbit in Sagittal View

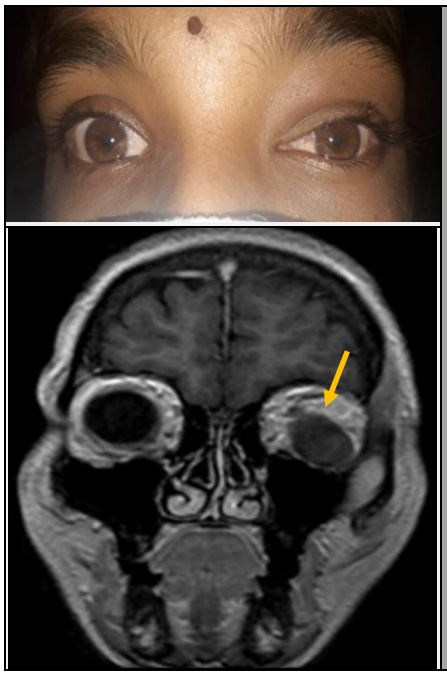


Figure 2. Eccentric Proptosis and CT Scan in Coronal View

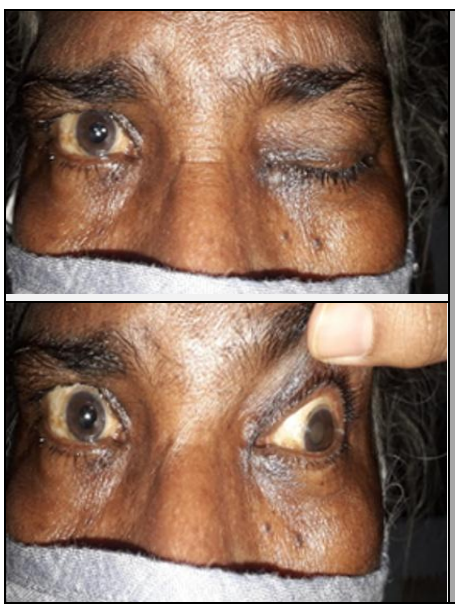


Figure 3. Presentation of Third Nerve Palsy

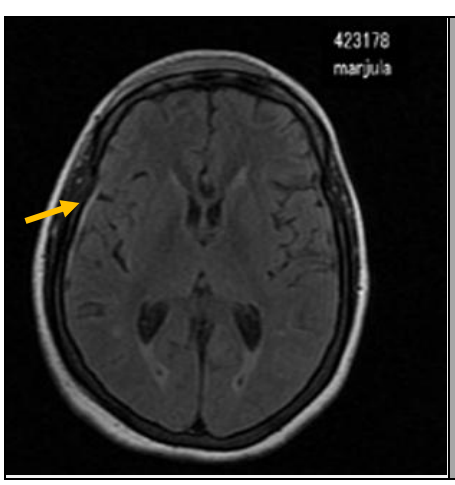
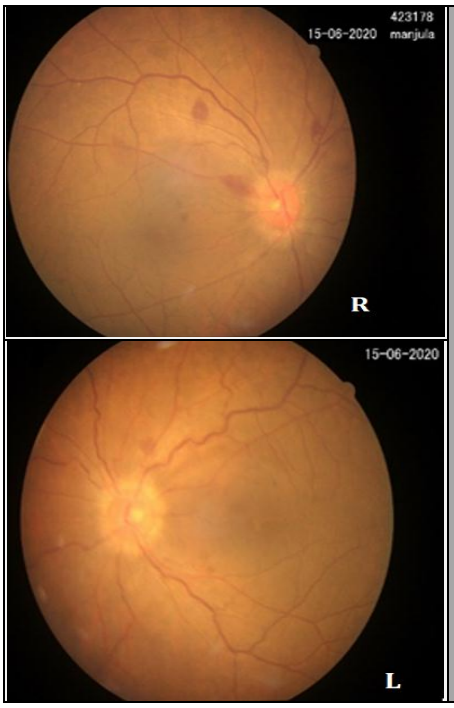


Figure 4. T1 Weighted-MRI Brain in Axial View, Right Temporoparietal Region SDH and



Both Eyes with Established Papilledema

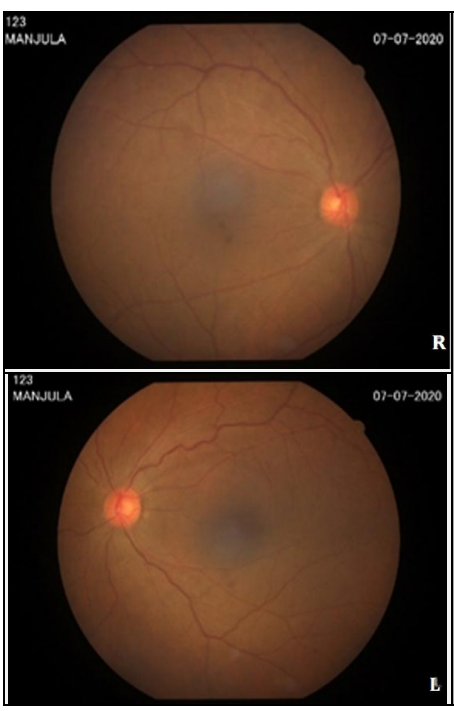


Figure 5. Normal Fundus after the Administration of Systemic Steroids

DISCUSSION

Ophthalmoplegia can also be broadly classified into restrictive and paralytic. In our report, the first 3 cases can be categorized into restrictive and the last 2 cases under paralytic types of ophthalmoplegia. Cavernous haemangioma is the second most common cause of proptosis after a thyroid eye disease.[2] The most common location for this tumour is the intraconal space. It mostly presents with painless, slowly progressive, axial proptosis and diplopia; in large lesions compressive optic neuropathy may occur.[3,4]

Age / Sex	BCVA		Clinical Presentation	MRI / CT	Diagnosis	Treatment	Outcome
	RE	LE					
48 / F	6 / 9	6 / 36	LE Axial proptosis, ptosis, diplopia	MRI: Orbital Cavernous Haemangioma	Painful ophthalmoplegia with multiple cranial nerve palsy due to retrobulbar lesion	IV steroids	Patient improved
26 / F	6 / 9	6 / 9	LE Chemosis, Eccentric proptosis	CT: possibility includes pseudotumor.	Painful ophthalmoplegia due to a mass in the superomedial aspect of the eyeball.	Systemic steroids	Lost follow-up
50 / M	6 / 6	6 / 6	LE Eccentric proptosis	MRI Orbit with contrast – left frontal mucocele	Painful ophthalmoplegia due to frontal paranasal sinus mucocele	Surgical	Lost follow-up
75 / F	6 / 12	6 / 60	LE Complete ptosis, outward and downward displacement of eyeball, non-reacting pupil	MRI brain: Posterior Communicating Artery Aneurysm? Subarachnoid haemorrhage.	Painful ophthalmoplegia due to third nerve palsy resulting from posterior communicating artery aneurysm	Referred to neurosurgeon for coiling	Lost follow-up
45 / F	6 / 9	6 / 9	Headache with history of trauma	CT: subdural haemorrhage in the right temporoparietal region	Painful ophthalmoplegia due to bilateral isolated sixth nerve palsy "false localizing sign":	Systemic steroids	Patient recovered

Table 1. Summary of Cases

Axial proptosis is the most common symptom that occurs due to the location of the lesion which is the intraconal space.[5,6,7,8] Approximately, 6 % of the intraorbital / retro bulbar masses of the orbit are due to cavernous venous malformation or cavernous haemangioma. Typically, middle-aged individuals are affected and will present with mass effect symptoms such as proptosis, pain, diplopia, and visual disturbance due to compression of the optic nerve. CT, especially with the use of contrast dye that allows for the enhancement of the haemangioma helps in obtaining a differential diagnosis. However, it is common for patients to sustain both magnetic resonance imaging (MRI) and CT as we did in our patient[9,10] (Figure 1). Colour Doppler and angiography may also help.[10,11] Treatment options for an orbital cavernous venous malformation can be either a nonsurgical or surgical excision methods. Surgical approach is determined by the location of the lesion. Nonsurgical methods may be indicated for small asymptomatic non-enlarging masses and that have all been described as surgical approaches are lateral orbitotomy, supraorbital, trans conjunctival, transanal, endoscopic, and extradural approaches.[11,12] In our case as the patient responded to steroids, we decided to follow up the patient. However, the mainstay of treatment will be excision.

Orbital pseudotumor (OP) or idiopathic orbital inflammatory disease was first described in 1903 by ADDIN and by Busse and Hochheim.[13] Inflammatory pseudotumor, most commonly occurs in the lung, liver, and orbit but nearly every organ in the body can be effected.[14,15] OP most commonly presents among third to sixth decades[14] with no strong sex predilection;[13] it is the third most common orbital disease following Graves 'orbitopathy and lymphoproliferative disease and accounts for 4.7 % to 6.3 % of orbital disorders.[13,15] Symptoms most commonly develop within hours or days but may be subacute or chronic and present with proptosis and inflammatory signs like pain, swelling and erythema; ptosis, chemosis, motility dysfunction, and optic neuropathy may also occur.[14] The main differential diagnoses that should be excluded are infections, an inflammatory reaction to trauma or foreign body, thyroid dysfunction, vasculitis (Wegner's granulomatosis, polyarteritis nodosa and giant cell arteritis), sarcoidosis, neoplasia (primary and metastatic tumours of breast, lung, prostate or kidney),

For lymphoma, arteriovenous fistula and malformation.[13] with moderate to severe clinical presentation, oral prednisone can be started with an initial dosage of 60 mg to 100 mg per day for 1 to 2 weeks followed by tapering, depending upon the clinical response.[13,14] If optic nerve dysfunction occurs intravenous corticosteroid should be initiated.[16] As additional therapy orbital irradiation may be useful especially when there is no clinical improvement after 2 weeks of adequate therapy or steroid-dependent patients, or a serious adverse reaction to steroid. Since recurrence / resistance is common other immunosuppressive agents have been used.[13,14] Cyclophosphamide, chlorambucil, mycophenolate mofetil, methotrexate are being used in refractory cases.[13,14] For the patients with severely progressive and disabling clinical course (e.g., orbital apex syndrome with optic nerve compression) or when the lesion is focal and easily approachable surgical debulking is indicated.[13] In our case, we lost the follow up of the patient due to the current pandemic situation.

Mucoceles of the paranasal sinuses were first described by Langenbeck (1820) under the name of hydatides.[17] Rollet (1909) suggested the name mucocoele.[17] Mucocele of a paranasal sinus is an accumulation of mucoid secretion and desquamated epithelium within the sinus with distension of its walls. Mucoceles can cause bone erosion and displacement of surrounding structures. The proximity of mucoceles to the brain may cause morbidity and potential mortality, if left without intervention.[18] Most commonly involved is the frontal sinus, whereas sphenoid, ethmoid, and maxillary mucoceles are rare.[19] The aetiology of mucoceles is multifactorial like inflammation, allergy, trauma, anatomic abnormality, previous surgery, fibrous dysplasia, osteoma, or ossifying fibroma. An important finding is the obstruction of natural ostia which in turn impairs the drainage of sinus. As the sinuses are in close relation to orbit and brain, mucoceles of the paranasal sinuses can spread both intraorbitally and intracranially.[20,21] CT and MRI can clinch the diagnosis of mucocoele. CT helps in determining the anatomy and extent of the lesion, specifically the intracranial extension and the bony erosion. MRI can differentiate mucoceles from neoplasms via contrast enhancement.[22] The mainstay of management of mucoceles is surgical which ranges from functional endoscopic sinus surgery to craniotomy, and craniofacial exposure, with or without obliteration of the sinus.[22] The goal is to drain the mucocele and ventilate the sinus along with eradication of the mucocele with minimal morbidity and prevention of recurrences.[23]

The main causes of acquired third nerve palsy include infections, central nervous system (CNS) or local; trauma, direct or indirect compression of the nerve anywhere along its path; vascular conditions, (ischemic / aneurysms); and neoplastic, inflammatory or demyelinating diseases.[24,25]

Intracranial aneurysms are the common causes for isolated oculomotor nerve palsy. The aneurysms in the posterior part of the Circle of Willis, posterior communicating aneurysms and basilar tip aneurysms have the highest rates of ruptures between 2.5 - 50 %, depending upon their size. Thus, these aneurysms are most commonly associated with isolated third nerve palsy.[24,26] The knowledge of associated syndromes based on different location can be helpful in management.

Sixth nerve palsy may be a false localizing sign in patients with increased intracranial pressure, and may be unilateral or bilateral. Various conditions can result in increased intracranial pressure like tumour, brain abscess, meningo-encephalitis, subdural hematoma, intracranial haemorrhage, ruptured aneurysm, and hydrocephalus. The causes should be obvious on neuroimaging or with spinal fluid analysis. The key finding on physical examination is papilledema. In our case, after treating the patient with systemic steroids, papilledema completely disappeared within one month and the patient became asymptomatic.

CONCLUSIONS

An appropriate and prompt neuroimaging helps in early intervention and prevention of sight threatening situations. Awareness of the association between neurological plus clinical findings in ophthalmoplegia with the anatomical structures which are involved is pivotal.

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Disclosure forms provided by the authors are available with the full text of this article at jemds.com.

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