

# Total Ophthalmoplegia – A Series of Case Reports

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## Abstract

Total ophthalmoplegia is defined as total paralysis of all the muscles of the eye, which in turn results in ptosis, immobility of the eye, dilated non reacting pupil and total loss of accommodation. In this case series the fungal sinusitis and total ophthalmoplegia case ultimately presented with auto evisceration of the crystalline lens, whereas the detailed ophthalmic evaluation finally diagnosed the case of nasopharyngeal carcinoma. The prognosis of fungal sinus infection, nasopharyngeal and maxillary carcinoma is always bad and a high index of suspicion is essential in such cases to reach the final diagnosis.

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## Introduction

Ophthalmoplegia means paralysis or weakness of one or more of the muscles that control the eye movements.<sup>1</sup> External ophthalmoplegia refers to the paralysis of extra ocular muscles. Whereas internal ophthalmoplegia refers to the paralysis of iris and the ciliary muscle.<sup>1</sup> Total ophthalmoplegia thus refers to the paralysis of all the muscles in the eye, which in turn results in ptosis, immobility of the eye, dilated non reacting pupil and loss of accommodation.<sup>1</sup> The condition may be myopathic, that means the muscles controlling eye movement are directly involved or neurogenic, that means the nerve pathways controlling eye muscles are affected.<sup>2</sup> Inter nuclear ophthalmoplegia, a disease which affects the nerves, is a major cause of total ophthalmoplegia.<sup>3</sup> This disorder is usually caused by multiple sclerosis.<sup>3</sup> Diseases of sinuses (fungal & cancerous) and nasopharyngeal carcinoma may spread to the orbit and thus causes total ophthalmoplegia.<sup>4</sup> The main presentation of total ophthalmoplegia is progressive limitation of eye movements with drooping of the eyelids (ptosis).<sup>5</sup> Eye movement disorders and ophthalmoplegia can also be seen with progressive supranuclear palsy, thyroid disease, diabetes mellitus, brainstem tumours, migraine, basilar artery stroke, pituitary stroke, myasthenia gravis, muscular dystrophy, and the Fisher variant of Guillain-Barré syndrome.<sup>6</sup> A tumour or a ruptured aneurysm in the cavernous sinus is usually presented with painful ophthalmoplegia. Painful ophthalmoplegia can also be seen in cases of Tolosa-Hunt syndrome.<sup>7</sup> Lesions of the cavernous sinus or orbit causes simultaneous injury to cranial nerves III, IV, VI as well as to the sympathetic nerves of the iris and to the ophthalmic distribution of the trigeminal nerve and thus presented with total ophthalmoplegia.<sup>8</sup> The occurrence of ocular nerve paralysis in a diabetic patient is always a dilemma from diagnostic, prognostic and therapeutic points of view. When the ophthalmoplegia is a manifestation of diabetic neuropathy, then a benign self-limiting course can be expected.<sup>9</sup> The prognosis of invasive aspergillosis or other fungal infections of the nasal sinuses involving the orbit is disastrous and usually fatal.<sup>10</sup> Death may occur due to cerebral infarction, probably due to

fungal thrombosis of the middle cerebral artery<sup>11</sup>, despite repeated local debridement followed by orbital exenteration and administration of antifungal agents.<sup>12</sup> Experience with these cases strongly indicates the necessity of prompt surgical eradication.<sup>13</sup> Ophthalmic along with neurologic manifestations may be the first and sole presenting feature of nasopharyngeal carcinoma<sup>14</sup> and may miss guide the doctor to make a final diagnosis. Hence a high index of suspicion is very much important in such cases. Ptosis and Horner's syndrome could be a part of neuroophthalmic manifestations of malignant nasopharyngeal carcinoma and thus prompting the patient's initial presentation to the ophthalmologist.<sup>15</sup> Squamous cell carcinoma of nasopharynx are said to be commonest variety.<sup>16</sup> They spread to the paranasal sinuses and orbital tissues causing multiple cranial nerve palsies and proptosis. Infiltration to the cervical sympathetic chain and cervical lymph node ultimately leads to Horner's syndrome.<sup>16</sup> In various studies it has been seen that the cranial nerve serving extra ocular muscles (CN 3,4,6), corneal sensation (CN 5) and orbicularis ocular muscle (CN 7) are affected in 20% of the cases, while optic nerve is affected in 15% of the cases of nasopharyngeal carcinoma.<sup>17</sup> Cranial nerve eight (CN 8) and hypoglossal nerve (CN-12) are affected in 10% of cases of such cases.<sup>17</sup> Maxillary carcinoma also involves the orbital apex, cavernous sinus, optic nerve, middle cranial fossa and presents as a case of total ophthalmoplegia.<sup>18</sup> Even after tumour resection and subsequent radio therapy, ophthalmoplegia, optic neuropathy and trigeminal neuropathy may progress in maxillary carcinoma.<sup>19</sup>

## Diagnosis

The patient's medical history, family history and detailed examination usually help to differentiate the various causes associated with ophthalmoplegia. In addition Computed tomography scan (CT scan) or magnetic resonance imaging (MRI) scan of the brain, orbit and sinuses are needed to rule out underlying sinus pathology, brain tumour, stroke, aneurysm, or multiple sclerosis. Other tests like hearing test (audiogram) and the Tensilon (edrophonium) test should be done in suspected cases. Thyroid disease and diabetes mellitus should be excluded by appropriate blood test.

**Treatment**

Specific treatments are available for multiple sclerosis, myasthenia gravis, diabetes mellitus, and thyroid disease. Surgical procedures can lift the drooping eyelids and patching over one eye can be used to relieve double vision. Dry eyes and exposure keratitis must be taken care of using artificial tears and lid tapping at night. Specific treatment for specific cause should be applied.

**Case Based Discussion**

**Case - 1 (Nasopharyngeal carcinoma & total ophthalmoplegia)**

31 years old female patient was presented to our outpatient department (OPD) with forward protrusion of right eyeball, drooping of her right eyelid and angle of mouth deviation to left side for last 7 months [Figure-1]. She came from a far island in Andaman & Nicobar Island. Deviation of mouth and drooping of eyelid was sudden in onset and not associated with any pain. There was no history of any fever and was not associated with systemic illness like diabetes mellitus, thyroid disease, malignancy and hypertension. She had gradual loss of her weight for last few months with hearing defect in the right side. On examination there was perception of light (PL) negative in right eye and 6/6 in the left eye. Digitally right eyeball was stony hard and intraocular pressure in left eye was 14mmHg. There was total loss of 3rd, 4th and 6th [Figure-2] cranial nerve function with complete ptosis in the right side. Pupil was fixed and dilated [Figure-2] in right eye whereas in left side it was normal. Anterior segment examination showed leukomatous opacity with vascularisation [Figure-1] involving lower part of right cornea. Conjunctiva was congested and chemosed. Anterior chamber was found normal. Corneal sensation was found absent in the right eye and normal in left eye. Fundus examination was normal in both the eyes.



**Figure 2:** Total ophthalmoplegia

Her blood pressure was (130/82mmHg) and there was cervical lymphadenopathy. Cardiovascular, respiratory and per abdomen examination was all appeared normal. On blood investigation sugar was found normal in range [random-120mg/dl]. Complete haemogram showed Hb-9.5, WBC-11000, neutrofil-66, lymphocyte-30, monocyte-04, ESR-36. NCCT brain and orbit showed neoplastic mass lesion in nasopharynx extending into the both nasal cavity, posteriorly into the prevertebral space, inferiorly into the pyriform fossa with intracranial extension [Figure-1]. There was bony erosion noticed in the petrous part of temporal bone [Figure-1]. The homogenous mass was invading optic foramina, optic nerves and optic chiasma. [Figure-1] The mass lesion was also invaded the sphenoidal sinus with evidence of bony erosions [Figure-1]. Fine needle aspiration biopsy from the cervical lymph node showed poorly differentiated metastatic lesion. Lubricating drops and lid tapping was advised. She was finally referred to the higher centre where she was underwent nasopharyngeal tumour resection along with orbital exenteration.

**Case – 2 (Sino-orbital mucormycosis & total ophthalmoplegia)**

64 years old male patient, a known case of type II diabetes and hypertension for last 15 years was presented to our OPD with complain of deviation of angle of mouth to the left side with drooping of his right upper eyelid. [Figure-3] There was mild proptosis and swelling of right sided face for last 15 days. There was no history of any pain. Incident was sudden in onset and associated with mild graded fever. On examination distance visual acuity was 6/60 in right eye with no improvement on pinhole, whereas in the left eye vision was 6/12. Intraocular pressure was 14mmHg in each eyes. Near vision was found N12 in both the eyes. Blood pressure was 130/80mmHg and urgent blood sugar was found 190mg/



**Figure 1:** Nasopharyngeal carcinoma

dl. Pulse rate was 130/min (regular). Extra ocular movement was found limited in all the direction of gazes in the right eye [Figure-4] along with presence of complete ptosis. Cornea had punctate lesions inferiorly with loss of corneal sensation [Figure-3]. Anterior chamber was found normal in both the eyes. There was fixed dilated pupil [Figure-3] in the right eye and in the left eye it was normal. Fundus examination was found normal in both the eyes. A diagnosis of right sided total ophthalmoplegia along with Bell's palsy and exposure keratitis was made and the patient was started on topical lubricating drops with lid tapping. On investigation there was hyperglycemia (350mg/dl), hyperkalemia, azotemia and neutrophilic lymphocytosis with raised erythrocyte sedimentation rate (ESR-35). He was admitted under medical supervision and was started managing with empirical antibiotics, anti-fungal and insulin. NCCT brain, orbit and sinuses showed marked mucosal thickening of all the sinuses of both the sides with full of inflammatory exudates with bony erosions [Figure-3]. Inflammatory lesions were extending upto the right orbital apex [Figure-3] causing proptosis. No evidence of cavernous sinus thrombosis was found and brain parenchyma appeared normal in the imagining studies. ENT opinion was obtained and endoscopic surgical debridement of sinuses was performed. Sample was sent for histopathological examination and was found multiple aseptatehypae, septate hypae and yeast like cells with pseudohyphae. Treatment continued with systemic anti-fungal, antibiotics and insulin. Topical lubricating and antibiotic drops were added. Patient was on irregular follow up with us with irregular medications. After 3 months he presented with perforated cornea with auto-evisceration of crystalline lens [Figure-3]. He was underwent evisceration followed by artificial eye implantation [Figure-3]. Exposure keratitis and associated dry eyes probably leads to corneal ulcer with subsequent perforation in this case and thus leads to auto-evisceration of crystalline lens.

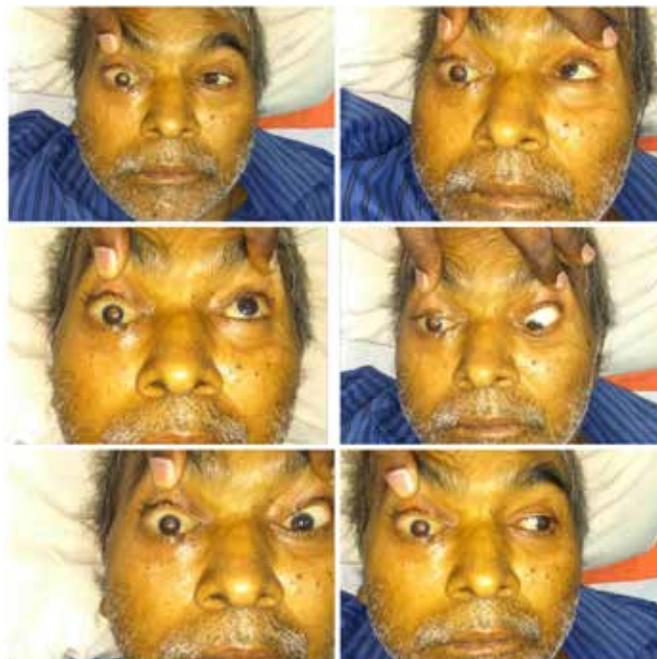


Figure 4: Total ophthalmoplegia

**Case-3 (Maxillary carcinoma & total ophthalmoplegia)**

60 years old male patient, a known case of diabetes mellitus for last 10 years and on oral hypoglycaemic agent was presented with complete dropping of the left upper eyelid , angle of mouth deviation along with difficulty in swallowing for last 20 days.[Figure-5] Patient was slightly drowsy and there was no associated pain and fever. Urgent blood (GRBS) sugar was found 475mg/dl. Blood pressure was 120/70 mmHg and pulse was 77/min (regular). On ophthalmic examination distance visual acuity was 6/36 in both the eyes with dense cataract. Intraocular pressure was 12 mmHg in each eyes. There was complete ptosis in the left eye with Bell's palsy [Figure-5]. Extra-ocular movements were restricted in all the directions of gaze with fixed dilated pupil in left eye [Figure-6]. Rest all the anterior segment examinations were found normal except for loss of corneal sensation in the left eye. Detailed fundus examination was normal in both the eyes. On investigation haemoglobin (Hb) was found-7.5gm%, neutrophil-67, lymphocyte-30, monocyte-03 with raised ESR - 36. Blood sugar (F) was-290mg/dl, sugar (PP)-310 mg/dl. Urine routine examination showed protein excretion in urine. Urea and creatinine was found normal. NCCT brain, orbit and para nasal sinus (PNS) revealed maxilloethmoidal tumour involving the left orbital apex, cavernous sinus, optic nerve, and middle cranial fossa with evidence of bony erosions [Figure-5]. MRI brain and PNS showed maxillary hypo dense mass likely malignancy [Figure-5]. He was then referred to higher centre where he was underwent surgical treatment and found to have maxillary ameoblastic carcinoma- a rare tumour.

**Prognosis**

The prognosis of progressive external ophthalmoplegia depends on the associated neurological problems; in



Figure 3: Sino-orbital mucormycosis

particular, whether there is associated severe limb weakness or cerebellar symptoms. Whereas prognosis for fungal sinus infection, nasopharyngeal and maxillary carcinoma is quite challenging.<sup>20</sup>



Figure 5: Maxillary carcinoma



Figure 6: Total ophthalmoplegia

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