ORIGINAL RESEARCH PAPER

Ophthalmology

Volume - 7 | Issue - 2 | February - 2017 | ISSN - 2249-555X | IF : 3.919 | IC Value : 79.96

Analysis of clinical profile and outcome in patients with multiple cranial nerve palsies in ophthalmology practice.

KEYWORDS: multiple cranial nerve palsies, ophthalmoplegia.

Dr. Rejitha Chinnu Varghese
Final Year Post graduate Department of Ophthalmology Yenepoya Medical College.

Dr. Rajarathna Hegde
Second Year Post graduate Department of Ophthalmology Yenepoya Medical College.

Dr Tariq Ahmed
Second Year Post graduate, Department of Ophthalmology, Yenepoya Medical College.

Dr. Sithara Sara Oomman,
Second Year Post graduate, Department of Ophthalmology, Yenepoya Medical College.

Dr. Rashmi S
Assistant Professor, Department of Ophthalmology, Yenepoya Medical College.

ABSTRACT Patients with multiple cranial nerve palsies pose a unique diagnostic challenge. They need to be carefully evaluated neurologically, ophthalmologically, systemically and radiologically for a prompt and accurate diagnosis. This can be an overwhelming task because locations and causes of multiple cranial nerve involvement are highly diverse. Tumour, vascular disease, trauma, infection, and Guillain-Barre/ Fisher syndromes are the most frequent causes. We report and analyse the clinical presentation, the course in hospital, management and outcome of seven patients who presented with diminished vision, external ophthalmoplegia, ptosis and lagophthalmos.

Introduction

The locations and causes of multiple cranial neuropathies are highly diverse. They can be involved unilaterally or bilaterally and simultaneously or sequentially. Tumour, vascular disease, trauma, infection and Guillain-Barre/ Fisher syndromes are the most frequent causes. In the largest series, neoplasms accounted for 30% of cases.[1] CT, MRI, blood and spinal fluid studies are all important diagnostic tests in the workup.

We report seven patients with multiple cranial neuropathies who presented with diminished vision, external ophthalmoplegia, ptosis and facial weakness. (cranial nerves 2th, 3rd, 4th, 5th, 6th and 7th involvement). And in addition dysphagia and dysarthria was also seen in two patients. (9th and 10th cranial nerves involvement)

Methodology:

This study is an analysis of patients who presented or were referred to ophthalmologist with diminished vision, external ophthalmoplegia, ptosis and lagophthalmos in a tertiary care hospital. All patients seen during the period of one year from January 2016 to December 2016 were included. Written informed consent was obtained from all the patients for reporting their case and any accompanying images. The clinical presentation and outcome of each patient was retrospectively studied. All the patients were admitted for evaluation and treatment. Detailed ENT, ophthalmological and neurological examination was done. MRI was done in all patients. CT was done in two patients. Patients who had clinical and radiological evidence of sinusitis underwent diagnostic nasal endoscopy and biopsy specimen was sent for microbiological and histopathological analysis (three patients).

Results: Out of 7 cases, 4 were female and 3 were male. The age of the patients ranged from 50- 65 years. The clinical features of the patients are described in Table 1.

Table1

	Case 1	Case 2	Case 3	Case4	Case 5	Case 6	Case 7
Cause	Aspergillosis	Mucormycosis	Chemical Injury	GB syndrome	Craniofacial trauma	Metastatic breast carcinoma	Mucormycosis
Age in years	63	55	50	65	55	60	58
Gender							
Comorbidity	Diabetes	Diabetes	Nil	Hypertension	Nil	Breast carcinoma	Diabetes
Diminished vision	present	present	present	present	present	present	present
Ophthalmoplegia, ptosis, lagophthalmos	present	present	present	present	present	present	present
Proptosis	absent	absent	present	absent	absent	present	present
Dysphagia, dysarthria	absent	present	absent	present	absent	absent	absent
Cranial Nerves involved	2,3,4,5,6,7	2,3,4,5,6,7,9,10	2,3,4,5,6,7	2,3,4,5,6,7,9,10	2,3,4,5,6,7	2,3,4,5,6,7	2,3,4,5,6,7
Laterality	ipsilateral	bilateral	ipsilateral	bilateral	ipsilateral	ipsilateral	Ipsilateral to bilateral
Time of involvement	simultaneous	simultaneous	simultaneous	sequential	simultaneous	simultaneous	sequential
Outcome	death	recovered	recovered	recovered	recovered	Lost to follow up	death

Volume - 7 | Issue - 2 | February - 2017 | ISSN - 2249-555X | IF : 3.919 | IC Value : 79.96

- Case1: Sixty three year old male with uncontrolled diabetes, dyselectronemia, renal failure, COPD, pulmonary TB, sinusitis and ophthalmoplegia. Right sided 2,3,4,5,6 and 7th cranial nerves were involved. Nasal biopsy revealed Aspergillosis. Patient's condition deteriorated rapidly, he developed pulmonary edema and died.

- Case 2: Fifty five year old female with uncontrolled diabetes presented with difficulty in swallowing, hoarse voice, sinusitis and right painful ophthalmoplegia. However there was no proptosis. Cranial nerves involved were right sided 2,3,4,5,6,7,9 and 10. Nasal endoscopy showed black eschar on turbinates and microbiological analysis revealed Mucormycosis. Patient recovered after treatment with intravenous Amphotericin.

- Case 3: Fifty year old lady developed right ear gangrene, right sided facial palsy and painful ophthalmoplegia after fall of alkali on right ear (Figure 1). Cranial nerves involved were right sided 2,3,4,5,6 and 7. MRI showed soft tissue thickening involving right temporal region, infratemporal fossa, maxillary region, external auditory canal, middle ear cavity, mastoid, and erosion of facial canal. Soft tissue thickening was also noted along the right peri-orbital region extending to and narrowing the orbital apex (compressing optic nerve) and cavernous sinus(Figure 2).She underwent debridement and skin grafting for pinna. Cranial nerve involvement in this case was due to inflammatory edema. She recovered after intensive steroid therapy.

Figure1

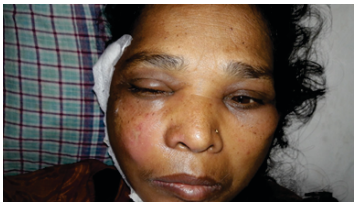
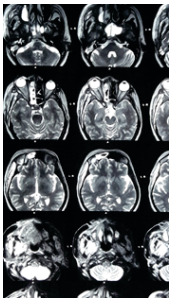


Figure2



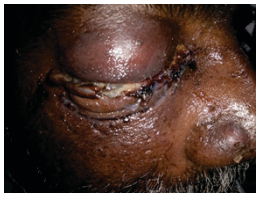
- Case 4: Sixty five year old female presented with slurred speech, difficulty in swallowing, reduced vision and giddiness. Cranial nerves involved- bilateral 2,3,4,5,6,7,9 and 10. She was diagnosed to have a variant of Guillain-Barre syndrome. She recovered completely after five cycles of plasmapharesis.

- Case 5: Fifty five year old male patient met with road traffic injury and sustained craniofacial trauma. He developed left eye ophthalmoplegia, ptosis, diminished vision and facial weakness. Cranial nerves involved were left sided 2, 3, 4, 5, 6 and 7th. Etiology was inflammatory edema. He recovered after steroid therapy.

- Case 6: Sixty year old female on palliative treatment for breast carcinoma presented with painful right sided proptosis and ophthalmoplegia. Imaging revealed metastasis in to orbit. Cranial nerves involved were left sided 2,3,4,5,6th. She requested to be discharged and was lost to follow up.

- Case 7: 58 year old male patient with uncontrolled diabetes had pansinusitis, painful right ophthalmoplegia, proptosis. Patient had rhino orbital cerebral Mucormycosis. (Figure 3). Nasal endoscopy revealed black eschar. Histopathology confirmed Mucormycosis. Cranial nerves involved were initially right sided 2, 3, 4, 5, 6 and 7th. Later even the left optic nerve and 6th nerve were involved. Inspite of treatment with Amphotericin and surgical debridement of involved tissues, the patient did not survive.

Figure 3



Discussion:

Patients presenting with multiple cranial neuropathies are not uncommon. The evaluation and management of these patients can often be overwhelming and challenging. Because the aetiologies are vast and complicated as well as have the potential for devastating outcomes.[2]

More than one ocular motor nerve can be either unilaterally or bilaterally. Before making a diagnosis of nerve paralysis, it is important to exclude disorders like ocular myopathies and myasthenia (muscle or neuromuscular junction). Once diagnosis of multiple ocular motor nerve paralysis is done, next important step is localisation. Dysfunction of the cranial nerves can occur anywhere in their course from intrinsic brainstem dysfunction (intra medullary) to their peripheral courses (extra medullary). Although brain stem contains all of the ocular motor nerves and their nuclei, it is extremely rare for only cranial nerves to get involved without the adjacent brain stem parenchyma being involved (with resultant disorder of supra nuclear motility and motor or sensory dysfunction). Hence when these signs are absent, the lesion is likely to be due to infra nuclear causes. Common locations for multiple infra nuclear nerve involvement are the subarachnoid space and the cavernous sinus- superior orbital fissure.[3]

Superior orbital fissure syndrome involves dysfunction of 3rd, 4th, 6th and first division of 5th cranial nerve. It presents with ophthalmoplegia, ptosis and loss of corneal sensation. Orbital apex syndrome involves dysfunction of 2nd, 3rd, 4th, 6th and first division of 5th cranial nerve. It presents with ophthalmoplegia, ptosis, loss of corneal sensation and optic nerve involvement. Cavernous sinus syndrome involves dysfunction of 3rd, 4th, 6th, first and second divisions of 5th cranial nerve. When the pathology spreads to skull base and parapharyngeal fossa, 9th and 10th cranial nerve can get involved. From here it can spread to retro parotid space and affect 7th nerve. Alternatively involvement of infra temporal fossa and parotid region can directly involve 7th nerve.

The aetiology of multiple cranial neuropathies can range from the relatively benign and treatable to malignant and life-threatening. The differential diagnosis is extensive, with the patient's age, immunocompetence, and tempo of progression all key considerations for clinical evaluation. In the largest series, cancer accounted for 30% of cases.4 Neoplastic processes are an important cause of multiple cranial neuropathy. Cranial nerve palsies can occur in metastatic carcinoma, due to involvement of skull base usually occurring late in the course of the disease.5 We had one patient with metastatic breast carcinoma in our series.

Aspergillosis and Mucormycosis are the commonest fungal infections causing cavernous sinus syndrome and leading to nerve palsies. Aspergillosis arises most commonly as a result of

Volume - 7 | Issue - 2 | February - 2017 | ISSN - 2249-555X | IF : 3.919 | IC Value : 79.96

haematogenous spread and occasionally by direct extension of infection from the paranasal sinuses, middle ear, or orbit. Mucormycosis can be a fulminant infection in immunocompromised and diabetic patients. After inhalation into the nasal cavity and paranasal sinuses, the Mucor fungi cause necrotizing vasculitis, thrombosis, or infarction of the nose and sinuses and can then rapidly extend into the orbits, deep face, and cranial cavity. The central nervous system may be invaded directly by extension through the skull base or indirectly through involvement of the carotid artery and cavernous sinus.[6] In this case series three patients had fungal infection; all three had uncontrolled diabetes. One patient survived after treatment with intravenous Amphotericin.

Trauma is another important cause. In our series we had one patient with mechanical injury and one with chemical injury who had inflammatory edema leading to cranial nerve palsies. Wegener's granulomatosis and other vasculitis can be the cause. Guillain-Barre syndrome and its variant Fisher syndrome can lead to multiple cranial nerve involvement. Thus a large number of pathologic processes initially are manifested by cranial-nerve dysfunction. CT, magnetic resonance imaging, radionuclide scanning, and blood and spinal fluid studies are all important diagnostic tests in the workup of multiple cranial neuropathy.[5]

Conclusion:

Patients with multiple cranial nerve palsies are not uncommon; however the management can be an overwhelming task. Patients need to be carefully evaluated neurologically, ophthalmologically, systemically and radiologically for a prompt and accurate diagnosis. MRI is the imaging modality of choice to identify the location and cause. If the cranial nerve involvement is due to any inflammatory causes, recovery is good with steroids. Neoplasms and fungal infections can be fatal. High index of suspicion is required for early diagnosis of Mucormycosis especially in diabetic patients and it needs prompt treatment with Amphotericin along with surgical debridement.

References:

1. Keane JR. Multiple cranial nerve palsies: analysis of 979 cases. Arch Neurol. 2005; 62(11):1714-7.
2. Carroll CG, Campbell WW. Multiple Cranial Neuropathies. Semin Neurol. 2009; 29(1):53-65.
3. Albert DM, Jakobiec FA. Third, Fourth and Sixth Nerve lesions and Cavernous Sinus. In Principles and Practice of Ophthalmology. 3rd ed; 2008: 4071-95.
4. Probasco JC, Munchel AT, McArthur JC, Blakeley JO. Clinical Reasoning: Multiple cranial neuropathies in a young man. Neurology. 2001; 80(6): 60–66.
5. Beal MF. Multiple Cranial-Nerve Palsies- A Diagnostic Challenge. N Engl J Med 1990; 322:461-63.
6. Lee JH, Lee HK, Park JK, Choi CG, Suh DC. Cavernous Sinus Syndrome: Clinical Features and Differential Diagnosis with MR Imaging. Am J Roent 2003; 181:2, 583-90