Chapter

4

Cranial Nerve Examination

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INTRODUCTION

In leprosy, a disease with an affinity for nerves, there is sufficient evidence of multiple cranial nerves being affected often. Yet, clinicians seldom gives importance to this part of examination. Nonetheless, it is paramount to examine all the commonly affected cranial nerves in leprosy to rule out their involvement, more so those which exit through bony canals where they can be compressed, especially during a lepra reaction. This can cause a great amount of function loss which may even be irreversible. Furthermore, these nerves supply vital structures of the body, the dysfunction of which results in enormous functional crippling. It is even shown that the manifestations of cranial nerve involvement may resurface even after completion of MDT. In this chapter, we have attempted to fill this lacuna by describing the examination of commonly affected cranial nerves in a simple and practical manner.

CURRENT KNOWLEDGE ON THE INVOLVEMENT OF CRANIAL NERVES IN LEPROSY

In leprosy, cranial nerve involvement is seen in about 10–17% of patients.[1] The infection can reach the nerves via direct exposure or through blood or lymph.[2] While it has been observed that cranial nerve involvement most commonly affects patients suffering from lepromatous leprosy, it has also been shown to occur in other spectra of the disease. Of the cranial nerves involved in leprosy, the commonly involved three cranial nerves are facial, olfactory and the trigeminal nerves.[3] Facial or trigeminal nerve involvement has commonly been reported in borderline tuberculoid (BT) leprosy, while the olfactory nerve involvement has been observed more frequently in borderline lepromatous or lepromatous (BL, LL) leprosy.[4] Auditory, oculomotor, glossopharyngeal, vagus, spinal accessory and hypoglossal nerves are rarely affected[5] and Optic nerve involvement is extremely rare.[6] The most common sign of trigeminal nerve involvement is hypoesthesia and/or anesthesia, of skin supplied by the maxillary division.[7] Multiple cranial nerve involvement in a single patient has been emphasized in many studies. The cranial nerves reported to be involved are I, V, VI, VII, VIII IX, X, and XII.[8–10] MRI evidence of CNS changes in the brain and spinal cord has also been described in leprosy patients.[11] Cranial nerves with their type and designated Roman numerical are summarized in Table 4.1.

Salient Clinical Aspects of Cranial Nerve Involvement

Involvement of olfactory nerve results into hyposmia/anosmia. Olfactory dysfunction has been shown to affect 40% of patients with leprosy.[12] Inability to identify smell of smoke/leakage of cooking gas/other hazardous gases can be a cause of consequences. The quality of

Table 4.1: Cranial nerves overview		
Type	***Cranial nerves***	
	Roman Numerical	***Names***
Pure sensory	I	Olfactory
	II	Optic
	VIII	Vestibulocochlear
Pure motor	III	Oculomotor
	IV	Trochlear
	VI	Abducens
	XI	Spinal accessory
	XII	Hypoglossal
Mixed	V	Trigeminal
	VII	Facial
	IX	Glossopharyngeal
	X	Vagus

life gets impaired as the food does not taste good with anosmia. The **optic nerve** involvement though rare, this may manifest as vision loss and visual field defects especially in patients with lepra reaction.

The ocular changes in leprosy are more commonly due to the trigeminal and facial nerve involvement.

Trigeminal and Facial Nerve Involvement

It presents as loss of corneal sensation, in addition to the loss of sensation in area supplied by its sensory branches on face, especially by the maxillary branch. Loss of corneal sensation can lead to injury to corneal epithelium and hinder its healing, which may progress ultimately to ulceration, scarring and perforation.[13]

Leprosy affects the facial nerve in various patterns. Zygomatic branch is preferentially involved due to its superficial location, but hematogenous spread of the lepra bacilli within epineural and perineural vessels may also be seen and could be the reason behind skip lesions and selective involvement of the branches of the facial nerve.[14] The facial nerve supplies muscles of facial expression and of which involvement of orbicularis oculi causes lagophthalmos. Combined involvement of V andVII nerves result in corneal xerosis, exposure keratitis, corneal ulceration, scarring, complicated cataract and blindness. Due to persistent corneal anesthesia, development of new ocular complications even after successful completion of treatment has been shown by many studies[15,16] Patients with lepromatous spectrum of the disease and elderly have increased ocular morbidity.[17]

Facial nerve involvement also affects other muscles of facial expression which manifests as facial asymmetry and Bell's palsy, more often in patients with BT leprosy undergoing type 1 lepra reaction.[5,18] In a patient who presents with bilateral facial palsy, leprosy should always be ruled out. Taste sensation from the anterior 2/3rds is carried by the facial nerve and taste impairment has been observed in about 40% of patients with leprosy.[19]

Involvement of Vestibulocochlear Nerve

It can lead to cochlear type of hearing loss as observed by researchers in a substantial number of patients of leprosy.[8,9] Sensorineural hearing loss is more common in lepromatous leprosy with erythema nodosum leprosum (ENL) reaction.[20]

CRANIAL NERVE EXAMINATION APPROACH

Following prerequisites which should be kept in mind before examining the patient.

W – **W**ash the hands properly/Wear gloves
I – **I**ntroduce yourself politely
P – **P**ermission (informed consent taken from patient after explaining the examination)
Privacy to be maintained
Position of the patient (Seated comfortably in a chair at your eye level).

Examination of Olfactory Nerve

It is an unmyelinated nerve sensory nerve covered with olfactory glia transmitting the smell sensation through olfactory pathway. It may directly get affected by as impairment of the olfactory bulb and olfactory receptors by the lepra bacilli [12] or by primary atrophic rhinitis caused due to mucosal thinning and defects in mucosal innervation with olfactory nerve.

Method of Examination

Enquire the patient for: (a) Any difference in ability to smell, (b) Any recent cold/epistaxis, (c) If they can recognize the smells of the common substances (as these may vary according to the region and subset of patients).

After checking for the patency of the nostrils and explaining the procedure, he is asked to close the eyes. Each nostril is tested separately. Place a familiar scent (non-irritating, non-pungent) like cardamom/soap/asafoetida (*hing*)/clove/peppermint below the nostril and ask him/her to identify the scent. Same procedure is to be repeated on the other side. Test is considered positive when he/she is able to identify the smell on both sides.

Optic Nerve Examination

Optic nerve is responsible for transmitting the special sense of vision. Leprosy can rarely cause visual field defects by damage to any part of the visual pathway and defects in the pupillary light reflex: Direct reflex and consensual reflex (Fig. 4.1).

Method of Examination

Examination of this nerve encompasses

1. *Pupillary reflexes:* Light and accommodation reflexes are tested to check both the optic and oculomotor nerves together (**Afferent: Optic; Efferent: Oculomotor)**
   - *Light reflex:* Light is shone into the patient's eye in a dark room (while keeping a card between the eyes) from the side. The examiner observes for pupillary constriction in the same eye (direct light reflex) and other eye (consensual light reflex). Each pupil constricts briskly if normal. The same procedure is repeated on the other eye. The schematic diagram for pathway of light reflex and possible defects are given in Fig. 4.2
   - *Accommodation reflex:* The examiner keeps his finger near nose of the patient (about 18″) and he is asked to focus on it after focussing on a distant object. The reflex comprises of convergence of the eyes along with pupillary constriction.

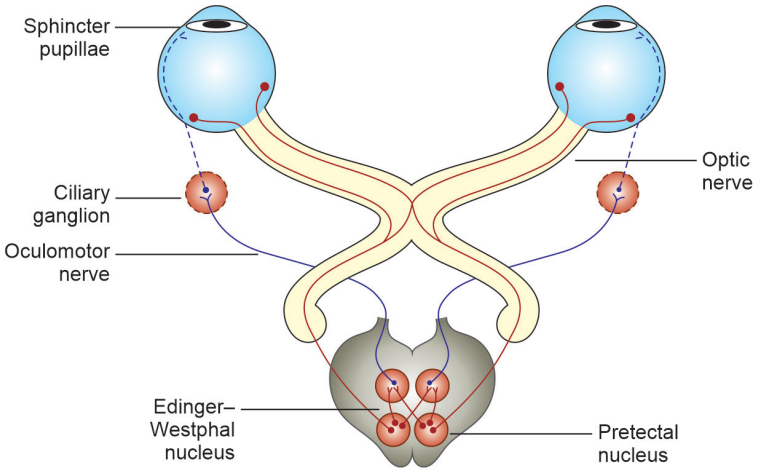

Fig. 4.1: Light reflex pathway

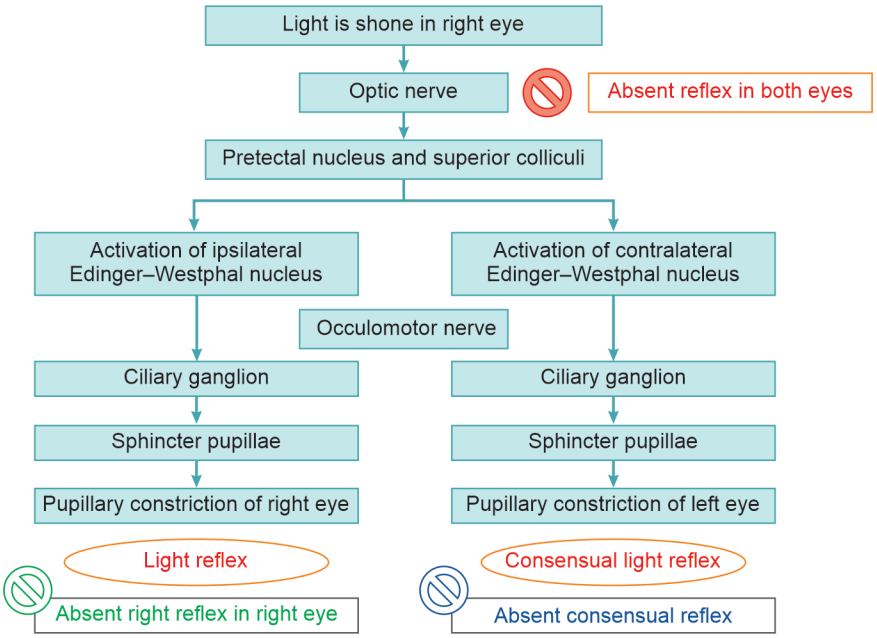

Fig. 4.2: Schematic representation of light reflex pathway

2. *Visual acuity* is the measure of how well the eyes can distinguish shapes and details of objects at a distance. It depends on the intactness of the central retina and the optic nerve. It also depends on the integrity of cornea. It is checked in each eye separately for both near and distant vision. Counting of fingers is simple test to detect gross visual defect for distant vision (start at 1 meter distance. Normal person can count till 6 meter distance). Distant vision can also be checked with the help of standard Snellen's chart. Near vision is checked with the help of Jaeger cards kept at a distance of 1 foot from the patient. In case of severe decrease in acuity, in addition to finger counting, hand movements and light perception are tested to assess the level of blindness.

Oculomotor, Trochlear and Abducens Nerve Examination

All three are mainly motor nerves. They are very rarely involved in leprosy. Oculomotor and trochlear nerves originate in the midbrain whereas abducens originates in the pons and all 3 of them traverse the supraorbital fissure to enter the orbit. Table 4.2 describes their innervations, functions and clinical signs of their damage. Clinical signs of 3rd nerve palsy are illustrated in (Fig. 4.3). The mnemonic LR6 $(SO_4)_3$ helps in remembering the innervation

Table 4.2: III, IVand VI nerves

Cranial nerves	*Innervation*	*Function*	*Clinical signs of damage (Fig. 4.3)*
III	a. Involuntary supply to pupil and lens • Ciliary muscle • Sphincter pupillae	• Controls the shape of lens to focus on closer objects • Constriction of the pupil	• Loss of accommodation • Dilated pupil • Ptosis • "Downward and outward" positioning of the affected eye [due to the unopposed abduction (lateral rectus muscle) (and adduction and downward movement (superior oblique muscle) • diplopia because of misalignment of the eyes
	b. Motor supply • Levator palpebrae superioris • 4 eye muscles (Superior rectus, inferior rectus, medial rectus and inferior oblique)	• Elevates the upper eyelid • Visual tracking and gaze fixation	
IV	Superior oblique muscle	• Forward rotation of the eyeball when the eye is adducted	• Failure to depress the eye during adduction • Diplopia • Difficulty in reading a book/ walking down the stairs • Affected eye is higher and medially deviated because of dominance of inferior oblique
VI	Lateral rectus muscle	• Abducts the eye on the same side	• Inability to abduct the ipsilateral eye • Diplopia and medial strabismus due to the unopposed action of medial rectus

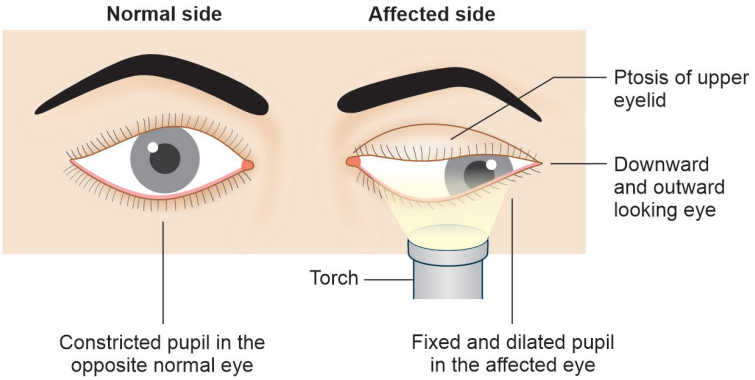

Fig. 4.3: Clinical signs of oculomotor nerve paralysis

by the extraocular muscles. It stands for: LR6: Lateral rectus muscle of eye being supplied by sixth cranial nerve. SO_4: Superior oblique muscle being supplied by fourth cranial nerve. The third (3) cranial nerve innervating the other extraocular muscles.

Method of Examination

The three nerves are additionally tested together for the movements of the eyeball. With the head kept stable, the patient is first asked to look in different directions (approximately corresponding to 12, 2, 4, 6, 8, 10 o'clock positions) on command by following an object kept at about 12" from patient's nose and then moved in the above mentioned directions. Both eyes are observed simultaneously. This can also be observed while doing the visual field mapping. Size of palpebral fissures is additionally observed for any narrowing due to muscle weakness (ptosis) as a result of oculomotor nerve lesion, if any.

Trigeminal Nerve Examination

It provides sensory innervation to the face. After its origin at the junction of pons and the middle cerebellar peduncle, and after its intracranial course, it exits the skull as 3 branches: ophthalmic branch; through superior orbital fissure, maxillary branch through *foramen rotundum* which enters the orbit through infraorbital foramen as infraorbital nerve and mandibular branch through foramen ovale (Fig. 4.4). Details of each branch are described in Table 4.3.

Method of Examination

Sensory examination: The patient is instructed to close his/her eyes and pain, temperature and light touch sensations are examined over forehead and upper side of nose (ophthalmic division), malar region and upper lip (maxillary division) and chin (mandibular division).

- *Corneal reflex:* After explaining the procedure, the patient is asked to fix gaze straight ahead. Then from the side, the lateral margin of cornea is touched lightly with a wisp of cotton. This results in blinking of both eyes. (Afferent—Trigeminal nerve; efferent—Facial nerve) as shown in Fig. 4.5.
- *Blink reflex/orbicularis oculi reflex/glabellar reflex*—The supraorbital ridges are tapped lightly which result in blinking. (Afferent—Trigeminal nerve; efferent—Facial nerve).

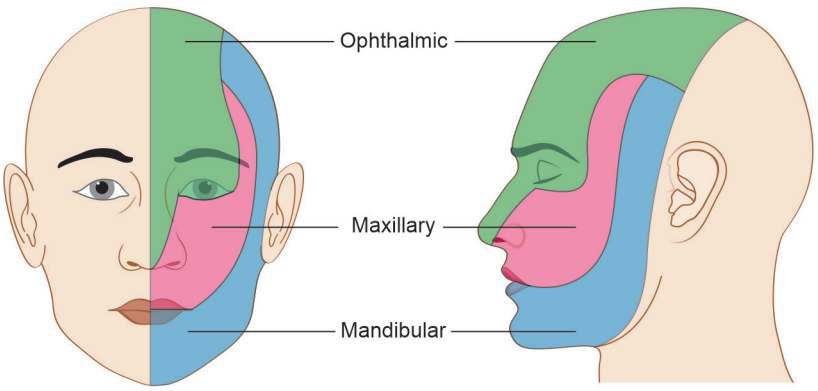

Fig. 4.4: Sensory innervation of trigeminal nerve branches

Table 4.3: Trigeminal nerve

Branches	*Description*	*Branches*	*Innervation*	*Function*	*Effects of damage*
Ophthalmic	• Smallest • Pure sensory	• Lacrimal • Frontal-2 branches – *Supraorbital* – *Supratrochlear* • Nasociliary	These branches supply • Forehead, scalp • Frontal and ethmoidal sinuses • Cornea • Upper eyelid with its conjunctiva • Dorsum of nose	Sensory	• Loss of sensations of the supplied area • Loss of corneal reflex
• Maxillary	• Second division • Pure sensory	• Gives various branches at various levels	• Lower eyelid with its conjunctiva • Cheeks • Maxillary sinus • Lateral nose and nasal cavity • Upper lip • Upper incisors, canines and molars, with the associated gingiva • Superior palate • Parasympathetic supply—nasal as well as lacrimal glands	Sensory	• Loss of sensations of the supplied area
Mandibular	• 3rd and the largest of the 3 • Mixed nerve with motor and sensory branches	• Sensory supply • Motor supply • Parasympathetic supply	• Mucosa and floor of oral cavity • External ear • Lower lip and chin • General sensation over anterior 2/3 of the tongue (taste sensations by branch of facial nerve) • Lower incisors, canines and molars with the associated gingiva • Muscles of mastication: • Digastric muscle-anterior belly • Mylohyoid muscle • Tensor veli palatini • Tensor tympani • To submandibular, sublingual and parotid glands	• Sensory • Help in mastication of food with various movements of mandible • Normal jaw jerk • Involved in elevation of hyoid bone while swallowing • Elevates soft palate and prevents regurgitation of food into nasopharynx • Dampens sounds like that of chewing by stabilizing malleus bone of middle ear	• Loss of sensations of the supplied area • Difficulty in mastication • loss of jaw jerk

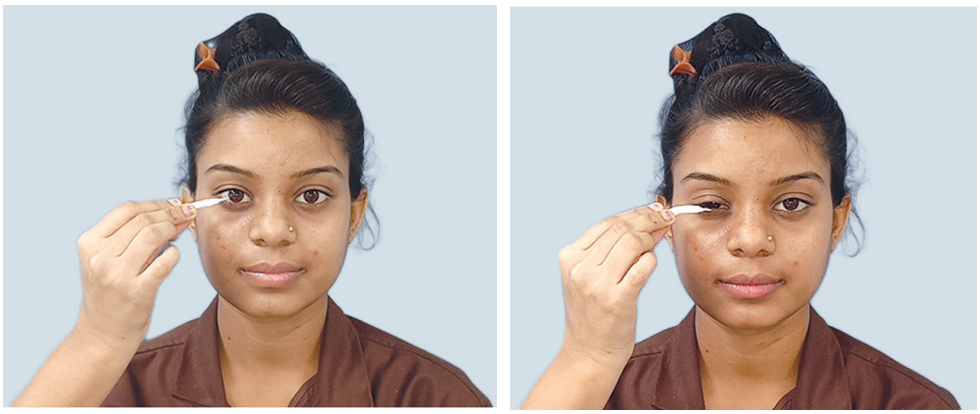

Fig. 4.5: Method of examination to elicit corneal reflex

Facial Nerve Examination

It is a mixed nerve having sensory, motor as well as parasympathetic innervations. It arises from the brainstem and after its intracranial course it comes out of the stylomastoid foramen of the temporal bone and divides into its terminal branches at the posterior edge of the parotid gland. Various branches and functions of facial nerve are described in Table 4.4, the areas supplied by its branches are depicted in Fig. 4.6. The patient is made to sit in good light and is observed for facial symmetry, symmetry of blinking/eye closure and any palsy. *Motor examination*: The patient is asked to raise the eyebrows, close the eyes tightly, frown, puff the cheeks and show the teeth. Any difficulty in doing the actions or asymmetry is noted.

Other cranial nerves are very rarely involved in leprosy and routine testing for their integrity is not needed in leprosy patients.

Table 4.4: VII nerve

Terminal branches	*Functions*		*Effects of damage*
• Temporal • Zygomatic • Buccal • Marginal mandibular • Cervical	• Motor	Posterior belly of the digastric muscle, muscles of facial expression, stylohyoid and stapedius muscles.	Peripheral nerve damage causes: • Ipsilateral paralysis of the muscles of facial expression
	• Sensory	Around the concha	
	• Special sensory	Via chorda tympani—Taste sensation to the anterior 2/3 of tongue	• Taste disturbance in the affected area
	• Parasympathetic	Supplies many glands of head and neck region: Lacrimal glands. Salivary glands: Submandibular and sublingual Nasal, palatine and pharyngeal mucous glands.	• Disturbances in lacrimation and salivation

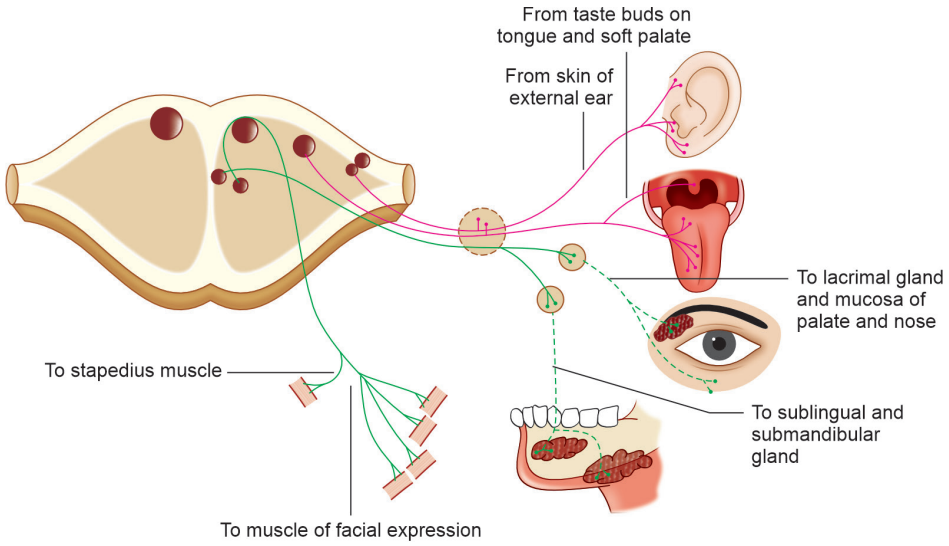

Fig. 4.6: Facial nerve innervation

SUMMARY

In the normal functioning of the human body, cranial nerves play a very essential role, and they can be involved in leprosy. Hence, it should be a routine practice to look for cranial nerve involvement in examining a leprosy patient. Investigations like CT and MRI may sometimes be required for the diagnosis. Patient suspected to have their involvement should be sent for Ophthalmology, ENT and neurology specialist care as needed to tertiary centers.

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