ONE-AND-A-HALF SYNDROME

Eye Learn

All about the Eye

Dr. Krati Gupta

Dr. Saurabh Deshmukh

www.eyelearn.in

One-and-a-half syndrome

The one-and-a-half syndrome is caused by a lesion of unilateral tegmentum of pons, causing damage to the PPRF (or abducens nucleus) and MLF.

Etiology

1. The most common cause of one-and-a-half syndrome was **cerebrovascular disease, and usually was brain stem lacunar infarction,**
2. followed by the **demyelinating etiology (multiple sclerosis),** and
3. Then the infectious cause including **neurocysticercosis and brainstem encephalitis**.
4. Other uncommon causes were **head trauma, brain stem tumor (primary or metastasis), astrocytomas,** etc.
5. Therefore, the vast majority of one-and-a-half syndrome companied by other positioning signs is due to the different lesions of brainstem
6. Only a very small number of patients showed isolated one-and-a half syndrome, while the lesions are smaller and localized, such as brainstem cysticercosis, brainstem tuberculosis, brainstem cavernous hemangioma and the local hemorrhagic infarction of brainstem.
7. **Pseudo–one-and-a-half syndromes may occur with myasthenia gravis, Wernicke's encephalopathy, or Guillain-Barré syndrome.**

Clinical signs include the following:

1. Horizontal gaze palsy on looking toward the size of the lesion ("one").
2. INO on looking away from the side of the lesion ("half"). This paralyzes adduction and causes nystagmus on abduction. As a result, the ipsilateral eye has no horizontal movement, and the only lateral ocular movement that remains is abduction and nystagmus of the contralateral eye.
3. Associated signs include skew deviation, gaze-invoked nystagmus on vertical gaze, and exotropia of the eye contralateral to the lesion.
4. Vertical ocular movements and convergence are usually intact.
5. Pupils are normal
6. Doll's head maneuver fails to elicit normal eye movements.

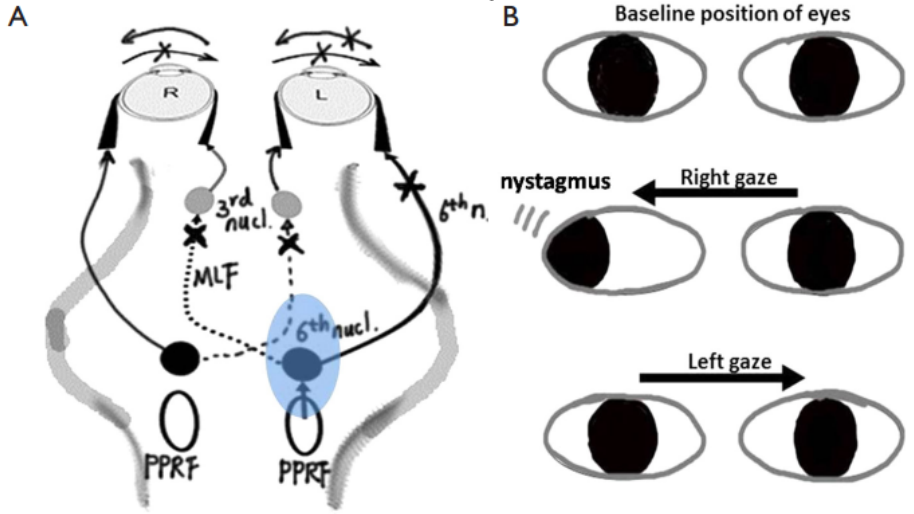

Figure 2 Diagram shows the mechanism for one-and-a-half syndrome. (A) Pathologic lesion (oval shaded area) in the dorsal pontine tegmentum involves both ipsilateral PPRF (or the abducens nucleus) and the ipsilateral MLF; (B) Position of eyeballs, with rightward gaze, the left eye has the impaired adduction, while the abduction of the right eye is intact, except for the nystagmus, and with left gaze, both eyes have impaired horizontal movement (abduction of the left eye and adduction of right eye). ×, interruptions of signal transmission or eye movement that are a result of the lesion; 6th nucl., abducens nuclei; 6th n, ipsilateral abducens nerve (cranial nerve VI); 3rd nucl., oculomotor nuclei.

Dr. Krati Gupta | Dr. Saurabh Deshmukh

Ancillary Testing

MRI detects tumor, myelinolysis, and vascular malformation, but only sometimes detects stroke and multiple sclerosis

Differential Diagnosis

1. Myasthenia gravis
2. Fisher syndrome
3. Wernicke encephalopathy
4. Frontoparietal infarct

Treatment

1. Stroke: supportive care
2. Tumor: search for primary site, lumbar puncture
3. Vascular malformation: radiation or surgery, as appropriate
4. Multiple sclerosis: immunomodulatory agents
5. Fisher syndrome: plasmapheresis or intravenous immunoglobulin
6. Wernicke encephalopathy: thiamine