Background

Cranial nerve VI, also known as the abducens nerve, innervates the ipsilateral lateral rectus (LR), which functions to abduct the ipsilateral eye. The sixth cranial nerve has a long subarachnoid course. The sixth nerve nucleus is located in the pons, just ventral to the floor of the fourth ventricle and just lateral to the medial longitudinal fasciculus (MLF). About 40% of its neurons project into the ipsilateral MLF only to cross over to the contralateral side and ascend to innervate that contralateral medial rectus subnucleus to participate in contralateral eye adduction. [1, 2, 3]

The abducens nerve emerges from the brainstem at the pontomedullary junction to enter the subarachnoid space, coursing upward between the pons and clivus to enter the Dorello canal. At the petrous apex, it angulates to enter the cavernous sinus and travels in close proximity to the internal carotid artery. The abduces nerve then proceeds through the superior orbital fissure and innervates the lateral rectus muscle.

Patients usually present with binocular horizontal diplopia (double vision producing a side-by-side image with both eyes open), worse in the distance, and esotropia in primary gaze. Patients also may present with a head-turn to maintain binocularity and binocular fusion and to minimize diplopia. Congenital sixth nerve palsy (Duane syndrome) is a well-recognized entity.

Examination for a sixth nerve palsy involves documenting the presence or absence of papilledema, examining the ocular motility, evaluating the eyelids and pupils, and excluding involvement of other cranial nerves (eg, V, VII, VIII).

Occasionally checking deep tendon reflexes (DTRs) and motor function to exclude corticospinal tract involvement may be important.

MRI is indicated for any brainstem findings to exclude pontine glioma in children (most have papilledema and nystagmus without other cranial nerve involvement) and in adults who show no improvement.

In young adults, a lumbar puncture (LP) for cerebrospinal fluid (CSF) analysis is completed to exclude meningitis in patients who have no history of diabetes or hypertension and who have a head scan

negative for other pathology. Elderly patients should undergo blood testing for erythrocyte sedimentation rate (ESR), C-reactive protein, and platelets to screen for giant cell arteritis (temporal arteritis). Poor or no resolution of sixth nerve palsy should prompt a full neurologic evaluation.

Pathophysiology

Only the ipsilateral lateral rectus that is solely innervated by the involved peripheral sixth cranial nerve is affected; therefore, only deviations in the horizontal plane are produced. In isolated cases of peripheral nerve lesions, no vertical or torsional deviations are present. Central nervous system lesions of the abducens nerve tract are localized easily secondary to the typical findings associated with each kind of lesion. Damage to the sixth nerve nucleus results in an ipsilateral gaze palsy. The lack of a contralateral adduction defect makes it easy to differentiate a nuclear lesion from a fascicular or nonnuclear lesion. [2]

Abducens palsy can be a false localizing sign with lesions that cause increased intracranial pressure and stretching of the sixth nerve as it ascends the clival area.

Abducens nerve palsy is frequently seen as a postviral syndrome in younger patients and as an ischemic mononeuropathy in the adult population.

Epidemiology

Frequency

United States

Sixth nerve palsies fall into the following categories: 3%-30% trauma, 0%-6% aneurysm, 0%-36% ischemic, 8%-30% idiopathic, and 10%-30% demyelination/miscellaneous.

The sixth cranial nerve is the most commonly affected of the ocular motor nerves. In children, it is the second most common after the fourth nerve, with an incidence of 2.5 cases per 100,000 in the population.

Mortality/Morbidity

A young patient should have an aggressive workup because of the greater likelihood of a neoplasm causing the palsy. Patients older than 55 years with isolated sixth nerve palsies may require a less aggressive initial workup if they have predisposing microvascular ischemic risk factors, but no history of cancer.

Age

Cranial nerve VI palsy can occur in all age groups; however, the etiology varies depending on the age group.

History

Clinical history of abducens nerve palsy includes the following:

- Binocular diplopia (worse at distance or lateral gaze)
- Esotropia
- Head-turn
- Vision loss
- Headache, vomiting, pain, or facial numbness
- Trauma
- Symptoms of vasculitis, particularly giant cell arteritis
- Hearing loss

Physical

Physical findings of abducens nerve palsy include the following:

- An esodeviation that increases on ipsilateral gaze and is often greater at a distance; prism measurements in different positions of gaze can reveal the magnitude of misalignment and its incomitance (asymmetry)
- An isolated abduction deficit
- Slowed ipsilateral saccades
- Papilledema (if increased intracranial pressure)
- Altered sensation in the V1 or V2 distribution with cavernous sinus lesions
- Nystagmus (usually in children, ie, secondary to pontine glioma)
- Otitis media
- Petrous bone fracture

- Tender, enlarged, nonpulsatile temporal arteries in giant cell arteritis
- Horner syndrome (Foville brainstem syndrome, carotid oculosympathetic plexus involvement in cavernous sinus)
- Contralateral hemiparesis may be seen in brainstem syndromes that involve the sixth cranial nerve (Millard-Gubler syndrome and Raymond syndrome)

Causes

Not all abduction deficits are cranial nerve VI palsies. Mimickers are orbital lesions, medial wall fractures, Duane syndrome, thyroid-associated orbitopathy, myasthenia gravis, and spasm of the near reflex. [4]

- Elevated intracranial pressure can result in downward displacement of the brainstem, causing stretching of the sixth nerve secondary to its anatomic location within the Dorello canal. This is believed to be the reason that about 30% of patients with pseudotumor cerebri have an isolated abducens nerve palsy and explains how lesions remote from the sixth cranial nerve can cause abducens paresis (false localizing sign).
- Subarachnoid space lesions can be causes of abducens nerve palsy (eg, hemorrhage, infection, inflammation, space-occupying tumor, cavernous sinus mass). Inflammatory (eg, postviral, demyelinating, sarcoid, giant cell arteritis)
- Vascular
- Metabolic (eg, vitamin B, Wernicke-Korsakoff syndrome)
- Neoplasm (children) - Pontine glioma
- Infectious (eg, Lyme disease, syphilis)
- Congenital absence of the sixth nerve (eg, Duane syndrome) [5]
- Trauma, particularly if it results in a torsional head motion [6, 7, 8, 9]
- Post–lumbar tap [10]

Differential Diagnoses

- Giant Cell Arteritis (Temporal Arteritis)
- Medial Wall Orbital Fracture
- Myasthenia Gravis
- Spasm of the near reflex
- Thyroid Associated Orbitopathy with medial rectus involvement

Laboratory Studies

See the list below:

- Complete blood cell (CBC) count
- Diabetes testing (glucose, glycosylated hemoglobin [HbA1C], glucose tolerance test)
- Erythrocyte sedimentation rate, C-reactive protein, and platelets in patients older than 50 years
- Acetylcholine receptor antibodies in the presence of variable strabismus or ptosis

The following are not mainstream tests for abducens palsy but can be considered:

- Rapid plasma reagin test
- Fluorescent treponemal antibody-absorption test
- Lyme titer
- Antinuclear antibody test

Imaging Studies

MRI is indicated for the following:

- Patients younger than 55 years with no vasculopathic history
- Associated pain or other neurologic abnormality [11]
- History of cancer
- Bilateral sixth nerve palsy
- Papilledema
- In the event no marked improvement is seen or other nerves become involved

An LP should be considered if MRI results are negative.

If a presumed microvascular ischemic sixth nerve palsy does not improve within 3-4 months or if other cranial nerves become involved, a full medical, neurologic, and imaging workup should be performed.

Other Tests

Check history for diabetes mellitus, cancer, thyroid disease, and hypertension.
Ask about history of recent trauma, ear infections (children), and fluctuation of symptoms.
An otoscopic examination may be performed in children to rule out a complicated otitis media (consider an LP).
Rule out other cranial nerve involvement.

Procedures

A temporal artery biopsy may be indicated in patients aged 50 years or older with findings and laboratory studies suggestive of giant cell arteritis.

Medical Care

Truly isolated cases of abducens nerve palsy are often benign. They can be followed with a serial examination, at least every 6 weeks, over a 6-month period to note decreasing symptoms (diplopia) and resolution of the paretic lateral rectus (increasing motility). [12, 3] Prism measurements are a simple objective method of documenting any changes in the esotropia.
Children with sixth nerve palsy who are in the amblyopic age group can be treated with an alternating patching to decrease their chances of developing any amblyopia in the paretic eye. Additionally, prescribing the full amount of hyperopic correction helps to decrease the esodeviation by relaxing the child's accommodative effort.
Adult patients and those children beyond the amblyopic age can be patched or have their lenses "fogged" with clear tape or nail polish to reduce their diplopia. Fresnel prisms also can be prescribed as an alternative.

Older patients in whom giant cell arteritis is suspected should start the standard treatment with prednisone or intravenous methylprednisolone.

Surgical Care

If, after 9-12 months of follow-up care, the remaining deviation is still unacceptable and is too large to be corrected with prisms, surgical corrective options should be discussed with the patient. The procedure that is chosen depends on the remaining function of the lateral rectus and the experience of the surgeon.

If some residual function exists in the lateral rectus, a graded recession of the medial rectus or botulinum toxin to the medial rectus, and resection of the lateral rectus or lateral rectus bupivacaine (Marcaine) injection can be performed.

When little or no residual function is present, a transposition of the vertical recti toward the lateral rectus (eg, Hummelsheim, Jensen, or Nishida procedure), can be considered in conjunction with weakening of the ipsilateral medial rectus.

Consultations

Patients with abducens palsy can benefit from consultation with a neurologist, ophthalmologist, or neuro-ophthalmologist, especially if the lesion does not resolve.

Activity

Patients who occlude an eye to alleviate diplopia should be warned that the resulting effects on depth perception may interfere with their ability to drive or perform certain occupations safely.

Prognosis

Microvascular sixth nerve palsy generally resolves within 6 months.

Overview

What is abducens nerve palsy ?

What is the clinical presentation of abducens nerve palsy ?

What is included in the physical exam for abducens nerve palsy ?

Which tests are performed in the workup of abducens nerve palsy?

Presentation

DDX

Workup

Treatment

Follow-up