Horner's Syndrome

Horner’s syndrome, also called oculosympathetic paresis, is a condition that can be produced by a lesion anywhere along the sympathetic pathway that supplies the head, eye, and neck.

Clinical Features

1. Ptosis.
2. Miosis.
3. Anhydrosis.
4. Enophthalmos.

 Causes

- *Anhydrosis* determines site of lesion.

<table>
<thead>
<tr>
<th>Central lesions</th>
<th>Pre-ganglionic lesions</th>
<th>Post-ganglionic lesions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anhydrosis of the face, arm and trunk</td>
<td>Anhydrosis of the face</td>
<td>No anhydrosis</td>
</tr>
</tbody>
</table>

- Stroke
- Syringomyelia
- Multiple Sclerosis

- Pancoast's Tumour
- Thyroidectomy
- Trauma

- Carotid artery dissection
- Carotid aneurysm
- Cavernous sinus thrombosis
- Cluster headache
Post-Ganglionic Lesions

1) Carotid Artery Dissection

- The most common cause of young-onset stroke (< 40 years). The second most common cause is carotid embolism.
- 75% affect the internal carotid artery.
- Causes: Mostly neck trauma or manipulation.

Clinical Pictures

1- Unilateral headache:

- Headache is commonly ipsilateral to the side dissection.
- Recurrence of the headache suggests extension or recurrence of the dissection.

2- Ipsilateral Horner’s syndrome:

- The Horner’s syndrome occurs as a result of compression of the ascending sympathetic supply within the carotid sheath, resulting in ptosis and miosis.
- Anhydrosis is classically not present as the sympathetic supply to the sweat glands is along the external carotid plexus and is therefore spared.

3- Contralateral hemisphere:

- Transient or complete strokes are found in 30-80% of patients.
- Signs: aphasia, neglect, visual disturbance, and hemiparesis.

Diagnosis

- It can be diagnosed with Contrast arteriography of the neck vessels.

Management

- It is similar to that of acute stroke to prevent cerebral infarction.
- Stenting can be used if there is ongoing ischemia.

*A thrombotic event resulting from cardiac embolism or antiphospholipid syndrome will only affect intracranial vessels; therefore, Horner’s syndrome will be absent.*

2) Carotid Artery Aneurysm

An aneurysm is a bulge or ballooning in the wall of the carotid artery. It is caused when a portion of the artery wall weakens.
Clinical Pictures

1. Transient ischemic attacks (TIAs) or stroke.
2. Symptoms can occur secondary to the pressure of the aneurysm on surrounding structures such as hoarseness or difficulty swallowing.
3. It might rupture and complicate to complete carotid dissection.

Diagnosis

- It can be diagnosed by ultrasound, CT or MRI scans of the neck.
- Follow up every 6 to 12 months is recommended if it is asymptomatic.

Management

- The mainstay of treatment of extracranial carotid artery aneurysms is surgical repair using endovascular or stent-graft repair.

3) Cavernous Sinus Thrombosis

- Cerebral venous thrombosis (CVT) is a thrombotic obstruction of the cerebral venous system and related anatomical structures (as dural sinuses) that may lead to ischemic lesions in the brain.
- Affect all ages, most commonly in women who have risk factors such as prothrombotic conditions, oral contraceptive use, and malignancy.
- Spreading of facial infections as otitis media and sinusitis may result in sinus thrombosis as well.

Clinical Pictures

- In a patient with cavernous sinus thrombosis, he will develop cavernous sinus syndrome, which includes:
  1. Ophthalmoplegia, proptosis and conjunctival congestion.
  2. Sensory loss on the forehead and cheek (trigeminal area)
  3. Horner syndrome.

Diagnosis

- CT/MRI (with or without venography) are tests of choice to confirm the diagnosis.
- Plain CT/MRI detects only brain edema and infarcts, but the thrombosis itself can only be visualized by means of venography.

Management

- Initial management involves treatment of the underlying causes.
- Medical treatment with antithrombotic therapy and antibiotics for infectious causes.
- Surgical Treatment should be offered if there progressive neurological worsening despite adequate medical treatment. Options as blood clot removal and placement of a shunt.
4) Cluster Headache

- It is a type of episodic headache that mostly affects the 20-40 years old adult men.
- It occurs in “cluster bouts” followed by months of remission; however, it may progress into chronic form if remission between bouts lasts less than one month.

Clinical Pictures

- **Headache:**
  - Intense agonizing pain
  - It occurs usually in a cyclical pattern (“clusters”) which lasts between 15 mins - 2 hours.
  - It affects periorbital and/or temporal areas.
  - Always unilateral and affects the same side.

- **Autonomic manifestations:**
  - Conjunctival injections and lacrimation.
  - Rhinorrhea and nasal stiffness.
  - Horner's syndrome: ptosis and miosis, but no anhidrosis.

- Restlessness during an attack.

- **The examination between the attacks should be normal.**

Diagnosis

- The diagnosis of cluster headache is based on his history and physical examination.
- Serious causes should be excluded.

Management

- **Acute:**
  - Oxygen therapy with 100% oxygen.
  - Medical: subcutaneous or intranasal triptans (e.g., sumatriptan).

- **Prophylaxis:**
  - First-line treatment: **CCB as verapamil**
    - Steroids can be used as a bridge until verapamil becomes effective.
  - Second-line treatments: Lithium, Topiramate and Ergot derivatives

Differential Diagnosis

<table>
<thead>
<tr>
<th>Chronic Paroxysmal Hemicrania (CPH)</th>
</tr>
</thead>
<tbody>
<tr>
<td>It has features of cluster headaches BUT:</td>
</tr>
<tr>
<td>- Attacks are shorter in duration with increased frequency, last between 3-45 minutes and occur 20-40 times per day.</td>
</tr>
<tr>
<td>- It responds very well to indomethacin.</td>
</tr>
</tbody>
</table>
Central Lesions

1) Stroke

- Stroke is defined as an acute neurological insult due to a focal cerebrovascular event that can either be a vascular occlusion (ischemic stroke) or rupture of a blood vessel (hemorrhagic stroke).
- The type of stroke that is associated with Horner’s syndrome is Wallenberg syndrome.

Wallenberg syndrome

- Wallenberg syndrome, also known as Lateral Medullary Syndrome, results from occlusion of either:
  - Posterior inferior cerebellar artery (PICA)
  - Branches of the vertebral artery.

<table>
<thead>
<tr>
<th>Clinical Features</th>
<th>Structures affected</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ipsilateral</td>
<td></td>
</tr>
<tr>
<td>• Horner’s Syndrome</td>
<td>• Sympathetic fibers</td>
</tr>
<tr>
<td>• Loss of pain &amp; temperature in face</td>
<td>• Trigeminothalamic tract (sensory neuron)</td>
</tr>
<tr>
<td>• Dysphagia</td>
<td>• Nucleus ambiguus (motor neuron)</td>
</tr>
<tr>
<td>• Hoarseness &amp; dysphonia (due to vocal cord paralysis)</td>
<td>• Inferior cerebellar peduncle</td>
</tr>
<tr>
<td>• Ataxia</td>
<td>• Vestibular nuclei</td>
</tr>
<tr>
<td>• Nystagmus &amp; diplopia.</td>
<td></td>
</tr>
<tr>
<td>• Vetrigo</td>
<td></td>
</tr>
<tr>
<td>• Falling toward the same side of the lesion.</td>
<td></td>
</tr>
<tr>
<td>Contralateral</td>
<td></td>
</tr>
<tr>
<td>• Loss of pain and temperature in the trunk and limbs</td>
<td>• Lateral spinothalamic tract (sensory neuron)</td>
</tr>
</tbody>
</table>

Management

- Management includes the same treatment of other types of strokes.
It is mainly supportive treatment and may include swallowing and speech therapy, and a feeding tube to help in swallowing in some cases.

2) Syringomyelia

- It is the development of fluid-filled cavity (syrinx) within the central canal of the spinal cord that produces progressive myelopathy and neurological manifestations.
- If extends into the brainstem then termed syringobulbia.
- It is associated with the Arnold-Chiari malformation in > 50% of patients.

Clinical Features

Symptoms begin insidiously in adolescence or early adulthood, progress irregularly, and may undergo spontaneous arrest for several years; most patients acquire cervical-thoracic scoliosis.

- May be asymmetrical initially → Discovered accidentally on MRI.
- The syrinx initially compresses the crossing fibers of the spinothalamic tract resulting in:
  - Dissociated sensory loss: Loss of pain and temperature sensations, while the sensation of light touch, vibration, and position remain intact on both sides.
  - Neuropathic arthropathy develops lately.
- Further expansion of the syrinx may damage:
  - Descending hypothalamic fibers in T1 to T4 → Horner’s Syndrome.
  - Lower motor neurons in the corticospinal tract at the level of syrinx → bilateral weakness and muscle atrophy.
  - Corticospinal tracts below the level of the syrinx → spastic paraplegia.
  - Posterior column (in advanced cases) → loss of position sense and vibration sense in the feet.
- Syringobulbia:
  - Tongue atrophy.
  - Loss of pain and temperature sensation in trigeminal nerve
  - Nystagmus
  - Dysphagia
  - Palatal and pharyngeal weakness

Diagnosis

- MRI is the investigation of choice which can visualize the syrinx.

Management

- Conservative therapy should be considered especially in the early stages:
  - Physiotherapy.
  - Analgesia as NSAIDs
- Surgical treatment is recommended for patients with progressive neurological symptoms:
  - Decompression by drainage of CSF fluid from the cavity.
3) Multiple Sclerosis

It is a chronic degenerative immune-mediated disease of the CNS that results in the demyelination of white matter in the brain and spinal cord, and hence UMNL signs are seen.

- It usually occurs in **young adults**, more common in **females**.
- The course is characterized by exacerbations followed by periods of complete/incomplete remission.

- **Multiple sclerosis (MS)** affects the central nervous system only.
- **Subacute combined degeneration (SCD)** of the cord affects central and peripheral nerves.

### Clinical Features

- **Visual**:
  - Optic neuritis: common earliest presenting feature.
  - Optic atrophy
  - Internuclear ophthalmoplegia as a result of a lesion in the medial longitudinal fasciculus.
  - **Uhthoff’s phenomenon**: worsening of vision following physical exertion, a warm bath or fever.

- **Motor**:
  - Pyramidal tract lesion → upper motor neuron weakness characterized by:
    - Spastic weakness: most commonly in the legs (*No hypotonia*).
    - Positive Babinski’s sign.
    - Brisk reflexes.
  - Absent abdominal reflex

- **Sensory**:
  - Dorsal spinal column affection → causes the following:
    - Loss of vibration.
    - Loss of fine touch sensations.
    - Numbness and paresthesia.
  - **Lhermitte’s syndrome**: an electric sensation that travels down the spine when the patient flexes his neck.

- **Cerebellar signs**: *(Charcot’s neurological triad)*
  - Scanning speech
  - Nystagmus
  - Intention tremors

- **Autonomic dysfunction**:
  - Bowel and bladder disorders.
  - Impaired sexual activity.
Diagnosis

- Multiple sclerosis should be suspected clinically with evidence of **UMNL signs**.
- MRI is the investigation which will show multiple sclerotic plaques at the **periventricular white matter**.

![MRI of the brain showing periventricular areas of hyperintensity](image)

*MRI of the brain shows periventricular areas of hyperintensity. This is relatively specific for multiple sclerosis.*

Management

There is **no cure** for MS. Treatment is focused on reducing the frequency and duration of relapses.

- **Acute relapse**:
  - **First line**: High dose steroids (e.g. IV Methylprednisolone) is given for 3-5 days to shorten the length of acute relapse. If the severity of symptoms decreased, the slow tapering of glucocorticoid therapy is then recommended.
  - **Second line**: plasmapheresis.

- **Reducing the replaces**:
  - **Beta-interferon**: The first-line drug in all forms of MS. It suppresses T cell activity.
  - **Glatiramer acetate**: An immunomodulating drug acts as a decoy for T cells.
  - **Natalizumab**: A recombinant monoclonal antibody that antagonizes α4-integrin on the surface of leucocytes, thus inhibiting invasion of leukocytes into the CNS.
  - **Fingolimod**: Sphingosine 1-phosphate receptor modulator which prevents lymphocytes from invasion into CNS.
Pre-Ganglionic Lesions

1) Pancoast's Tumor

- Lung cancer is one of the leading causes of death worldwide.
- 70% of cases are attributed to smoking.
- Pancoast's is considered a peripheral lung carcinoma (Usually Non-small cell lung cancer) that is located in the apex of the lung.
- It often affects: Cervical sympathetic nerves & Brachial plexus.

Clinical Pictures

- Horner's Syndrome: with anhydrosis of the face only.
- Severe pain in the axilla and shoulder.
- Edema of arm and facial swelling from the compression on adjacent structures.
- Paraneoplastic syndrome manifestations.

Diagnosis

- Chest x-ray shows apical opacity (see the image).
- CT chest for further assessment.
- Positron Emission Tomography (PET), it is more accurate than CT in differentiating between benign and cancerous lesions.
- Bronchoscopy and taking biopsy from the lesion.

Management

- Treatment is lung cancer is based on the stage of the cancer.
  - Curative: Chemotherapy and radiotherapy + Surgery in small respectable tumors.
  - Palliative: Chemotherapy and radiotherapy + palliative surgeries.
2) Thyroidectomy

- Thyroidectomy is a procedure commonly performed to treat different thyroid disorders.
- It varies from total thyroidectomy to only removal of the affected thyroid lobe (Thyroid lobectomy).

Complications

- Thyroidectomy is associated with various post-operative complications depending on the extent of resection. The greater the extent of resection, the greater the risk of complications.
- *Horner's syndrome* is considered one of the known complications, and should be considered if the patient complained of:
  - Ptosis.
  - Hydrosis.
  - Anhydrosis of the face only.

3) Trauma

- Direct penetrating or blunt trauma to the neck may affect the sympathetic chain resulting in Horner's syndrome.
- Diagnosis is based in the classical features of Horner's syndrome:
  - Ptosis.
  - Hydrosis.
  - Anhydrosis of the face only.

Subscribe using the following link in order to get the full access to our website:

SUBSCRIBE