

# Central Vestibular Disorders

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Disorders of the central nervous system can lead to dizziness and dysfunction of the vestibular system. To diagnose a central disorder one has to be familiar with the various neurological disorders that usually cause vestibular dysfunction. Although many conditions can lead to a central vestibular disorder they can typically be grouped into infective, vascular, neoplastic, craniovertebral junction disorders, demyelination (multiple sclerosis), cerebellar ataxias and focal seizure disorders.

## □ Infective causes

Brain abscesses especially those involving the cerebellum and the temporal lobe, petrous apex infections and encephalitis can lead to a central vestibular disorder.

## □ Vascular Disorders

### **Migraine**

Headaches and dizziness have been linked in many different settings. In classical basilar migraine with aura, vestibulocochlear symptoms are commonly seen. To diagnose this central vestibular disorder one needs to be familiar with the typical symptoms associated with migraine. These might include visual symptoms (blind spots, a zigzag pattern in their visual field, flashing or colored lights and double vision), dysarthria, paresthesia, paresis, vertigo, tinnitus, hearing loss and ataxia. The migraine aura should develop gradually and not last more than 60 minutes. In one study neurotological symptoms were shown to be significantly higher in those with migraine as compared to those with tension headaches<sup>1</sup>. In this study there were no cochlear symptoms in any of the patients with tension headache and vertigo as opposed to the migraine group.

### **Etiology**

The most accepted theory is that of intracerebral vasoconstriction which leads to an aura followed by extracranial vasodilatation which leads to the headache.

### **Management**

Migraine management consists of symptomatic treatment and prophylaxis.

Symptomatic treatment might include simple analgesics, antiemetics and sedatives.

If these fail to abort the attack then a 5HT<sub>1D</sub> receptor agonist such as sumatriptan (Imitrex) is usually recommended.

Prophylactic treatment is needed in most patients with frequent attacks. Propranolol, calcium channel blockers, amitriptyline or acetazolamide may be used. Acetazolamide is especially useful in patients with a familial essential tremor or associated seizures<sup>2</sup>.

## **Vertebrobasilar insufficiency (VBI)**

VBI is more common in the elderly with atherosclerotic disease of the vertebral and basilar systems<sup>3</sup>. Vertigo is associated with other symptoms resulting from a deficit in the posterior circulation. These symptoms might include drop attacks, ataxia, visual defects, weakness and paresthesia. Sometimes attacks are precipitated by hypotension, neck movement (cervical spondylosis) or upper arm exercises (subclavian steal syndrome).

### **Management**

Management is usually conservative and includes reduction of risk factors for atherosclerosis and treatment with ASA.

## **Brainstem infarction**

Vertigo in a brainstem infarct is associated with focal neurological deficits. Infarction involving the dorsolateral medulla (Wallenberg's syndrome) usually leads to:

- ❖ Vertigo
- ❖ Nystagmus
- ❖ Horner's syndrome
- ❖ Loss of pain and temperature involving the ipsilateral side of the face and the contralateral side of the body
- ❖ Ipsilateral paralysis of palate and larynx
- ❖ Ipsilateral facial and abducens nerve palsies
- ❖ Ipsilateral dysmetria, dysrhythmia and dysidiadochokinesia<sup>4</sup>.

Involvement of anterior inferior cerebellar artery (AICA) will result in infarction of the dorsolateral pontomedullary region and inferolateral cerebellum (lateral pontomedullary syndrome)<sup>5</sup>. As the labyrinthine artery originates from the AICA in most patients there is usually an infarction of the membranous labyrinth. Focal neurological signs include:

- ❖ Vertigo
- ❖ Ipsilateral hearing loss and tinnitus
- ❖ Ipsilateral facial weakness
- ❖ Ipsilateral cerebellar dysfunction
- ❖ Loss of pain and temperature sensation ipsilateral face and contralateral body
- ❖ Nystagmus<sup>4</sup>.

## **Cerebellar infarction**

An isolated cerebellar infarction can occur without involvement of the brainstem. This is usually embolic in nature.

Persistent vertigo lasting days to weeks associated with vomiting and ataxia is the typical presentation. Attention should be directed to the presence of cerebellar signs and gaze paretic nystagmus.

## **Cerebellar hemorrhage**

This is often seen in hypertensive patients. Initially the symptoms are similar to a cerebellar infarct i.e. vertigo, nystagmus and ataxia. If unrecognized the resultant pressure will lead to brainstem compression and hydrocephalus which is usually fatal. It

is therefore essential that suspected cases should have an urgent CT or MRI scan as surgical decompression is only successful in the early stages.

## □Neoplastic Disorders

Brainstem and cerebellar neoplasm can lead to dizziness and ataxia. Because of their location other neurological signs are likely to be present as well. Brainstem gliomas when slow growing are usually treated with radiotherapy.

Tumours of the fourth ventricle (medulloblastomas, ependymomas, papillomas, teratomas and epidermoid cysts) usually present with headaches as the result of obstructive hydrocephalus. Atypical positional nystagmus and vertigo however may be the presenting symptom<sup>6</sup>.

Cerebellar tumours can also cause hydrocephalus as they enlarge and will present usually with headaches, imbalance and papilledema. Positional vertigo and nystagmus as well can be a presenting complaint<sup>7</sup>.

## □Disorders of craniovertebral junction

Disorders of craniovertebral junction are often difficult to diagnose and manage. It is important however to recognize as sometimes successful surgical treatment is possible. Symptoms usually result from a cervicomedullary compression. Vestibulocochlear symptoms such as vertigo, tinnitus and hearing loss are often seen in association with lower cranial nerve palsies, limb weakness and numbness.

### **Basilar impression**

A bony abnormality of the skull base sometimes leads to the skull descending upon the spinal column. With the odontoid process of C2 impinging on the foramen magnum, the anterior aspect of medulla is often compressed. This in turn will cause lower cranial nerve problems and sometimes a vestibular dysfunction.

Diseases which are often associated with basilar impression include: Paget's disease, rickets, osteomalacia, osteogenesis imperfecta and rheumatoid arthritis<sup>8</sup>.

### **Assimilation of the atlas**

This is primarily seen in association with the Klippel-Feil syndrome. The atlas is usually fused partially or completely with the skull base limiting the movement between the two. As a result the odontoid may impinge on structures in the foramen magnum. Patients with this condition are prone to atlantoaxial dislocation.

### **Atlantoaxial dislocation**

Normally the transverse ligaments keep the odontoid process in its normal position. Laxity of this ligament will lead to dislocation of odontoid process on flexion and extension of the neck. Dislocation will result in compression of structures in the foramen magnum.

Various diseases have been known to predispose to this condition. Patients with Downs' syndrome, Hurler's syndrome, rheumatoid arthritis and retropharyngeal infections are especially prone to atlantoaxial dislocation<sup>9</sup>.

## **Chiari malformation**

In this congenital abnormality of the cervicomedullary junction the caudal end of the brainstem and cerebellum herniates into the cervical canal. There are two distinct types usually distinguished by the degree and contents of brain herniation that occurs thru the foramen magnum. A Chiari type II malformation (where there is significant herniation of both the medulla and cerebellar tonsils) is more common. It usually presents early in life and is associated with other congenital anomalies (myelomeningocele etc). The Chiari type I malformation is much more difficult to diagnose and often presents in early adulthood. It is best diagnosed with a MRI scan. Chiari type I is often associated with vestibular symptoms<sup>10</sup>.

## **□ Multiple Sclerosis (MS)**

A diagnosis of MS is made when there are symptoms and signs of multifocal demyelination in time and location. It usually presents in the third and fourth decades of life. New lesions are not usually seen after age 55 although an affected individual will still carry the previous burden of any permanent deficit they may have.

Symptoms may include visual loss as the result of retrobulbar (optic) neuritis, weakness, numbness, vertigo, ataxia and even hearing loss.

Examination findings are variable depending on the extent of the disease process.

Nystagmus is often pendular and may demonstrate atypical positional features. Cerebellar signs may also be present when the cerebellum is affected.

The investigation of choice is an MRI scan which will detect white matter change in most patients. Other useful diagnostic tools include triple evoked potentials and a CSF examination looking for oligoclonal banding

Treatment may include the use of steroids in an acute attack and disease modifying agents (interferon beta-1b etc as prophylaxis<sup>11</sup>).

## **□ Cerebellar ataxia syndromes**

### **Spinocerebellar atrophy**

This is a progressive degenerative disorder with an autosomal dominant inheritance. There are six different types diagnosed to date. The basic defect in all types is an expansion of a CAG triplet repeat. In this way, it is similar to fragile-X syndrome, Huntington disease and myotonic dystrophy, all of which exhibit a triplet repeat expansion of a gene. In the case of spinocerebellar atrophy the gene is found on chromosome 6.

Clinically the major problem is progressive cerebellar ataxia. There is often associated macular degeneration. There may also be pyramidal and extrapyramidal features.

## **Cerebellar atrophy**

In this disorder there is a progressive cerebellar ataxia without other symptoms. This mainly affects the midline cerebellum. There are typical signs of vestibulocerebellar degeneration that includes various abnormal eye movements.

## **Familial episodic ataxia**

This episodic form of ataxia is rare but important as some patients improve dramatically with acetazolamide administration. There are two types, EA1 and EA2. The locus for EA1 is on chromosome 12p while EA2 defect is on chromosome 19p.

## **Friedreich's ataxia**

This is a progressive degenerative disorder of spinocerebellar pathways, posterior columns and pyramidal tracts, resulting in a worsening ataxia. There is usually loss of vibration and position sensation, loss of reflexes and often scoliosis. The oculomotor defects include saccadic smooth pursuit, saccadic dysmetria, ocular flutter and failure of VOR cancellation. Heart failure often results in these patients from a hypertrophic cardiomyopathy.

The chromosomal defect (X25) was found to be on chromosome 9q13.

## **Refsum's disease**

This is an autosomal recessive disorder due to a phytanic acid oxidase deficiency. Phytanic acid accumulates in serum and a variety of tissues. Symptoms often include cerebellar ataxia, visual loss, weakness due to polyneuropathy, sensorineural hearing loss and ichthyosis. There is often cardiomyopathy and ECG changes.

Treatment is by dietary restriction of foods which contain phytanic acid.

## **Ataxia telangiectasia**

Ataxia telangiectasia is another form of cerebellar ataxia associated with telangiectasia of the skin and conjunctiva. The defect is on chromosome 11q. There is often associated immune deficiency and raised serum  $\alpha$ -fetoprotein.

## **Paraneoplastic cerebellar degeneration**

This is a rapidly progressive cerebellar degeneration associated with various malignant diseases. Small cell lung carcinoma, breast carcinoma, ovarian carcinoma and Hodgkin's lymphoma are most commonly associated with an autoimmune antibody production which cross reacts with cerebellar Purkinje's cell.

## **Alcoholic cerebellar degeneration**

This is a selective degeneration of the anterior vermis of cerebellum as the result of alcohol abuse. This usually results in mainly truncal ataxia.

## **▣ Focal seizures**

Sometimes patients with temporal or parietal seizures will experience as part of their aura vertigo, tinnitus and auditory hallucinations. This is possibly the result of absent electrical activity in the auditory and vestibular cortical projections

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