

CENTRAL NERVOUS SYSTEM CAUSES OF VERTIGO

CENTRAL NERVOUS SYSTEM CAUSES OF VERTIGO

Central vertigo is a false sensation of motion caused by a lesion in the CNS that results in dysfunction of **the vestibular nuclei or their projections to the cerebellum**. The vestibular nuclei are located in the caudal pontine tegmentum and dorsolateral medulla and can be subdivided into four separate subnuclei: the superior, lateral (Deiters), medial, and inferior vestibular. These nuclei receive afferents from the peripheral vestibular system by way of the vestibular division of cranial nerve VIII. They also receive afferents from the cerebellum, the reticular formation in the pons, the spinal cord, and the vestibular nuclei on the opposite side. Projections from the vestibular nuclei reach the cerebellum, extraocular nuclei, and spinal cord. *Lesions in any of these areas can result in the symptom of vertigo.*

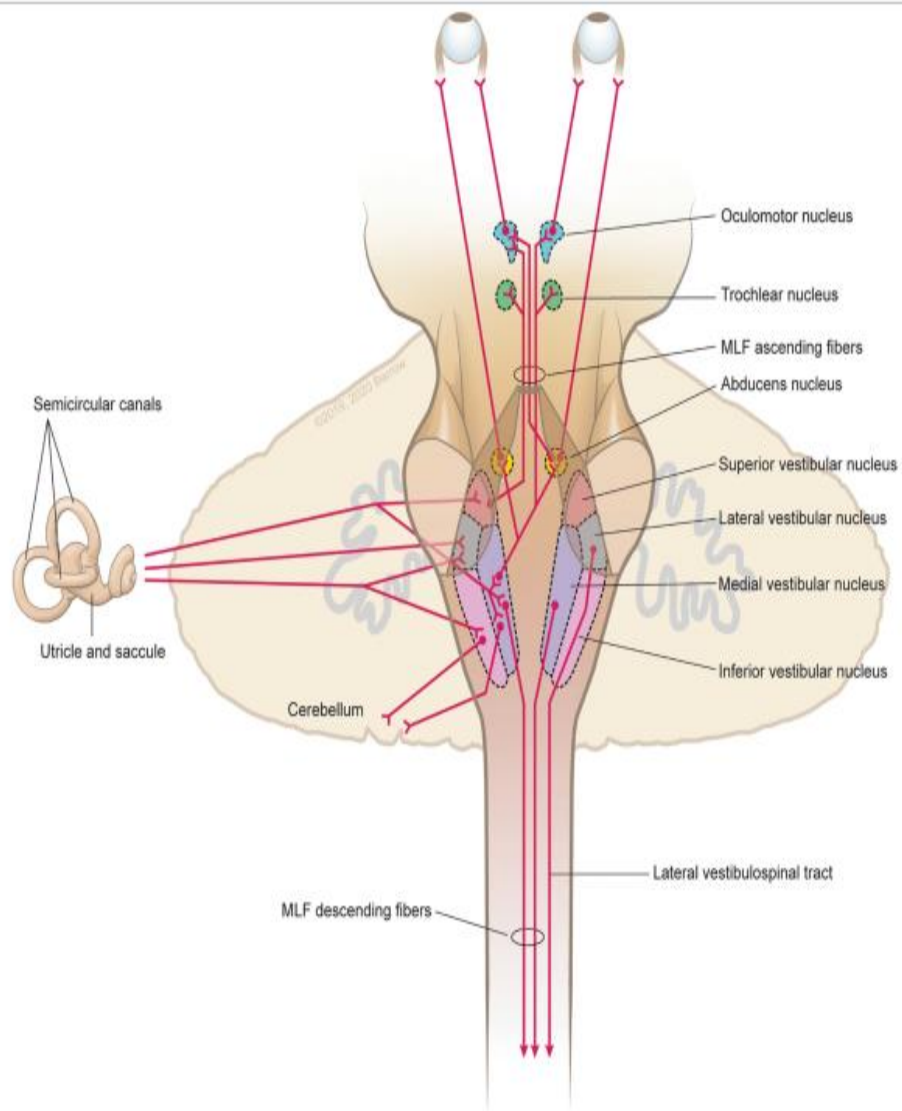


FIGURE 7-1

Structures of the central nervous system vestibular system.

MLF = medial longitudinal fasciculus.

Vestibular Migraine

Vestibular migraine is a common cause of vertigo seen in patients with a history of migraine. Vestibular migraine causes ***episodic vertigo that can appear positional, spontaneous, or visually induced.***

Vestibular symptoms may occur during headaches but also commonly occur without headache.

Between episodes, some patients may experience chronic dizziness and imbalance.

Vestibular migraine remains a clinical diagnosis, and diagnostic criteria have been developed by the Bárány Society and the International Headache Society. Although a clear understanding of the pathophysiology is lacking, it may be related to the presumed pathology of migraine. **Pathologic nystagmus and central vestibular dysfunction have been seen in the majority of patients with vestibular migraine studied, although they are often nonspecific**

Diagnostic criteria:

A. At least five episodes fulfilling criteria C and D

B. A current or past history of 1.1 *Migraine without aura* or 1.2 *Migraine with aura*¹

C. Vestibular symptoms of moderate or severe intensity, lasting between 5 minutes and 72 hours

D. At least half of episodes are associated with at least one of the following three migrainous features:

1. headache with at least two of the following four characteristics:

- a) unilateral location
- b) pulsating quality
- c) moderate or severe intensity
- d) aggravation by routine physical activity

2. photophobia and phonophobia

3. visual aura

Not better accounted for by another ICHD-3 diagnosis or by another vestibular disorder

Vestibular symptoms are rated **moderate** when they interfere with but do not prevent daily activities and **severe** when daily activities cannot be continued.

Duration of episodes is highly variable. About 30% of patients have episodes lasting minutes, 30% have attacks for hours and another 30% have attacks over several days. The remaining 10% have attacks lasting seconds only, which tend to occur repeatedly during head motion, visual stimulation or after changes of head position. In these patients, episode duration is defined as the total period during which short attacks recur. At the other end of the spectrum, there are patients who may take 4 weeks to recover fully from an episode. However, **the core episode rarely exceeds 72 hours.**

One symptom is sufficient during a single episode. Different symptoms may occur during different episodes. Associated symptoms may occur before, during or after the vestibular symptoms.

History and physical examinations do not suggest another vestibular disorder *or* such a disorder has been considered but ruled out by appropriate investigations *or* such a disorder is present as a comorbid condition but episodes can be clearly differentiated. Migraine attacks may be induced by vestibular stimulation. Therefore, ***the differential diagnosis should include other vestibular disorders complicated by superimposed migraine attacks***

Vestibular symptoms, as defined by the Bárány Society's Classification of Vestibular Symptoms and qualifying for a diagnosis of A1.6.6 *Vestibular migraine*, include

:a) spontaneous vertigo:

- internal vertigo (a false sensation of self-motion);
- external vertigo (a false sensation that the visual surround is spinning or flowing);

b) positional vertigo, occurring after a change of head position;

c) visually-induced vertigo, triggered by a complex or large moving visual stimulus;

d) head motion-induced vertigo, occurring during head motion;

e) head motion-induced dizziness with nausea (dizziness is characterized by a sensation of disturbed spatial orientation; other forms of dizziness are currently not included in the classification of vestibular migraine)

Other symptoms

Transient auditory symptoms, nausea, vomiting, prostration and susceptibility to motion sickness may be associated with A1.6.6 *Vestibular migraine*.

However, since they also occur with various other vestibular disorders, they are not included as diagnostic criteria.

Relation to migraine aura and migraine with brainstem aura

Both *migraine aura* and *migraine with brainstem aura* (formerly: *basilar-type migraine*) are terms defined by ICHD-3. Only a minority of patients with A1.6.6 *Vestibular migraine* experience their vertigo in the time frame of 5-60 minutes as defined for an aura symptom. Even fewer have their vertigo immediately before headache starts, as required for 1.2.1.1 *Typical aura with headache*. Therefore, episodes of A1.6.6 *Vestibular migraine* cannot be regarded as migraine auras.

Although vertigo is reported by more than 60% of patients with 1.2.2 *Migraine with brainstem aura*, ICHD-3 requires at least two brainstem symptoms in addition to visual, sensory or dysphasic aura symptoms for this diagnosis. Fewer than 10% of patients with A1.6.6 *Vestibular migraine* fulfil these criteria. ***Therefore, A1.6.6 Vestibular migraine and 1.2.2 Migraine with brainstem aura are not synonymous, although individual patients may meet the diagnostic criteria for both disorders***

Relation to benign paroxysmal vertigo

While A1.6.6 *Vestibular migraine* may start at any age, ICHD-3 specifically recognizes a childhood disorder, 1.6.2 *Benign paroxysmal vertigo*. The diagnosis requires five episodes of vertigo, occurring without warning and resolving spontaneously after minutes to hours. Between episodes, neurological examination, audiometry, vestibular functions and EEG must be normal. A unilateral throbbing headache may occur during attacks but is not a mandatory criterion. 1.6.2 *Benign paroxysmal vertigo* is regarded as one of the precursor syndromes of migraine. Therefore, previous migraine headaches are not required for diagnosis. Since the classification of A1.6.6 *Vestibular migraine* does not involve any age limit, the diagnosis can be applied in children when the respective criteria are met, **but only children with different types of vertigo attacks (eg, short-duration episodes of less than 5 minutes and longer-lasting ones of more than 5 minutes) should receive both these diagnoses.**

Overlap with Menière's disease

Migraine is more common in patients with Menière's disease than in healthy controls. Many patients with features of both Menière's disease and A1.6.6 Vestibular migraine have been reported. In fact, migraine and Menière's disease can be inherited as a symptom cluster. **Fluctuating hearing loss, tinnitus and aural pressure may occur in A1.6.6 Vestibular migraine, but hearing loss does not progress to profound levels.** ***Similarly, migraine headaches, photophobia and even migraine auras are common during Menière attacks.*** The pathophysiological relationship between A1.6.6 Vestibular migraine and Menière's disease remains uncertain. In the first year after onset of symptoms, differentiation between them may be challenging, since Menière's disease can be monosymptomatic with only vestibular symptoms in the early stages of the disease.

When the criteria for Menière's disease are met, particularly hearing loss as documented by audiometry, Menière's disease should be diagnosed, even when migraine symptoms occur during the vestibular attacks. Only patients who have two different types of attacks, one fulfilling the criteria for A1.6.6 Vestibular migraine and the other for Menière's disease, should be diagnosed with both disorders. A future revision of ICHD may include a vestibular migraine/Menière's disease overlap syndrome.

Multiple Sclerosis

- MS causes inflammatory demyelinating lesions throughout the CNS and is known to cause lesions specifically in areas that result in vertigo (ie, the brainstem and cerebellum). It has been estimated that 20% of patients with MS will experience **true vertigo** during their lifetime, and in **about 5%** of patients with MS, it is **the presenting symptom** of the disease.

- The most common CNS sites known to cause vertigo in MS are lesions ***at the root entry zone of cranial nerve VIII (the lateral pontomedullary junction) and the medial vestibular nucleus***. Additionally, patients can have symptoms from lesions scattered throughout the cerebellum. In one retrospective analysis of a university-based population of patients with MS presenting with acute vertigo due to demyelinating plaques, three-fourths of the patients had a lesion in the root entry zone of cranial nerve VIII and one-fourth had a lesion in the medial vestibular nucleus. ***It should be noted that a root entry zone lesion may cause vertigo that behaves as if caused by a peripheral vestibular lesion although the lesion may be in the CNS.***

- Vertigo due to MS may present ***acutely*** during an exacerbation, or it can persist in ***a chronic form*** as a result of disease burden.
- During an exacerbation, vertigo typically begins acutely and can be accompanied by nausea and vomiting. Patients may be ataxic and may also report diplopia. Symptoms can be explained by active (enhancing) lesions in the areas described above. The vertigo usually improves or remits as the exacerbation resolves.
- Treatment is usually indicated and consists of corticosteroids for most patients with MS who are able to tolerate them. Vertigo can be managed symptomatically with antiemetics or vestibular suppressants such as benzodiazepines (which are not recommended for long-term use). ***Vestibular therapy is not usually of strong benefit for central vertigo in MS; however, it has been shown to improve balance and disability due to dizziness or general disequilibrium in patients with MS***

Central vertigo from MS is usually seen along with various focal findings on neurologic examination. Abnormal saccades with reduced velocities, nystagmus (potentially in multiple directions), impaired suppression of the vestibulo-ocular reflex, and internuclear ophthalmoplegia (INO) are the prominent features that can be observed. INO is the most common eye movement disorder seen in MS and is caused by demyelination of the medial longitudinal fasciculus in the pons or midbrain. It is a disorder of impaired conjugate lateral gaze, resulting in slowing adduction or even paralysis of the adducting eye if severe enough. The abducting eye exhibits nystagmus, and patients report diplopia. An INO can be unilateral or bilateral in patients with MS, and variants exist. It is worth noting that the presence of an INO does not necessarily mean a patient will have vertigo. Similarly, not all patients with MS with vertigo have an INO on their examination.

Patients with MS may have saccadic dysmetria from cerebellar involvement, particularly when the cerebellar peduncle is affected. Cerebellar lesions can also cause impaired smooth pursuit, and gaze-evoked, downbeat, or acquired pendular nystagmus that may be associated with oculopalatal tremor and often with dizziness, imbalance, and oscillopsia (a perception of objects bouncing or oscillating). Acquired pendular nystagmus likely results from damage to the neural integrator network in the brainstem and cerebellum.

Another important type of vertigo that patients with MS may experience is **central positional vertigo**. This can be more challenging to diagnose and *may be confused with benign paroxysmal positional vertigo (BPPV)*. A central positional vertigo is much rarer than BPPV. BPPV is more prevalent in patients with MS than in the general population. A retrospective analysis of 1153 patients with MS with acute vertigo found that more than 50% of the patients had BPPV, and all were treated successfully with canalith repositioning maneuvers.

- Central positional vertigo is similar to BPPV in that it is triggered by position change. However, several pearls can help differentiate it from the more common BPPV:
 - *Patients with BPPV typically have a brief period of latency during a provocative maneuver such as the Dix-Hallpike test. Central positional vertigo often has no latency, so nystagmus commences immediately upon positioning.*
 - *The nystagmus in BPPV fatigues after some time in the head-hanging position, whereas central positional vertigo may exhibit nystagmus that persists and is prominent even after repeat positioning.*
 - *The pattern of nystagmus is perhaps the most important difference. Classic posterior canal BPPV presents with both upbeat and torsional components, whereas central positional vertigo is more likely to present without both components simultaneously*



FIGURE 7-2

Axial postcontrast T1-weighted MRI shows an acute demyelinating lesion (*circle*) in the right middle cerebellar peduncle adjacent to the cerebellar nodulus.

Reprinted with permission from Barrow Neurological Institute.
© 2020 Barrow Neurological Institute, Phoenix, Arizona.

Therefore, since the clinical presentation of both types of vertigo can look identical, it is important that neurologists pay ***close attention to the directional features of nystagmus*** to better differentiate them and treat accordingly. Several case studies have shown the most common lesion responsible for central positional vertigo in patients with MS to be in the superior cerebellar peduncle (brachium conjunctivum); however, other lesions in the cerebellum are also known to cause central positional vertigo. Small lesions in this region may be missed if thin MRI slices are not obtained when imaging the posterior fossa

Stroke and Transient Ischemic Attack

- Strokes and transient ischemic attacks (TIAs) are known causes of central vertigo, dizziness, and imbalance when ***the posterior circulation*** is affected. A cerebellar ischemic stroke in the posterior inferior cerebellar artery (PICA) territory, the anterior inferior cerebellar artery (AICA) territory, or the superior cerebellar artery territory may be associated with vertigo or nystagmus, or both.
- ***Vertigo and nystagmus more commonly occur when the area of infarction affects the cerebellar peduncles, flocculus, nodulus, vermis, and paravermian regions and are less common with small lesions that are far lateral in the cerebellar hemisphere.*** Vertigo may also manifest from a brainstem infarction, which can affect the lateral medulla, medial medulla, pons, or midbrain

- Vertigo with unilateral hearing loss can be caused by ***a labyrinthine infarction***. The labyrinth is supplied by ***the internal auditory artery, usually a branch of the AICA***. ***This can be missed on brain imaging***, so clinical suspicion is imperative.

- In fact, AICA territory infarct can present with both peripheral and central findings***. Involvement of the root entry zone of the facial nerve or the labyrinth could lead to peripheral findings such as facial paresis, hearing loss, and vertigo. At the same time, expansion of stroke can lead to involvement of the pons or cerebellum, or both, which can cause central vertigo and ataxia. Recent literature points out that audiovestibular loss in isolation can be an impending sign of AICA territory infarction, with initial symptoms of only vertigo and hearing loss occurring days or weeks before the presentation of a posterior fossa stroke. Most patients with this presentation seem to have evidence of reduced basilar artery flow near the AICA origin. ***This type of infarct should be considered in patients with vascular risk factors who experience audiovestibular loss even if MRI is unrevealing***

• **Chronic vertigo due to the late effects of stroke** is not an uncommon symptom; it is often a residual symptom that can persist long after the infarct occurs. It may be caused by central or peripheral damage or a combination of the two, as discussed above. **Initial management usually is a short course of a vestibular suppressant followed by physical therapy.** Central lesions may not respond as quickly or as successfully to therapy as peripheral insults; however, therapy can be helpful in improving balance overall after a stroke.

• **Visually induced vertigo is common in these patients,** and they report symptoms that are exacerbated or triggered by complex visual surroundings. Therapy programs with visual-vestibular stimulation during therapy can result in greater improvements in such patients.

Tumors and Other Structural Lesions in the Central Nervous System

Neoplasms and vascular lesions can cause central vertigo and other neurologic symptoms based on their location in the CNS, including vestibular schwannoma, cavernous malformation, hemangioblastoma, and medulloblastoma.

Vestibular schwannoma,

- also known as acoustic neuroma, is a tumor that arises from the Schwann cells around cranial nerve VIII. Although the tumor forms from the vestibular portion of the nerve, it most commonly presents with hearing loss as it can compress the cochlear division of the nerve.
- **Vertigo is an infrequent presentation of vestibular schwannoma, estimated to be seen initially in less than 15% of patients.** Slowly progressive unilateral hearing loss and tinnitus are more common initial symptoms, although in a small percentage of patients hearing loss may occur suddenly. **When patients present with dizziness, they may describe spinning vertigo, lightheadedness, or gait imbalance.**
- As vestibular schwannomas enlarge, they may lead to peripheral vestibular loss that contributes to the feeling of loss of balance. However, these tumors can also grow to compress the brainstem and thereby cause central vertigo.

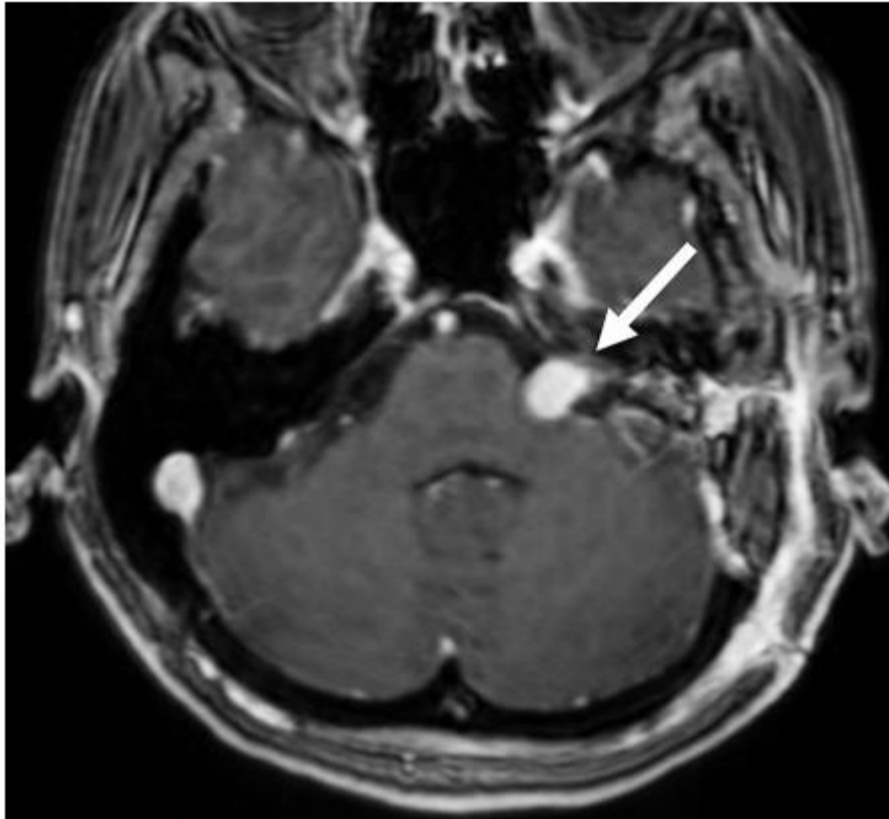


FIGURE 7-3

Vestibular schwannoma. Axial postcontrast T1-weighted MRI shows an enhancing left cerebellopontine angle extraaxial mass (*arrow*), consistent with vestibular schwannoma.

- An audiogram showing significantly asymmetric hearing loss may lead to suspicion of vestibular schwannoma. Asymmetry of 15 dB at 3 kHz on audiometry is associated with increased positive yield of finding an abnormality on MRI that explains the patient's hearing loss.

- If a vestibular schwannoma is suspected, *an MRI of the brain with and without gadolinium* can be used to evaluate for an enhancing lesion in the internal auditory canal or cerebellopontine angle . A cerebellopontine angle meningioma can present similarly in many respects.

Observation and sequential imaging, radiosurgery, and microsurgery are all management options for vestibular schwannoma. Decisions for treatment are often based on the patient's age and surgical risk factors, quality-of life considerations, the size of the tumor and its rate of growth, the status of vestibular and hearing function, and the desire or need to preserve hearing function

Cavernous malformations

are either sporadic or inherited vascular malformations. When sporadic, they are usually single lesions that may be associated with a developmental venous anomaly. Cavernous malformations are made up of dilated capillaries with thin walls and are often surrounded by hemosiderin from recurrent hemorrhage. They are described on MRI as having ***a popcorn ball appearance***

- About **one-fourth** of cavernous malformations are found in the posterior fossa, usually in the **pons or the cerebellum**; these tend to have higher annual bleeding rates than supratentorial cavernous malformations. **Vertigo can be a presenting symptom of a cavernous malformation, especially if active hemorrhage is present.** Nausea, vomiting, and diplopia can also accompany the acute vertigo in these cases

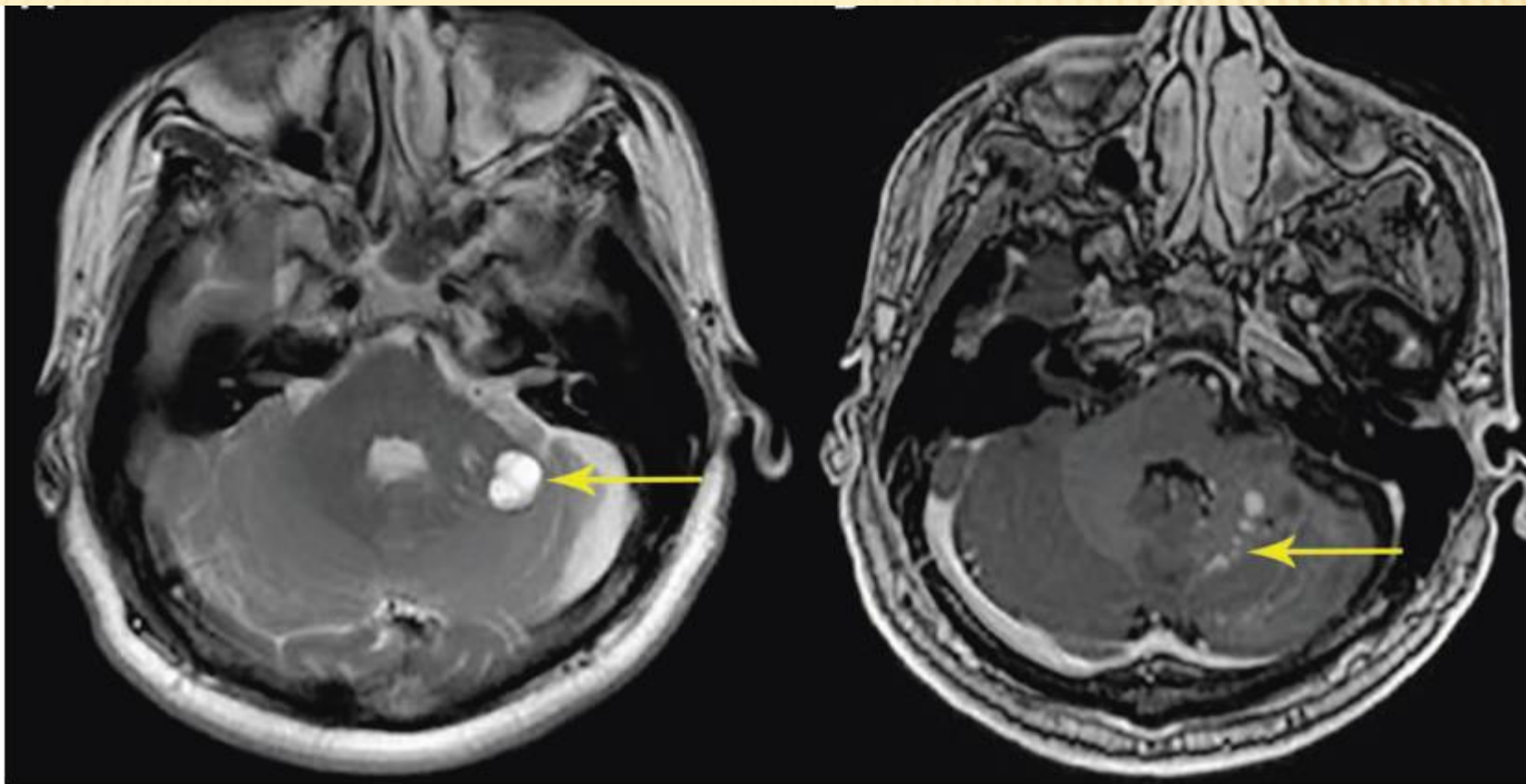


FIGURE 7-4

Cavernous malformation. Axial T2-weighted (A) and postcontrast T1-weighted (B) images of a left cerebellar cavernous malformation. A T2-hyperintense cystic component and T2-hypointense rim of hemosiderin surround the lesion (A, arrow). An adjacent developmental venous anomaly, a common association, is seen on the postcontrast image (B, arrow).

Hemangioblastomas

are tumors that grow slowly in the spinal cord, cerebellum, or brainstem. They can be sporadic but are often seen in patients with von Hippel-Lindau disease, along with various other tumors. Because of the strong association with von Hippel-Lindau disease, finding a CNS hemangioblastoma often prompts genetic evaluation for the disease in patients who have not yet been diagnosed.

- **Hemangioblastoma can cause vertigo and ataxia because of compression of structures in the brainstem or cerebellum or because of hemorrhage in those areas.**
- **In the cerebellum, where they usually present, hemangioblastomas often appear as enhancing nodules within a cyst.**
- **Rarely, hemangioblastomas can occur in the cerebellopontine angle and may be misdiagnosed as vestibular schwannoma since the symptoms and MRI findings can be similar.**

Medulloblastoma

is the most common malignant brain tumor diagnosed in children. It presents with headache, nausea, and vomiting, often because of fourth ventricle involvement causing increased intracranial pressure. In addition, patients may have dizziness or vertigo due to brainstem compression and cerebellar involvement. Midline cerebellar lesions may cause more gait or truncal ataxia than lateral cerebellar tumors, which can cause more limb dysmetria.

- Medulloblastoma is usually seen on MRI in the cerebellum, with some areas of enhancement and possible obstruction of the fourth ventricle.
- Central patterns of nystagmus can be seen in primary gaze or with end gaze on examination; however, medulloblastoma is also known to cause central positional vertigo only. Therefore, BPPV is sometimes suspected but should be considered unlikely when patients are young and the symptoms do not improve with repositioning maneuvers or when the nystagmus is sustained.²

Meningitis and Encephalitis

- **Acute bacterial meningitis** may cause bilateral hearing and vestibular loss, especially in children. The most common organisms known to cause this include *Streptococcus pneumoniae*, *Neisseria meningitidis*, and *Haemophilus influenzae* type b. Cases attributed to *H. influenzae* type b have decreased since a vaccine was introduced. The causative lesion may be in the inner ear end organs, the vestibulocochlear nerve, brainstem, or auditory and vestibular pathways. Infection may spread to the inner ear from the subarachnoid space via the cochlear aqueduct or the cochlear modiolus.
- **Dizziness and bilateral vestibulopathy may sometimes occur with chronic meningitis.** Inflammation within the central audiovestibular pathways and cranial nerve nuclei may be responsible for these symptoms. The list of specific causes of chronic infectious meningitis is long and includes tuberculosis, fungal infections such as coccidioidomycosis and cryptococcosis, and Lyme disease.

- Occasionally, certain types of ***localized brainstem encephalitis*** may cause dizziness. The term ***rhombencephalitis*** refers to inflammation affecting the brainstem or cerebellum, or both; it may be associated with dizziness, unsteadiness, nausea, diplopia, headache, and altered awareness. *Listeria monocytogenes* is the most common infectious cause of rhombencephalitis.

Sarcoidosis, a noninfectious disorder of unknown etiology, is a granulomatous process that can affect multiple body systems. Less commonly, it can affect the nervous system exclusively, presenting as neurosarcoidosis. Sarcoidosis has a predilection for the basal meninges and can affect the vestibulocochlear nerve exit or, rarely, can manifest with granulomas in the cerebellopontine angle. Combined evidence from retrospective review has shown that ***audiovestibular manifestations of sarcoidosis are primarily caused by cranial nerve VIII neuropathy***

Carcinomatous or lymphomatous meningitis may also cause multiple evolving cranial neuropathies and brainstem symptoms. This involves seeding of malignant cells to the leptomeninges. It can be seen in solid cancers, such as breast or lung, and with hematologic malignancies. Primary brain tumors can also spread to the meninges. ***Headache, cranial neuropathies, nausea, and dizziness are common at presentation.*** Imaging reveals diffuse leptomeningeal enhancement, often in the cerebellar folia and ventral surface of the brainstem, when patients have dizziness or unsteadiness

Chiari Malformation

Chiari malformations can be classified as types I through IV based on the anatomic structures involved in the malformation. Chiari malformation type I, the most common type, is a congenital lesion that may not manifest with symptoms until adulthood. In Chiari malformation type I, the cerebellar tonsils extend below the foramen magnum. Diagnosis is radiographic, and most sources agree that it is defined as tonsillar herniation of greater than or equal to 5 mm below the foramen magnum . Of note, the degree of herniation does not necessarily correlate with the extent of symptoms experienced by patients.

Chiari malformation type I can be asymptomatic but when symptomatic often presents with some combination of posterior headache, neck pain, weakness, dysphagia, or vertigo and gait imbalance. ***The vertigo in patients with Chiari malformations is usually induced by position change such as neck extension.*** This may be caused by pressure being applied to the brainstem or cerebellum or their blood supply. ***Vertigo is usually episodic and brief, often relieved by changing position.*** Nausea and vomiting may also accompany the vertigo. Although several patterns of nystagmus have been reported, most commonly a downbeat nystagmus is observed when examining a patient in primary gaze, especially when the patient is supine with the head slightly tilted back.

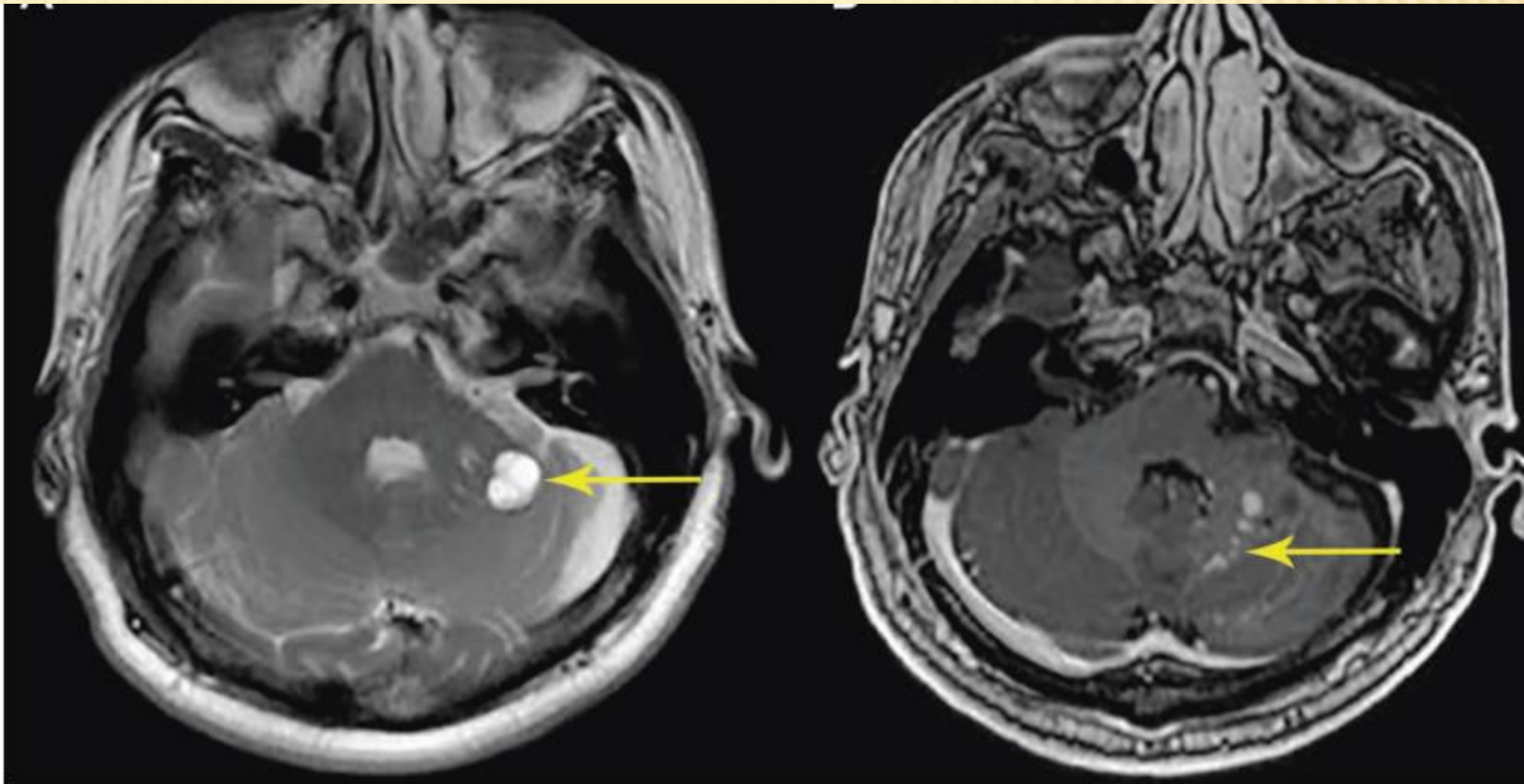


FIGURE 7-4

Cavernous malformation. Axial T2-weighted (A) and postcontrast T1-weighted (B) images of a left cerebellar cavernous malformation. A T2-hyperintense cystic component and T2-hypointense rim of hemosiderin surround the lesion (A, arrow). An adjacent developmental venous anomaly, a common association, is seen on the postcontrast image (B, arrow).

Superficial Siderosis

Superficial siderosis is an uncommon disorder that can affect various areas in the CNS, including the brainstem, spinal cord, cerebellum, supratentorial brain, nerve roots, and cranial nerves. It frequently leads to progressive symptoms of vertigo, ataxia, and hearing loss. The two types of superficial siderosis are cortical superficial siderosis and infratentorial superficial siderosis. Cortical superficial siderosis affects the supratentorial brain and can be seen in cerebral amyloid angiopathy. ***Infratentorial superficial siderosis more commonly affects the vestibular end organs, cranial nerve VIII, the brainstem, the cerebellum, and the spinal cord***

- **The hearing loss** seen in superficial siderosis usually **affects high frequencies** early on. It can be asymmetric at first but will progress and cause profound damage bilaterally with time. It is typically more severe than what would be expected for hearing loss due to presbycusis (hearing loss associated with aging). Hearing aids can be used in earlier stages, and cochlear implantation has been shown to have some benefit based on systematic review of available studies.

- ***When vestibular damage is present, patients may report dizziness or vertigo and exhibit gait instability.*** Since some gait instability in superficial siderosis is usually because of cerebellar damage, the vestibular system is often forgotten as a potential site of damage. However, cranial nerve VIII has a long course from the end organs through the internal auditory canal, making it vulnerable to damage. Damage to cranial nerve VIII can be assessed using various vestibular tests, such as videonystagmography, rotary chair testing, vestibular evoked myogenic potentials, and video head impulse testing. ***Overall, most patients with superficial siderosis appear to have both peripheral and central vestibulopathy***

- ***the diagnosis is made by MRI.*** Hemosiderin is seen easily on MRI sequences, including gradient recalled echo (GRE), T2-weighted, and susceptibility-weighted imaging (SWI). Superficial siderosis appears as rims of hypointensity

Neurodegenerative Disease

Vertigo and dizziness are commonly seen in *patients with Parkinson disease, multiple system atrophy, progressive supranuclear palsy, and cerebellar ataxia.*

Cerebellar ataxia has numerous potential etiologies, such as genetic disease, vitamin deficiencies, paraneoplastic disease, environmental/toxin exposures, and as a result of adverse effects of medications. Although the possible causes are myriad, the manifestations can be similar. Vertigo is often paroxysmal, and bedside examination usually reveals central nystagmus. Typical patterns include spontaneous downbeat nystagmus and direction-changing horizontal end-gaze nystagmus. Downbeat nystagmus results from degeneration of the cerebellum, leading to floccular hypofunction.

In Parkinson disease and the atypical conditions that cause parkinsonism, such as multiple system atrophy, **central orthostatic hypotension** may be a cause of presyncopal dizziness. This is because of involvement of the central autonomic network that helps to regulate visceromotor, neuroendocrine, and pain responses

Episodic Ataxias

Seven autosomal dominant episodic ataxias have been identified, aptly named episodic ataxia type 1 through episodic ataxia type 7. Of these types, most cases encountered are usually episodic ataxia type 1 or episodic ataxia type 2. In patients with episodic ataxia type 2, vertigo is severe and episodic, often accompanied by nausea and vomiting as well as unsteadiness. **Patients with episodic ataxia type 2 usually start having episodes during adolescence, and each episode can last hours. Stress is a common trigger, as are heat, exertion, alcohol, and caffeine.** Genetic testing usually reveals mutations in the CACNA1A gene, specifically in the P/Q-type calcium channel $\alpha 1A$ subunit.

Autoimmune Vestibulocerebellar Disorders

The literature on autoimmune vestibulocerebellar disorders is actively expanding as more is learned about the autoantibodies that are biomarkers for these disorders. Typically, the antibodies target antigens in the vestibulocerebellar pathways, vestibular nuclei, or vestibular end organs. This results in autoimmune syndromes characterized by symptoms such as dizziness and ataxia that can progress rather quickly. Clinicians must be proficient not only in diagnosis but also in initiating treatment in hopes of halting this progression.

- The presentation of patients with these disorders is **subacute**, meaning that patients usually have an onset of cerebellar symptoms over weeks to months. Symptoms may begin first with a prodrome of nausea and vomiting. Truncal and appendicular ataxia begin after this, along with vertigo, dysarthria, diplopia, and dysphagia. Extraocular movements are abnormal and may include any the following: positional vertical nystagmus (upbeat or downbeat), spontaneous downbeat nystagmus, spontaneous or gaze-evoked horizontal nystagmus, opsoclonus, periodic alternating nystagmus, or internuclear ophthalmoplegia.

- The diagnostic workup centers on testing for specific autoantibodies after a clinical syndrome is suspected. In general, antibody testing should be done on both serum and CSF samples. Diagnostic certainty results from the combination of a characteristic clinical syndrome and positive result of the accompanying antibody in either serum or CSF, or both.