

## CASE 2: HEARING LOSS AND DIZZINESS (Slide CC4-1)

1. **dizziness** - When clinicians talk of dizziness we are usually referring to *vertigo* which is a sensation of apparent movement or spinning that is in contradiction with actual movement. Vertigo can be caused by a lesion anywhere in the vestibular pathway, including the semicircular canals, the vestibular ganglion, the vestibular nerves, the vestibular nuclei and the cerebellum. Keep in mind that when patients complain of "dizziness," they may not feel actual vertigo, but rather, anything from generalized weakness, to trouble walking, to nausea, to light headedness, to vision problems. It is very important to always ask clarifying questions such as "did you feel the room spinning" (as our patient did) to determine exactly what the patient means by "dizziness."

**decreased hearing** - Hearing loss is usually divided into two categories:

1. *Conductive hearing loss* can be caused by obstruction of the external auditory canal, damage to the tympanic membrane or to the middle ear ossicles.

2. *Sensorineural hearing loss* is caused by damage to the cochlea, the hair cells or to the acoustic nerves. It is important to realize that unilateral hearing loss cannot be caused by lesions in the central nervous system since, as you will learn in a later lab, all auditory pathways contain bilateral information once they enter the brainstem.

Conductive and sensorineural hearing loss can be distinguished with a simple tuning fork (Rinne Test). In a normal individual the tone will sound louder when the vibrating tuning fork is held just outside the ear than when the handle is held against the mastoid process (air conduction > bone conduction). In conductive hearing loss the mechanical system for transmitting sound waves to the oval window of the cochlea is damaged and, therefore, bone conduction > air conduction. In sensorineural hearing loss the mechanical system is intact but the cochlea or neural pathways are damaged and, therefore, air conduction > bone conduction as in the normal individual (but hearing, of course, will be diminished in the affected ear).

**facial pain and decreased corneal response** - Pain in a sensory nerve distribution is usually caused by peripheral sensory nerve injury rather than by a central lesion. Facial pain is usually due to injury to the trigeminal nerve (CN V). Much less commonly, facial pain could occur due to focal seizure activity in the facial area of the sensory cortex..

Corneal sensation is supplied by the ophthalmic division of the trigeminal nerve (CN V<sub>1</sub>).

**decreased taste** - taste to the majority of the tongue (anterior 2/3) is supplied by the chorda tympani/nervus intermedius portion of the facial nerve (CN VII), with some, more posterior areas being supplied by CN IX and X. Taste sensation is then relayed via the nucleus solitarius to the thalamus and cortex as will be discussed in a later lab.

2. *Review the point of exit of each of the cranial nerves from the brainstem. Also, review the point of exit of each of the cranial nerves through the skull.*

Our patient has symptoms related to CN V, VII and VIII. The presence of unilateral sensorineural hearing loss implies peripheral CN VIII injury. Meanwhile, the involvement of CN V, as well, suggests that the lesion must lie within the cranial vault. Thus, a lesion must be present in the area where CN V, VII and VIII exit from the brainstem. This region is referred to as the *cerebellopontine angle*.

A number of different kinds of lesions can occur in the cerebellopontine angle, including brain metastases (recall that our patient recently had a melanoma, which frequently metastasizes to brain), meningiomas, gliomas, astrocytomas and epidermoid tumors. However, the most common lesion of the cerebellopontine angle by far is the ACOUSTIC NEUROMA: This slow growing tumor makes up about 10% of all intracranial tumors. It is a nerve sheath tumor which develops at the transition zone between Schwann cells and oligodendrocytes covering CN VIII. This transition occurs at the point where CN VIII enters the internal acoustic meatus. The tumor almost always arises from the vestibular division of CN VIII. It grows initially within the bony internal acoustic meatus, but then expands into the cerebellopontine angle. As it grows the symptoms produced are often more related to the pressure sensitivity of the structures involved rather than the proximity to the tumor. Thus, although the tumor arises from the sheath of the

vestibular nerves, vertigo is usually not a prominent symptom (unlike our patient). Initially, most patients experience tinnitus (ringing in the ear) and unilateral hearing loss due to compression of the acoustic nerve. As the tumor expands it compresses CN V. The fibers supplying corneal sensation are most sensitive to compression and, therefore, *one of the earliest signs of a cerebellopontine angle tumor may be unilateral decreased corneal sensation*. Later, more general decreased facial sensation can occur due to CN V compression. The facial nerve (CN VII) travels along with CN VIII in the internal auditory canal in the petrous portion of the temporal bone and is severely distorted by acoustic neuromas, however, facial droop is usually a late symptom of this tumor. Rarely, diminished unilateral taste may be reported, as in our patient. As the tumor continues to grow, cerebellar and corticospinal pathways are compressed causing ipsilateral ataxia and contralateral hemiparesis, respectively. Impairment of swallowing and the gag reflex (CN IX and X) and unilateral impaired eye movements (CN III and VI) occur only in very large tumors. Ultimately, if left untreated, the tumor compresses the fourth ventricle, causing CSF outflow obstruction, hydrocephalus, herniation and death.

#### Clinical Course:

The ENT specialist suspected an acoustic neuroma, or possibly a metastatic lesion in the left cerebellopontine angle. She, therefore, referred the patient to a neurosurgeon for further evaluation. In the meantime, she scheduled:

1. an audiometric exam which showed greatly diminished hearing in the left ear, particularly for higher tones,
2. a nuclear bone scan which did not show any melanoma metastasis to the skeleton, and
3. an MRI scan of the head, described below.

MRI: *Please review the section on MRI scanning in the Introduction to Clinical Materials prior to this lab.*

MRI is clearly the test of choice in suspected lesions of the cerebellopontine angle, since posterior fossa structures can be imaged far more clearly with MRI than with CT where "shadowing" artifact occurs due to dense bones at the base of the skull. In addition, MRI can often pick up tiny acoustic neuromas that haven't even expanded beyond the internal acoustic canal, allowing early intervention and treatment. Coronal, sagittal and axial (horizontal) images of the brain were obtained with and without intravenous magnetic contrast medium (gadolinium). The slide shows images obtained in the spin echo mode with TR = 500 and TE = 20. Are these T1 or T2 weighted images? Hint: note that the CSF appears dark, the white matter appears white and the gray matter appears gray. The top two panels show horizontal cuts through the mid cerebellum without (left) and with (right) gadolinium. Identify the mass in the left cerebellopontine angle which enhances with contrast due to increased vascularity. Note that the mass is located entirely outside the brainstem and causes distortion of the pons and fourth ventricle. In addition, note that the mass has a lateral knob that appears to emerge from the internal auditory meatus in the petrous portion of the temporal bone. Fortunately for the patient, this scan is virtually diagnostic of an acoustic neuroma. There is no evidence of melanoma metastases. The lower two panels show left (left lower panel) and right (right lower panel) parasagittal images, both without gadolinium. The acoustic neuroma can be seen in the left but not in the right parasagittal image. Use these images to review the course of each structure that can be compressed by acoustic neuromas as described above.

#### Hospital Course:

The patient was seen by the neurosurgeon in the office, and a short time later, admitted for neurosurgical removal of the tumor. The left occipital bone was opened behind the transverse sinus, the dura was opened, and the left cerebellar hemisphere was gently retracted to reveal the tumor. The tumor was carefully dissected away from the adjacent cerebellum, pons, CN V, VII, IX and X, and branches of the posterior inferior cerebellar artery. The functioning of the facial nerve was monitored continuously during the resection using a stimulating electrode placed on CN VII and EMG (electromyography) leads placed in the orbicularis oculi and labial muscles

.Thus, although the facial nerve was severely distorted by the tumor, its function was preserved. CN VIII, however, was sacrificed as it was completely encapsulated by tumor.

Pathology Report:

Acoustic Schwannoma (Neuroma).

Post-operative course:

The patient suffered from vertigo and had nystagmus at rest for 1 to 2 days post-operatively which then resolved. She also had a complete left facial paralysis which resolved over the course of several months and has done well subsequently. Fortunately, if acoustic neuromas are removed early, a surgical cure can be obtained.