

ANSWER CASE HISTORY VII

1). Where is the lesion, and what structures are responsible for his symptoms and signs?

The inability to look to the right with either eye, signifies a loss of either the abducens nucleus or the PPRF on the right. The right facial weakness represents a lower motor lesion of the facial nerve, most likely from involvement of the nucleus of motor VII or the axons of the nerve as they pass over the abducens nucleus. The contralateral nystagmus suggests damage to the right vestibular nerve or nuclei (paralytic nystagmus) and the hearing deficit points to a lesion of the right cochlear nerve or nuclei. The falling to the right and the tremor point to a lesion of the right inferior or middle cerebellar peduncle.

Dizziness may mean many things and in this patient it represents true vertigo, which is an illusion of movement, the sensation of either the individual or environment rotating. Vertigo is often associated with nausea and trouble walking. Let's refer to this case as "facial colliculus-vestibulocochlear".

2). What is the etiology of the lesion?

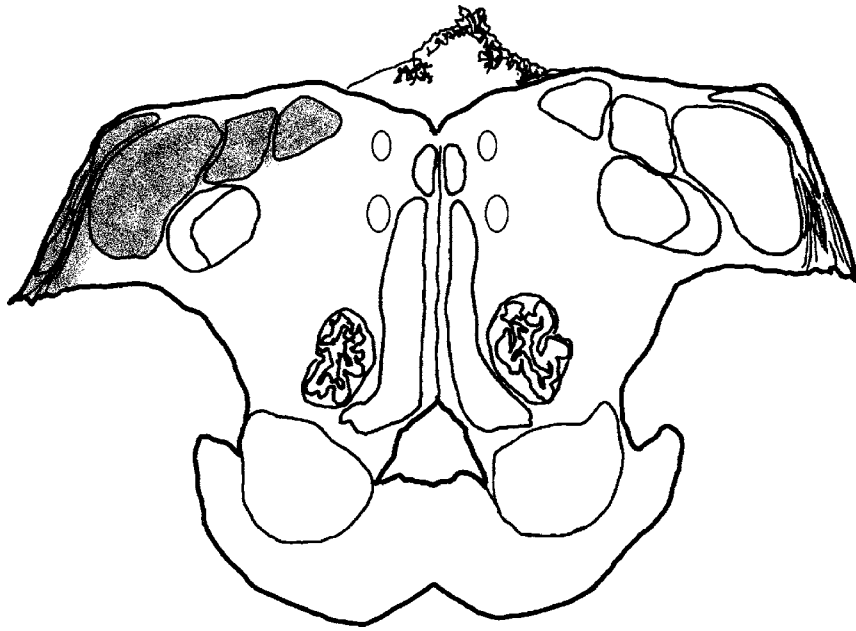
A relatively acute onset with some progression implies a vascular etiology and with the progression thrombosis must be considered. The vascular territory of this deficit is the anterior inferior cerebellar artery (see Brain stem and Cerebellar dissection lab). A hemorrhage is another concern. Occasionally tumors present with a rapid onset and may mimic a stroke. In this age group, metastatic lesions are often a concern.

3). How can the lesion be confirmed?

CT scans rarely define brain stem ischemic infarctions, but they can detect hemorrhages. The MRI scan is the best way to define brain stem and cerebellar anatomy.

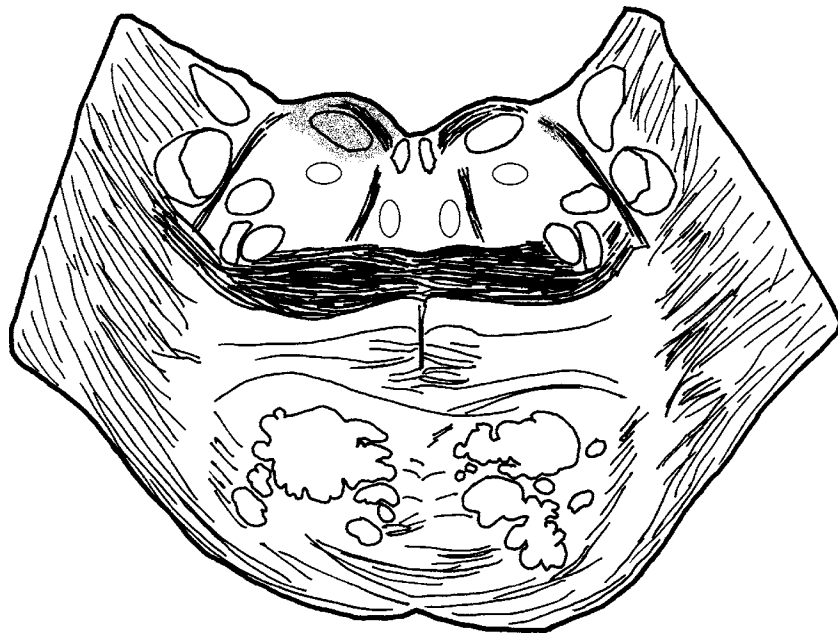
4). What treatment should be considered? What treatments may be developed to treat stroke?

Anticoagulation with heparin is a reasonable approach when there is clinical evidence to suggest ongoing clot formation. This should not be started until a hemorrhagic etiology is ruled out. Tissue plasminogen activator factor (tPA) is a therapeutic option for acute thrombosis, the main risk being changing an infarction into a hemorrhagic infarction. Ways to prevent sequelae of ischemia are also being developed. These include glutamate receptor antagonists to prevent excitotoxic damage and free radical scavenger treatments to prevent oxidative injury from any free radical formation produced by ischemia.



RIGHT

LEFT



RIGHT

LEFT

ANSWER CASE HISTORY VIII

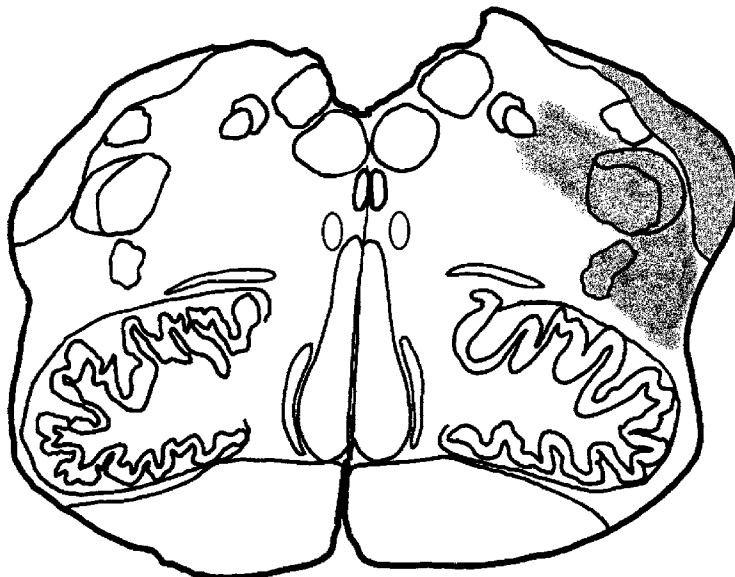
A 55-year-old woman noticed that she suddenly became nauseated and experienced vertigo. Her voice was hoarse and she was having difficulty swallowing. She also noticed a burning pain in and around the **LEFT** eye. Three days later the pain had subsided but she decided to see a neurologist. On her way to the hospital she started hiccuping and her husband noticed that her pupils were not symmetrical.

Upon examination it was found that both muscle tone and reflexes were normal and equal in all four extremities. The nausea and vertigo had subsided and no nystagmus was present. Taste was normal on both sides of the tongue. There was **NO** detectable paralysis of the jaws, facial muscles, or tongue, but an inspection of the pharynx showed that the uvula was directed toward the **RIGHT**. Laryngoscope investigation also suggested paralysis of the **LEFT** vocal cord. There was loss of pain and temperature sensations on the **LEFT** side of the face and **RIGHT** side of the entire trunk including the arm and leg. The exam revealed that her **LEFT** pupil was smaller than the right and the **LEFT** side of her face was dryer and warmer than the right side. The patient was no longer hiccuping.

LATERAL MEDULLARY (WALLENBERG'S) SYNDROME

The loss of pain and temperature sensation on the **RIGHT** side of the body results from interruption of the anterolateral system or spinothalamic tracts on the **LEFT**. The descending autonomic fibers destined for the lateral cell column have also been interrupted on the **LEFT**, and this accounts for the constricted pupil in the **LEFT** eye and the warm and dry (**LEFT** side) face. The initial pain in the **LEFT** side of the face, followed by the loss of pain and temperature from the **LEFT** side of the face, indicates involvement of the **LEFT** spinal nucleus and tract V. The paralysis of the **LEFT** vocal cord and deviation of the uvula to the **RIGHT** indicate damage to the **LEFT** nucleus ambiguus. The **hiccup** resulted from the lesion invading the area ventral and lateral to nucleus and tractus solitarius. The tremor relates to cerebellar lesion or left inferior cerebellar peduncle involvement.

In this case, a thrombus (a blood clot obstructing a blood vessel) had developed in the posterior inferior cerebellar artery. This led to the destruction of a region of the **LEFT** medulla which centered on the region including the spinal nucleus and tract V, nucleus ambiguus the anterolateral system. The initial nausea and vertigo resulted from slight irritation of the vestibular nuclei (they lie slightly rostral and dorsal to the main focus of the lesion)



ANSWER CASE HISTORY IX

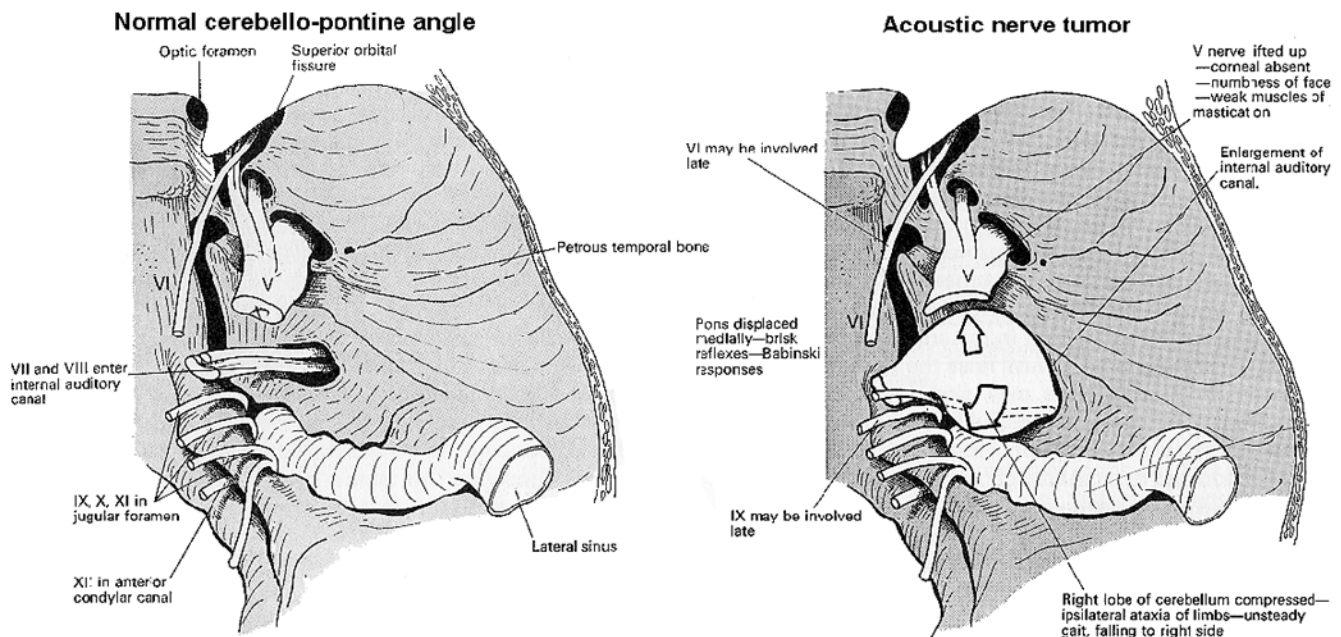
A young girl, aged 14, was admitted to the hospital for diagnosis. According to the history, her birth had been normal and uncomplicated. There had been no unusual illness or accidents during her childhood, and her growth and development were normal. About a year before this admission, the girl began to complain of “ringing or buzzing” in her **RIGHT** ear, a condition which had persisted since. As time went on the girl began to experience vertigo (sensation of turning) and nausea. When walking she tended to veer to the **RIGHT**.

Examination revealed that there was almost 100 percent loss of hearing in the **RIGHT** ear. Sometimes there was a spontaneous slow conjugate movement of the eyes to the **RIGHT**, and a fast return movement to the **LEFT**. When there was no spontaneous nystagmus, caloric stimulation of the **RIGHT** ear with either cold or hot water had **NO** effect. However, stimulation of the **LEFT** ear with **COLD** water resulted in **RIGHT** nystagmus while stimulation with **WARM** water elicited **LEFT** nystagmus. All muscles on the **RIGHT** side of her face showed evidence of weakness. There was a loss of taste from the **RIGHT** side of the tongue and a “dry” (no tears) **RIGHT** eye. Some incoordination of the **RIGHT** arm and leg was also apparent.

ACOUSTIC NEUROMA

The ringing (**tinnitus**) and later deafness in the **RIGHT** ear is due to involvement of the auditory portion of the **RIGHT C.N. VIII**. The vertigo and nystagmus are related to involvement of the vestibular portion of the **RIGHT C.N. VIII**. The weakness of the muscles on the **RIGHT** side of her face, the loss of taste from the **RIGHT** side of the tongue and the loss of tearing in the **RIGHT** eye indicate involvement of the **RIGHT C.N. VII**. The incoordination of the **RIGHT** arm and leg is due to involvement of the overlying cerebellum on the **RIGHT**.

The findings suggest a tumor of the posterior fossa on the **RIGHT** side, and surgery was recommended. At operation, a tumor was found in the **RIGHT** cerebellopontine angle, involving C.N.s VII and VIII. There was also some pressure on the inferior part of the cerebellum. This is a typical **ACOUSTIC NEUROMA**. The usual acoustic neuroma (nerve tumor) in adults is a schwannoma that originates from the vestibular portion of C.N. VIII just within the internal auditory meatus. The tumor grows and extends into the posterior cranial fossa to occupy the angle between the cerebellum and the pons (cerebellopontine angle).



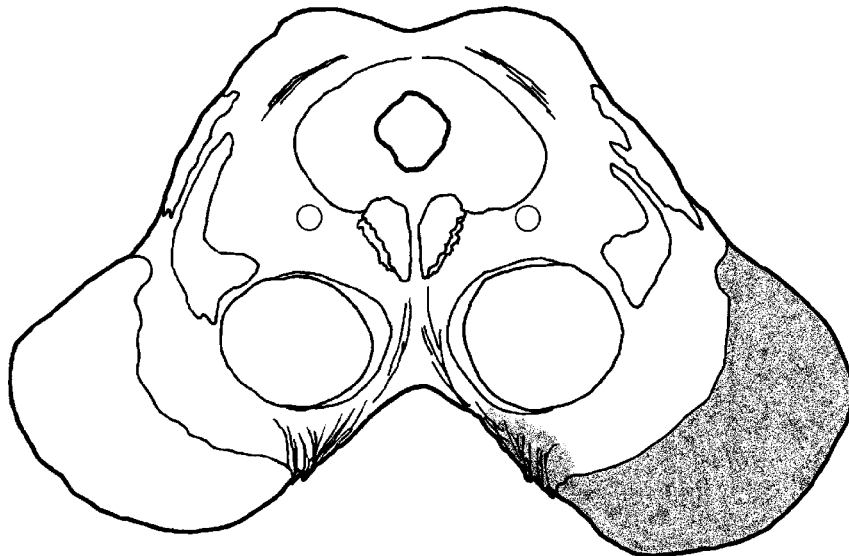
ANSWER CASE HISTORY X

Upon examining a 55 year old male, the neurologist found weakness of the **RIGHT** arm and leg; a Babinski sign was present. Examination of the tongue in the mouth revealed no atrophy or wrinkling, but upon protrusion the tongue deviated to the **RIGHT**. There was marked weakness of muscles in the lower half of the face on the **RIGHT**, but no atrophy of these muscles. There was a ptosis of the **LEFT** eyelid. When the neurologist lifted the eyelid, the pupil was dilated and the eye was turned down and out.

HINT! THERE IS ONE LESION SITE.

WEBER SYNDROME OR ROSTRAL ALTERNATING HEMIPLEGIA

The hemiplegia of the **RIGHT** arm and leg and the Babinski sign are due to involvement of corticospinal fibers in the **LEFT** cerebral peduncle. The tongue deviation to the **RIGHT** and the weakened muscles of the lower face on the **RIGHT** are due to the interruption of corticobulbar fibers in the **LEFT** cerebral peduncle. The dilated pupil in the **LEFT** eye, ptosis of the **LEFT** eyelid and outward and downward deviation of the **LEFT** eye are due to involvement of CN III (remember, the levator palpebrae elevates the eyelid). The lesion involves the **LEFT** cerebral peduncle and the rootlets of the **LEFT** CN III.



RIGHT LEFT

ANSWER CASE HISTORY XI

1. Where is the lesion and how can you explain the patient's complaints and the findings on exam?

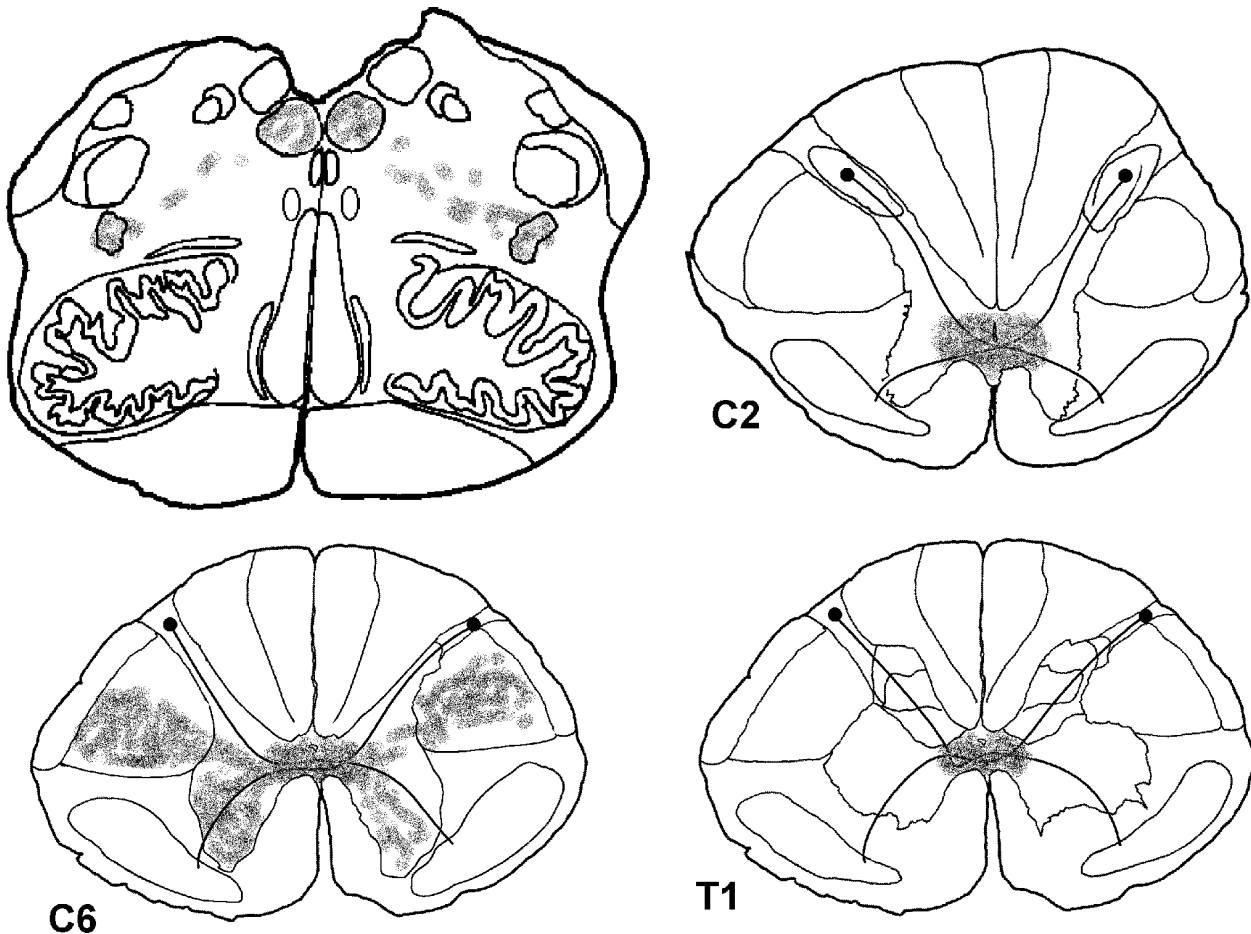
The tongue findings suggest a lower motor neuron lesion on the right (either nerve or nucleus) and the decrease gag, nasal speech, and hoarseness point to a lower cranial nerve or medulla lesion (nucleus ambiguus). The weakness in the upper extremities is consistent with a lower motor lesion; however, in the lower extremities the findings suggest an upper motor lesion. The sensory findings point to a lesion affecting the crossing fibers of the spinothalamic pathway (ALS) including the descending sensory fibers of cranial nerve V. The urinary symptoms suggest a bilateral interruption of descending motor pathways to the bladder (travel medial to the LCST) and the symptoms are consistent with the acute phase of an upper motor neuron (spastic bladder). The lesion must extend from around C7-8 up to the medulla in the central gray matter of the spinal cord and floor of the IV ventricle.

2. What are the possible etiologies?

A tumor extending in the area of the deficits is a possibility. The relatively slow progression is in keeping with a tumor and not a vascular lesion. Another possibility is a syrinx, a fluid filled cavity that may communicate with the central canal or IV ventricle.

3. How would you evaluate this patient?

The best way to evaluate this patient would be to obtain an MRI scan of the cervical spine and medulla.



SYRINGOMYELIA

DISCUSSION

Syringomyelia is a cystic cavity that develops in the central region of the spinal cord. This lesion is usually seen in the cervical segments of the spinal cord. However in some patients it may involve the medulla oblongata and then it is called syringobulbia and in some instances it may extend to the thoracic or lumbar cord. Sometimes syringomyelia may be associated with an abnormality of the posterior fossa structures (cerebellum, brain stem) called a Chiari malformation.

Although many pathogenetic mechanisms have been postulated, the etiology of syringomyelia is unknown. The clinical findings on neurological examination are explained by the localization of the lesion. The cystic lesion involving the central gray matter leads to dissociative sensory loss since it disrupts the fibers of the spinothalamic tract which conduct pain and temperature stimuli and decussate in the spinal cord. Lateral extension of the cavity may cause compression of the lateral corticospinal tract(s) and leads to spasticity, weakness and hyperactive reflexes as well as upgoing toes to plantar stimulation (Babinski's sign). Sphincter function may be affected. Sometimes intermediolateral regions of the lower cervical and thoracic cord, where the sympathetic pathways are localized may be damaged and this may cause decreased sweating or development of the Horner's syndrome (miosis, ptosis, anhydrosis). The muscle wasting and weakness is caused by forward extension of the cavity which leads to damage of the ventral horn cell.

Several neurological conditions may present with similar clinical findings and have to be differentiated with syringomyelia. The most important one to remember is to rule out an intramedullary tumor.

Some cases of syringomyelia may be treated by surgical decompression of the spinal cord and brain stem or by drainage of the cavity.