CLINICAL VIGNETTES FOR A LIFETIME (I HOPE!!)

As you near completion of this course I want you to begin to think in terms of "Case Vignettes" instead of "facts." In the front of Module 3 I have included the 14 case scenarios listed below. They are:

1. Duschenne's Muscular Dystrophy
2. Guillain-Barre Syndrome
3. Spinal cord hemisection-Brown Sequard Syndrome
4. Lateral Medullary (Wallenberg) Syndrome
5. Acoustic Neuroma
6. Weber Syndrome (Rostral Alternating Hemiplegia)
7. Cerebellar Cases (2)
8. Broca's Aphasia
9. Huntington's Disease
10. Cataract
11. Several Hearing Loss Cases
12. Thalamic Lesion-Pure Sensory Loss
13. Subdural Hematoma
14. "David and Goliath" (my favorite!!)

There are **nine** more vignettes on this www. site that you should look over for Exam #3. While at first glance your cortisol and NE might go up, I think you will enjoy reviewing these cases since they are associated with material that we have covered this year.

When the 9 scenarios below are added to the 14 above, you will have 23 neuro vignettes in your long-term memory (your "blackboard" can't hold them all long enough!).

Please go for the "Big Picture" on all of the Clinical Vignettes, as that is my goal in having you review them. I have given the "Big Picture" for the 9 "new" vignettes discussed below, so I will leave the other 14 (above) up to you.

15. Clinical Vignette--ALZHEIMER'S

A 52 year-old man is noted to have memory difficulties. He forgets to pay several bills and forgets numerous business appointments. His wife reports that he appears depressed and is not his usually jovial self. He acts strangely and inappropriately and has an affair with a 15 year-old high school sophomore. Neurological examination shows the following abnormalities.

1. disorientation for year, month, and day
2. inability to remember 3 unrelated objects at 15 minutes
3. inability to accurately draw geometric shapes
4. inability to think abstractly.

CT and MRI show no evidence of brain atrophy; MRI-SPECT (spectroscopy) shows reduced metabolism/blood flow in frontal, temporal and parietal regions bilaterally.

JKH Comment:

This patient has dementia, most likely of the pre-senile Alzheimer type. In the early stages of this disorder there may be clinical evidence of cognitive or intellectual deterioration; however, CT and MRI may not yet show evidence of brain atrophy (shrinkage). In the later stages there should be anatomical evidence of brain atrophy and pathological studies would show loss of neurons. There are other pathological changes e.g. neurofibrillary tangles, amyloid deposits. MRI-SPECT shows physiological evidence of reduced perfusion and presumed reduced metabolism. This is more sensitive test than CT/MRI. Although there is reduced brain perfusion, techniques to increase cerebral blood flow will not enhance brain function. The reduced brain perfusion is a manifestation of primary neural depression; therefore as there is reduced brain mass, there is need for reduced blood flow. In Alzheimer dementia, there is a central cholinergic disorder. Drugs that enhance acetylcholine function are effective in enhancing memory (physostimine is an inhibitor of the catabolic enzyme of ACh and therefore increases the level of ACh; carbachol=cholinergic agonist).

Big Picture!

DISORIENTATION FOR YEAR, MONTH AND DAY; ACTS "STRANGELY:" FORGETS; NEUROFIBRILLARY TANGLES AND SENILE PLAQUES; LOW ACETYLCHOLINE

16. Clinical Vignette--PARKINSON'S

Louise Bermel is a 60 year old female who presents to the office with difficulty using her right arm. Ms. Bermel states that the difficulty in using her right arm has occurred over the past 2 months. She is right-handed. The clumsiness and intermittent tremor have become progressively worse. No other symptoms are present. To direct questioning she says the right shoulder aches, "it may be bursitis," and that she has noted saliva on her pillow in the mornings. Her children feel she is slower and looks depressed. She denies any visual disturbances, vertigo, palpitations, sensory symptoms or unconsciousness. She denies headaches and there is no history of trauma.

The problems with her shoulder and arm are making it more difficult for her to carry out her work. There is no history of neurological diseases in the family except for her mother who had a slight shake of her head in old age. There is no history of early cerebrovascular or coronary artery disease in the family. There is no family history of hypertension. Her father died of pneumonia and her mother died of cancer of the breast.

Neurologic Exam: Normal cranial nerves II-XII and finger to finger and heel to shin on gait testing (cebellar). Her right arm swings less and on turns her neck looks stiff. Deep tendon reflexes, sensation, and fine coordination are almost normal. There is no significant weakness. Initiation of gait is slow (what is this called?) and a little unsteady, but when pulled from behind she does not lose her balance. There is a slight tremor of her right hand intermittently at rest that disappears when she reaches for something. It is also noted when she is walking.

JKH Comment:

This is a case of Parkinson's.. It often begins unilaterally. An MRI could be done, but the presence of a tremor would interfere with getting a good exam. Treatment is designed to maintain normal function--the lifestyle to which she was accustomed--which has tremendous psychological benefit for her. The surgical possibilities are pallidotomy (lesion of Gpi) , deep brain stimulation of the subthalamic nucleus (mess up its firing!) and fetal transplantation of cells that will produce DA..

Big Picture

SLOW INITIATION OF GAIT; SLOWER AND DEPRESSED; RESTING TREMOR; REFLEXES AND SENSATION NORMAL

17. Clinical Vignette LOW BACK PAIN

A 45 year old delivery man comes to see you complaining of low back pain that has been intermittent for the past 6 months. The pain is in the middle of the lower back and usually radiates into the left buttock. The pain is made worse by sneezing, coughing, or when he hits a pothole while driving. In some cases these maneuvers cause the pain to radiate down the back of his left leg into the bottom lateral aspect of his foot (what a clue!!). Over the past 6 weeks, he has noted that it is difficult for him to stand on his tip toes (what a great second clue!!) and that this is primarily because of weakness in his left foot.

On exam he has a normal neurological exam except that his left ankle jerk is absent and he has weakness of his left gastrocnemius. There is abnormal sensation over the lateral aspect of the left foot. He cannot stand on his toes of his left foot. When you have him lying down, you cannot elevate his left leg above 35 degrees because of shooting pain into his left buttock and down the back of his left leg.

JKH Comment

The clinical picture is compatible with a left S1 radiculopathy and most likely it is due to a L5-S1 herniated disc. The needle exam would reveal signs of denervation in the S1-innervated muscles, particularly the gastrocnemius, glutei, and the S1 paraspinous muscles. NCV (nerve conductin velocities) would probably be normal. To define a herniated disc and make sure that this was not some other problem, an MRI of the spine would be appropriate. Abnormal MRIs of the lumbosacral spine are common and must always be interpreted with respect to the clinical picture. Symptomatic medical treatment is probably as successful as surgical decompression and the patient's condition will improve. Surgery is favored when an active process associated with denervation and weakness is combined with disabling pain. If the EMG does not indicate active denervation (lower CMAP for instance), conservative medical therapy may be more reasonable.

Big Picture

RADIATING PAIN IN DERMATOMAL DISTRIBUTION; WEAKNESS; HYPOTONIA OR ATONIA; ABNORMAL SENSATION

18. Clinical Vignette-Amyotropic Lateral Sclerosis

A 65 year old man presents with a six month history of progressive fatigue, weakness and leg cramps. On a few occasions he choked on food. His wife noted diffuse twitching of muscles on his chest and upper back. Two months ago he developed a foot drop in his left leg. He has not complained of any sensory symptoms. There has been no cognitive decline. He has no difficulty with bowel or bladder function. His family history is noncontributory.

Examination showed that the patient had normal mental status. Motor examination showed severe, bilateral diffuse muscle wasting in both upper and lower extremities. The most atrophied were the deltoid, triceps, biceps, hand muscles and quadriceps on either side and the left anterior tibialis. There were prominent fasciculations (WHAT ATE FIBRILLATIONS?) in all muscle groups. The muscle tone was increased in both upper and lower extremities. There was diffuse weakness in all 4 extremities with complete left foot drop. Neck extensors were profoundly weak so that the patient was barely able to keep his head up. The tendon reflexes were hyperactive in all four extremities. The plantar reflexes were extensor (Babinski sign). **Sensory examination and coordination were normal**. His gait was characterized by decreased arm swing and limping of the left leg.

JKH Comment

This is a case of amyotrophic lateral sclerosis. The increased muscle tone, hyperreflexia and positive Babinski sign indicate upper motor neuron (UMN) damage (lateral corticospinal tract). Severe, diffuse muscle wasting and fasciculations are suggestive of a lower motor neuron (LMN) involvement (anterior horn cells). In amyotrophic lateral sclerosis, not all of the LMNs die at the same time (the disease is progressive). Loss of anterior horn cells leads to atrophy of their target muscles (therefore no spasticity), while those ventral horn cells that are still functional will cause spasticity in their targeted muscles due to the loss of the LCST input, an upper motor neuron deficit.

Big Picture

FASCICULATIONSAND ATROPHY (LMN PROBLEMS) COMBINED WITH UMN PROBLEMS LIKE A BABINSKI SIGN, SPASTICITY, INCREASED TENDON REFLEXES; NORMAL SENSATION AND COGNITION. DO YOU THINK THE PATIENT HAS CLONUS?

19. Clinical Vignette--Syringomyelia

A 56-year old woman has a long history of numbness in and clumsiness of her hands. Initially the numbness started in her right hand but progressed slowly to include both hands and both arms. Over the past year the numbness has actually included the shoulders, neck, scalp and most recently the area of the face next to the hairline and the angle of the jaw on the right. Besides the numbness, she has noted increasing difficulty in opening jars and over the past 2-3 years has had trouble brushing her hair. Most recently she has begun to have trouble with slurred speech, swallowing, and a hoarse voice.

Her other complaints include trouble with walking. It seems that she is having trouble with tripping over her toes. She has smoked cigarettes for many years and recently stopped because she has burned her fingers a number of times and did not feel the pain **(big clue!).** She had to have a skin graft of her right hand because of a severe burn she sustained from a stove.

Her exam revealed a normal mental status. Cranial nerve exam showed a bilateral decrease in pin prick sensation at the hair line and angle of the jaw. The palate moved poorly and her voice had a nasal characteristic. Her gag reflex was depressed. There was some atrophy of the left side of her tongue with fasciculations noted. The motor exam revealed diffuse weakness and atrophy of the upper extremities. There were some fasciculations noted in the biceps bilaterally. The reflexes in the upper extremities were barely obtained. In the lower extremities there was mild weakness and increased tone and reflexes. Bilateral Babinski signs were noted. The patient's gait was stiff and she could not walk on her heels. The sensory exam in the lower extremities was normal. The sensation in the upper extremities was intact to vibration and joint position sense but she could not feel pin prick in either upper extremity or across the shoulders, the neck or the scalp.

JKH Comment:

This is a case of syringomyelia, which is a cavitation/gliosis in the central canal (and interruption of the crossing dorsal horn fibers headed into the ALSs or lateral spinothalamic fibers; LSTT) of the spinal cord. The destructive process has obviously moved rostrally into the brain stem. 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 medullary lesion (remember nucleus ambiguous??). 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) in the spinal cord and the right spinal nucleus V.

Big Picture

"CAPE-LIKE" DISTRIBUTION OF PAIN (PIN PRICK) AND TEMPERATURE LOSS DUE TO DAMAGE TO CROSSING ALS/SPINOTHALAMIC FIBERS IN THE SPINAL CORD. ALSO CAN SPREAD INTO THE BRAINSTEM WHERE THERE IS LMN AND OTHER DAMAGE. DON'T CONFUSE THIS WITH "GLOVE AND STOCKING" DISTRIBUTION SEEN IN NEUROPATHIES.

20. Clinical Vignette--Wernicke-Korsakoff Syndrome

A 50 year-old CEO develops abdominal pain and is found to have a tender abdomen on examination. Liver function studies are markedly abnormal and serum amylase is markedly elevated. An abdominal CT shows a mass. At surgery, he is found to have a pancreatic pseudocyst. (The pseudocyst is, by far, the most common cystic mass of the pancreas. Approximately 10% of patients develop pseudocysts following several episodes of alcohol induced pancreatitis [severe complications of pancreatitis are more common when alcohol induced]). These cysts are usually noncomplicated. If complicated, and there is no history of pancreatitis, the diagnosis of mucinous cystic neoplasm must be entertained. This may seem to be an obvious distinction, but many mucinous cystic neoplasms carry a diagnosis of pseudocyst before the correct diagnosis is made.. In the surgical recovery room, he awakens and is delirious and tremulous. He is treated with CNS tranquilizers and this calms him. Two days later, he awakens but his recent memory is quite poor but remote (past) memory is intact. He can not remember any of three unrelated objects at 10 minutes.

JKH Comment:

Based upon the clinical features, this patient most likely has alcohol-induced damage even though no statement is made to suggest "chronic alcoholism." Initially, he is "delirious" and "tremulous" and this suggest major alcoholic withdrawal syndrome which occurs after abrupt cessation of alcohol. As the patient awakens and shows poor recent memory (**like H.M**.), this is consistent with **Wernicke-Korsakoff Syndrome**. One way of telling this disease from anoxia of the hippocampi (both exhibit the **absence of new memories**) is that in the Korsakoff's there is "**confabulation**,**"** that is, the patient tries to cover his memory loss by making up stories.

Pathological changes include necrotic and hemorrhagic microscopic lesions involving the **mammillary** bodies (memory problem due to connections with hippocampus via fornix) periaqueductal midbrain, paraventricular thalamus, and hypothalamus. This disorder

results from specific deficiency of vitamin B-1 (thiamine). This is an essential cofactor for decarboxylation of pyruvate and alpha-ketoglutarate. Replacement therapy with thiamine should cause improvement in memory function.

Big Picture

TOO MUCH ALCOHOL!! (DON'T FORGET ABOUT ALCOHOL'S EFFECT ON THE MEDIAL ZONE (VERMIS) OF CEREBELLUM). PATIENT HAS SIMILAR MEMORY PROBLEMS AS H.M AFTER HIS BILATERAL HIPPOCAMPAL REMOVAL BUT IN ADDITION EXHIBITS "CONFABULATION." LESION INVOLVES MAMMILLARY BODIES AND PERHAPS ADJACENT HYPOTHALAMUS AND THALAMUS.

21. Clinical Vignette-Multiple Sclerosis

Leslie Biggins, a twenty-eight year old, male, graduate student presents complaining of double vision when he looks to the right for the past three days. Mr. Biggins states that the double vision was first noticed upon awakening three days ago. He feels fine and has no other complaints. He has never been ill with the exception of an occasional upper respiratory infection and the usual childhood diseases of measles, chickenpox and mumps. His family history is negative except for hypertension in his father, which is well controlled with medication. His maternal grandmother was afflicted with "rheumatism" and was in a wheel chair for many years before her death.

He reports no headaches, muscle weakness, dizziness or sensory anomalies. He sleeps poorly and is up most of the night because of a need to urinate frequently. He feels that this is a direct consequence of caffeine intake. Two months ago he became incontinent of urine in bed, and he has cut off coffee altogether. He feels that this has improved matters somewhat, but not entirely. He has noticed an inability to concentrate and fatigue has become an inordinate problem.

He is not currently taking any medication and does not smoke or drink. He is a graduate student in clinical psychology and is under much pressure to finish his dissertation. His wife is anxious to start a family. He admits to being "stressed out" and depressed. This has had a significant negative impact regarding his relationship with his wife and recently he has had problems with impotence.

Neurological Examination: (I have noted only important ones!)

Mental status: Mr. Biggins is cooperative and is oriented as to place and time. He has appropriate memory for immediate (working), recent (hippocampal) and past events. He is able to compute arithmetically and can attend to similarities and differences.

Eyes: On right gaze the right eye abducts but the left eye cannot pass the midline. Nystagmus is present in the right (good) eye during right lateral gaze. On left gaze both eyes turn left normally without nystagmus or diplopia.

Motor System--Upper and lower limbs are strong.
Tone in both lower extremities is elevated. Clonus is elicited in the left ankle.
Sensation: Proprioception, vibration, pain, and temperature are all normal, as is stereognosis and two-point-discrimination.
DTR (deep tendopn reflexes): Symmetrical and 3+.
Babinski + Bilaterally.
Cerebellar funct.: Gait, toe-heel, finger-nose, rapid alternating movement are normal and symmetric.

The MRI shows multiple high signal (white) white-matter lesions. The lesions are mostly around the lateral ventricles. A number of the lesions are oval in shape and some lesions also appear as dark "holes" in the T_1 images (sequences when the ventricles are dark).

JKH Comment

This is multiple sclerosis. Mr. Biggins was placed on prednisone, 60 mg per day. The diplopia resolved by the tenth day of treatment and the dose was tapered and discontinued. Hopefully you remember what happens with a lesion (demyelination) of the left MLF (can't turn left eye medially to the right). Moreover, both LCSTs (increased reflexes, clonus, and Babinskis) are exhibiting demyelination.

Big Picture

THE KEY HERE IS MULTIFOCAL LESIONS. WE HAVE EMPHASIZED IN THE PROBLEM SOLVINGS SINGLE LESIONS THAT ACCOUNT FOR NUMEROUS CLINICAL DEFICITS. MS HITS ALL OVER THE PLACE AND COMMONLY AFFECTS THE MLF, LCSTS AND OPTIC NERVES.

22. Clinical Vignette--EPILEPSY

A two-year-old boy presents to the ER. Just after awakening, the mother noticed that his right arm, then leg, began to shake. He was also "out of it" but not unconscious; he would not respond to her. This continued while she brought the child to the ER. He had been fine that day except for feeling "warm." On arrival to the ER, the right-sided rhythmic jerking movements continued. The rest of the exam was significant for a temperature of 104^0 and a left otitis media. Past medical history was significant for speech delay, as he did not yet have any words beside "mama" and "dada."

JKH Comment:

The two-year-old boy presented above has a **simple partial seizure** involving predominantly the left hemisphere (right sided jerking movements). The MRI showed a subependymal heterotopia on the left. Gray matter heterotopias are collections of normal neurons in an unusual location, due to an arrest in fetal neuronal migration. Because of its localization, these neurons have escaped the cortical GABA-ergic inhibition that normally prevents over excitation. Because of this lack of inhibition, these misplaced neurons are highly epileptogenic. Increased body temperature, such as one sees during a fever, increases the excitability and can provoke a seizure.

Big Picture

THE KEY HERE IS THE MAINTENANCE OF CONSCIOUSNESS WITH THE RHYTHMIC JERKING OF THE CONTRALATERAL LIMBS.

23. Clinical Vignette—Myasthenia Gravis

A 25 year old woman came to your office complaining of intermittent double vision for the last three weeks. She also has complained of fatigue. She has felt best during the early morning hours, but later, during the course of the day, she gradually develops double vision and diffuse weakness. Her boy friend has observed that her right eyelid has been drooping frequently. She used to play competitive basketball while in college, but now she has been short of breath after climbing only 2 flights of stairs. Also, on a few occasions, she choked on food and her friends noted that her speech was slurred or thick.

On examination her mental status was normal. She had marked ptosis on the right side. Eye movement examination showed decreased movement in all directions in the right

eye. There was also slightly decreased abduction in the left eye. On repetitive blinking she developed ptosis of the left eyelid and her right-sided ptosis got much worse. Motor examination showed normal muscle bulk and tone. Muscle testing revealed that she was initially strong, but rapidly became "tired" or weak with repeated effort. She was unable to hold her arms abducted at 90 degrees for more than 30 seconds. All sensory modalities, reflexes, coordination and gait examination were normal. Plantar reflexes were flexor.

JKH Comment:

Myasthenia gravis is an immune mediated disorder of neuromuscular transmission. This condition is characterized by fluctuating weakness and fatigability of voluntary muscles. In the vast majority of cases, extraocular muscles are involved. Some patients may also have involvement of the muscles of the tongue, pharynx, and soft palate (dysphagia; difficulty swallowing and dysarthria; slurred speech). Weakness gets worse with any sustained activity and improves after a period of rest.

MG is caused by immune mediated destruction of acetylcholine receptors at the postsynaptic membrane and in some patients it may be associated with a thymoma (tumor) of the thyroid gland (about one in ten MG patients).

Big Picture

THE KEY HERE IS FATIGUE, EYE MUSCLE(S) WEAKNESS THAT GETS WORSE AS THE DAY PROGRESSES, DECREMENTAL CMAP and NORMAL SENSATION