PowerPoint 2:

Slide 1:

If you have a patient who has a complicated weakness that is involving just one extremity, and it is involving more than one nerve root and more than one peripheral nerve or muscle, then you have to consider the possibility of plexus lesion. This could be a brachial plexus lesion or a lumbar plexus lesion. In a plexus lesion you would expect to have lower motor neuron findings, so if it's a complex lesion with lower motor neuron findings and probably sensory involvement as well, you would probably have to consider plexus lesion. The EMG and nerve conduction study can be very helpful in the evaluation of a patient with a possible plexus lesion. If it appears to be complex and affecting multiple levels, if it were a root lesion, radiculopathy, we would expect the sensory nerve action potential (SNAP) to be intact because this would be proximal to the dorsal root ganglion. If it is a plexus lesion, you would expect the sensory nerve action potential (SNAP) to be affected because this would be distal to the dorsal root ganglion.

Slide 2:

I would recommend quickly reviewing the brachial plexus and the lumbosacral plexus prior to taking the shelf exam. I don't know that they necessarily ask whether it is a root, trunk, or a particular division involvement in the brachial plexus or lumbosacral plexus, but I do believe that it's important to formalize yourself with the organization of the lumbosacral plexus and brachial plexus before the shelf exam.

Slide 3:

Peripheral nerve lesions can cause weakness as well. Before taking the shelf exam, I would familiarize yourself with the major peripheral nerves of the upper extremities. For an example, the median nerve, which can cause carpal tunnel syndrome or the median neuropathy of the wrist. Patients typically have numbness and tingling over the lateral 3½ (approximately) fingers of the hand and can have weakness of thumb abduction because the median nerve supplies the opponens pollicis, abductor pollicis brevis, and flexor pollicis brevis and can drop things due to carpal tunnel syndrome. Often, patients with carpal tunnel syndrome have paresthesias when they wake up in the morning and have to shake out their hand.

Next we have the ulnar nerve which can get compressed at the wrist but is more commonly compressed at the elbow. There is sensory loss over the medial aspect of the hand in the ulnar distribution, and the ulnar nerve also supplies most of the intrinsic hand muscles to the hand so they often have weakness in the hands and drop things as well. What is interesting is we often think of carpal tunnel syndrome when a patient wakes up with paresthesias in the hand but you can also have paresthesias upon awakening with ulnar neuropathy. If the patient bends their elbow at night keeping their hand close to their face and head that can compress the ulnar nerve at the elbow and can cause worsening of the ulnar neuropathy symptoms.

You can also have peripheral nerve damage to the radial nerve, typically in something like Saturday Night Palsy, where the radial nerve gets compressed in the radial groove causing weakness of wrist and finger extension. They come in with a wrist drop. Generally these

patients recover relatively well unless they have any underlying diabetes or peripheral neuropathy which might result in some residual damage to the nerve. Those are the most common nerves to get compressed in the arm.

In the legs, we often think of sciatica, but most sciatica is actually caused by L5 or S1 radiculopathies. A more common peripheral nerve to be compressed in the legs would be the peroneal nerve. The deep peroneal nerve supplies dorsiflexion, toe extension, and the web between the toes. The superficial peroneal contributes to eversion as well as numbness over the lateral leg and dorsum foot. I see patients with a peroneal neuropathy that either have sensory loss over the lateral aspect of the lower leg or dorsum of the foot or both depending on which fascicles of the nerve are being affected. So, if you see a patient that has foot drop, weakness of dorsal flexion, your two primary considerations would be a peroneal neuropathy or an L5 radiculopathy. There are certainly other causes of foot drop like ALS and a variety of other conditions, but foot drop really should be considered L5 radiculopathy or peroneal neuropathy until proven otherwise.

Slide 4:

The next area in the neuro axis to cause weakness would be the neuromuscular junction. The most common neuromuscular junction syndrome would be myasthenia gravis. The lesion in myasthenia gravis is postsynaptically with antibodies to the acetylcholine receptor on the muscle membrane. This is an autoimmune process. The usually treatment for myasthenia gravis would be Mestinon or pyridostigmine. So, if they ask on the shelf exam what the initial treatment of choice would be, it would be pyridostigmine. They sometimes offer IVIg as an alternative on the shelf exam but we really just use IVIg or perhaps plasma exchange for refractory cases. The usual treatment would consist of pyridostigmine. We also use steroids in these patients but those are really not the first line use in myasthenia gravis. On the test they also ask for the diagnostic test of choice for the evaluation of myasthenia gravis, and in medical school we often here about the Tensilon test, which is edrophonium, a short acting acetylcholinesterase inhibitor. This can helpful in the evaluation of myasthenia gravis however it is not 100% positive for myasthenia gravis because it's been found in positive in patients with ALS, stoke, and other conditions. The diagnostic test of choice, and I hope I am not leading you astray, would be a nerve conduction study. In reality this would not be a typical nerve conduction study - This would be a repetitive stimulation. Clinically a myasthenic would get weaker with repetitive activity and better with rest. So, if you repetitively stimulate the nerve you can get a decremental response in amplitude on the repetitive stimulation. In myasthenia gravis, they typically present with more of a proximal more than distal weakness. They may have difficulty combing their hair or getting things off of a high shelf, getting out of the bathtub or other proximal muscle weakness.

They can also have bulbar symptoms, they can have the dysarthria, dysphagia, diplopia, and dyspnea. In a patient with acute onset bulbar symptoms you have to consider a vascular event, but if you have someone who has chronic bulbar symptoms that are perhaps getting worse, you have to consider motor neuron disease (ALS) or myasthenia gravis. So, if there is eye

involvement, diplopia or ptosis, then it is more likely to be myasthenia gravis because the ALS does not typically have the eye findings.

In myasthenia gravis, you can use other tests in evaluating such as an antibody panel, an acetylcholine antibody panel which includes binding, blocking, and modulating antibodies. There is a 4th antibody that can be checked, the MUSK antibody. I believe, though, that the answer on the exam would be a nerve conduction study, in particular repetitive stimulation.

Another neuromuscular junction disorder would be Lambert Eaton Myasthenic Syndrome. The lesion is presynaptically in the voltage gated calcium channel. They are not able to release acetylcholine into the synapse, causing weakness. In both myasthenia gravis and Lambert Eaton you are going to check a CT of the chest without contrast. In myasthenia gravis you are looking for thymoma, and in Lambert Eaton you are looking for small cell lung cancer, which is the most commonly associated with tumor. There have been some non-paraneoplastic causes for Lambert Eaton but it really should be paraneoplastic until proven otherwise. In Lambert Eaton they improve with activity, so if you were checking knee jerk reflexes on these patients they may have little to no response with patellar response initially, but if you have the patient extend the leg at the knee against resistance for 10 seconds, you can amplify the reflexes. Clinically they improve with activity, so they also have an incremental response with repetitive stimulation. In these patients they really have difficulty releasing the acetylcholine to pass on the message to the muscle membrane so acetylcholinesterase inhibitors are only marginally effective in these patients.

Slide 5:

The last section in the neuro axis cause of weakness would be muscle disorders. There are a variety of different myopathies, including inflammatory myopathy, endocrine and electrolyte disorders, metabolic myopathies, drugs and toxins, and infections. If you have an inflammatory myopathy you have to consider the possibility of polymyositis or dermatomyositis, so if you have patient with muscle tenderness, proximal weakness, and elevated CK with NO rash then you think about polymyositis but if there is an associated rash then consider dermatomyositis.

Myopathy patients often have associated cramping pain and muscle tenderness with their proximal muscle weakness. Myopathies can certainly cause proximal muscle weakness but another consideration for proximal muscle weakness would be a neuromuscular junction syndrome.

Also under muscle disorders would be acute neurologic symptoms from something like neuroleptic malignant syndrome. In this situation the patient would present with fever, muscle rigidity, altered mental status, and high CK, particularly after having taken an antipsychotic. This could be caused by either typical or atypical antipsychotic. The treatment would be to stop the neuroleptic, hydrate and cool the patient, and you can give benzodiazepines for agitation. You can also use dantrolene for treatment of neuroleptic malignant syndrome.

Serotonin syndrome is another consideration and the intensity of clinical findings reflects the degree of serotonergic activity. They can have mental status changes with anxiety, agitation,

delirium, restlessness, disorientation, and easy startle. They can have autonomic manifestations and neuromuscular hyperactivity.

With rhabdomyolysis, you can see this with crush injury, seizures, alcohol abuse especially if there is a hyperkinetic state with delirium tremens. You can see rhabdomyolysis with exertion especially with environmental heat illness, vascular surgery, and malignant hyperthermia.

Slide 6:

I would recommend quickly reviewing the dermatomal distribution using the dermatomal map that is provided in this PowerPoint. I would review this quickly right before the shelf exam. That way in the vignette, they give you a patient who has numbness over the proximal anterior thigh, then maybe it's an L2 nerve root lesion or whatever, but I would definitely recommend reviewing the dermatomal distribution map right before the shelf exam.

Slide 7:

I would also recommend quickly looking over the sensory distribution of major peripheral nerves of the upper and lower extremities and in particular the median, ulnar, and radial nerves in the arm and the peroneal nerve in the leg.

Slide 8:

It is helpful to review the localization associated with the different lobes of the brain. With a frontal lobe lesion you would expect to find contralateral weakness because the motor strip is located in the frontal lobe. You can also see urinary incontinence, particularly with bilateral lesions which makes sense if you think about a patient with normal pressure hydrocephalus and the dilated ventricles. If you think about the homunculus with the leg fibers being medial so the leg fibers are being dilated over the ventricles causing gait issues. You also have the micturition fibers being dilated as they extend over the ventricles as well. So, in normal pressure hydrocephalus you can explain the gait issues and the urinary incontinence. In the frontal lobe lesion you could see expressive aphasia if Broca's area is being affected, and classically you can see executive dysfunction and personality change particularly with the prefrontal lobe lesions.

Slide 9:

The parietal lobe is, I think, one of the most interesting lobes of the brain. You get all sorts of interesting findings like neglect, apraxia, and agnosia. With bilateral parietal lesions you get Balint's syndrome. You also get contralateral sensory loss, and visual field changes with an inferior quadrantanopia. You can also have apraxia and other interesting exam findings. If you are having trouble localizing a lesion and it's something odd, then perhaps it is coming from the parietal lobe.

Slide 10:

In the temporal lobe there can also be visual field changes. We typically see the "pie in the sky" defect or the superior quadrantanopia. You can also have language dysfunction such as a

receptive aphasia or Wernicke's aphasia. With bitemporal tip being affected you can have Kluver-Bucy syndrome or amnesia particularly with amygdala or hippocampal involvement and impaired recognition of facial emotional expression.

Slide 11:

With an occipital lobe lesion we get more significant visual field changes, in this case a homonymous hemianopsia. The farther back you go in the optic radiations, the more of the entire half of the vision would get affected. So there could be macular sparing in an occipital lobe lesion because this is a watershed area that is supplied by terminal branches of PCA and MCA. If you have the PCA infarct causing an occipital lobe infarct than you would macular sparing because of the MCA supply to the macula.

Slide 12:

Brainstem lesions are also very interesting. I would familiarize yourself with Wallenberg syndrome right before taking the shelf exam. They like the Wallenberg syndrome. I have never heard students say that they ask you about the Weber, Claude, or Benedict syndromes but I would definitely yourself with Wallenberg syndrome. So if you have a patient with crossed findings, with a loss of pain and temperature on ipsilateral face but contralateral limbs and trunk, those cross findings would definitely make you think about a brainstem lesion. Wallenberg syndrome would be caused by a lesion in the lateral medulla like a PICA or a vertebral artery infarct. They also get loss of vibration, proprioception, and ataxia on the ipsilateral limbs. They can have Horner's syndrome as well as other findings associated with Wallenberg.

Slide 13:

I would recommend familiarizing yourself with localization for various visual field cuts, particularly in the parietal lobe, temporal lobe, and occipital lobe. A quick review of this picture would probably be advantageous.