Neurologic Consultation - 5

Specialty: Consult - History and Phy.

Description: Approximately one and a half years ago, patient fell down while walking in the living room from the bedroom. At that time, he reports both legs gave away on him and he fell. He reported that he had some lightheadedness just before he fell and was slightly confused, but was aware of what was happening around him.

Report

HISTORY OF PRESENT ILLNESS: ,The patient is a 79-year-old right-handed man who reports that approximately one and a half years ago, he fell down while walking in the living room from the bedroom. At that time, he reports both legs gave away on him and he fell. He reported that he had some lightheadedness just before he fell and was slightly confused, but was aware of what was happening around him. He was able to get up shortly after falling and according to the patient and his son, subsequently returned back to normal.,He was then well until the 3rd of July 2008 when his legs again gave way on him. This was not preceded by lightheadedness. He was rushed to the hospital and was found to have pneumonia, and the fall was blamed on the pneumonia. He started using a walker from that time, prior to that he was able to walk approximately two miles per day. He again had a fall in August of 2008 after his legs gave way. Again, there was no lightheadedness associated with this. He was again found to have pneumonia and again was admitted to hospital after which he went to rehabilitation and was able to use his walker again after this. He did not, however, return to the pre-July baseline. In October of 2008, after another fall, he was found to have pneumonia again and shingles. He is currently in a Chronic Rehabilitation Unit. He cannot use a walker and uses a wheelchair for everything. He states that his hands have been numb, involving all the fingers of both hands for the past three weeks. He is also losing muscle bulk in his hands and has noticed some general weakness of his hands. He does, however, note that strength in his hands has not been normal since July 2008, but it is clearly getting worse. He has been aware of some fasciculations in his legs starting in August 2008, these are present both in the lower legs and the thighs. He does not report any cramps, problems with swallowing or problems with breathing. He reports that he has had constipation alternating with diarrhea, although there has been no loss of control of either his bowel or bladder. He has had some problems with blood pressure drops, and does feel presyncopal when he stands. He also reports that he has no feeling in his feet, and that his feet feel like sponges. This has been present for about nine months. He has also lost joint position sense in his feet for approximately nine months.,PAST MEDICAL HISTORY:,1. Pneumonia. He has had recurrent episodes of pneumonia, which started at approximately age 20. These have been treated repeatedly over the years, and on average he has tended to have an episode of pneumonia once every five years, although this has been far more frequent in the past year. He is usually treated with antibiotics and then discharged. There is no known history of bronchiectasis, inherited lung disease or another chronic pulmonary cause for the repeated pneumonia.,2. He has had a catheter placed for urinary retention, his urologist has told him that he thinks that this may be due to prostate enlargement. The patient does not have any history of diabetes and does not report any other medical problems. He has lost approximately 18 pounds in the past month.,3. He had an appendectomy in the 1940s.,4. He had an ankle resection in 1975.,SOCIAL HISTORY: ,The patient stopped smoking 27 years ago, he smoked approximately two packs a day with combined cigarettes and cigars. He has not smoked for the past 27 years. He hardly ever uses alcohol. He is currently retired.,FAMILY HISTORY: , There is no family history of neuropathy, pes cavus, foot deformities, or neuromuscular diseases. His aunt has a history of type II diabetes.,CURRENT MEDICATIONS: , Fludrocortisone 0.1 mg p.o. q.d., midodrine 5 mg p.o. q.i.d., Cymbalta 30 mg p.o. per day, Prilosec 20 mg p.o. per day, Lortab 10 mg p.o. per day, Amoxil 500 mg p.o. per day, vitamin B12 1000 mcg weekly, vitamin D 1000 units per day, Metamucil p.r.n., enteric-coated aspirin once a day, Colace 200 mg p.o. q.d., Senokot three tablets p.o. p.r.n., Reglan 10 mg p.o. q.6h., Xanax 0.25 mg p.o.

q.8h. p.r.n., Ambien 5 mg p.o. q.h.s. p.r.n. and Dilaudid 2 mg tablets p.o. q.3h. p.r.n., Protonix 40 mg per day, and Megace 400 mg per day.,ALLERGIES:, He has no medication or food allergies.,REVIEW OF SYSTEMS:, Please see the health questionnaire and clinical notes from today.,GENERAL PHYSICAL EXAMINATION:,VITAL SIGNS: BP was 137/60, P was 89, and his weight could not be measured because he was in a wheelchair. His pain score was 0.,APPEARANCE: No acute distress. He is pleasant and well-groomed.,HEENT: Atraumatic, normocephalic. No carotid bruits appreciated.,LUNGS: There were few coarse crackles in both lung bases.,CARDIOVASCULAR: Revealed a normal first and second heart sound, with no third or fourth heart sound and no murmurs. The pulse was regular and of normal volume.,ABDOMEN: Soft with no masses and normal bowel sounds. There were no carotid bruits.,EXTREMITIES: No contractures appreciated.,NEUROLOGICAL EXAM:,MSE: His orientation, language, calculations, 100-7 tests were all normal. There was atrophy and fasciculations in both the arms and legs.,CRANIAL NERVES: Cranial nerve examination was normal with the exception that there was some mild atrophy of his tongue and possible fasciculations. His palatal movement was normal and gag reflex was normal.,MOTOR: Strength was decreased in all muscle groups as follows: Deltoid 4/4, biceps 4+/4+, triceps 5/5, wrist extensors 4+/4+, finger extensors 4-/4-, finger flexors 4-/4-, interossei 4-/4-, hip flexors 4+/4+, hip extensors 4+/4+, knee extensors 4/4, and knee flexors 4/4. Foot dorsiflexion, plantar flexion, eversion, toe extension and toe flexion was all 0 to 1. There was atrophy in both hands and general atrophy of the lower limb muscles. The feet were both cold and showed dystrophic features. Fasciculations were present mainly in the hands. There was evidence of dysmetria and past pointing in the left hand.,REFLEXES: Reflexes were 0 in all sites in the arms and legs. The jaw reflex was 2+. Vibration was severely decreased at the elbow and wrist and was absent in the fingers. Vibration was absent in the toes and ankle bilaterally and was severely decreased at the knee. Joint position sense was absent in the toes and severely decreased in the fingers. Pin perception was absent in the feet and was decreased to the upper thighs. Pin was decreased or absent in the fingers and decreased above the elbows. The same distribution of sensory loss was found with monofilament testing.,COORDINATION: Coordination was barely normal in the right hand. Rapid alternating movements were decreased in the left hand greater than the right hand. The patient was unable to stand and therefore gait, Romberg's test and balance could not be assessed.,DIAGNOSTIC STUDIES: , Previous diagnostic studies and patient reports. There were extensive patient reports, all of which were reviewed. A previous x-ray study of the lateral chest performed in October 2008 showed poor inspiration with basilar atelectasis and an infiltrate. An x-ray of the cervical, thoracic and lumbar spine showed some evidence of lumbar spinal stenosis. A CTA of the neck with and without contrast performed in November 2008 showed minor stenosis in the left carotid, a mild hard and soft plaque in the right carotid with approximately 55% stenosis. The posterior circulation showed a slightly dominant right vertebral artery with no stenosis. There was no significant stenosis, but there was minor extracranial stenosis noted. An MRI of the brain with and without contrast performed in November 2008 showed no evidence of an acute infarct, major vascular occlusion, and no abnormal enhancement with gadolinium administration. There was also no significant sinusitis or mastoiditis. This was an essentially normal brain MRI. A CBC performed in January 2009 showed an elevated white cell count of 11.3, a low red cell count of 3.43, elevated MCH of 32.4 and the rest of the study was normal. An electrolyte study performed in January 2009 showed a sodium which was low at 127, a calcium which was low at 8.3, and a low protein of 5.2 and albumin of 3.1. The glucose was 86. TSH performed in January 2009 was 1.57, which is within the normal range. Vitamin B12 was greater than a 1000, which is normal and the folate was 18.2, which was normal. A myocardial stress study performed in December 2008 showed normal myocardial perfusion with Persantine Cardiolite SPECT. The ECG was non-diagnostic. There was normal regional wall motion of the left ventricle. The left ventricular ejection fraction was 68%, which is within the normal range for males. A CT of the lumbar spine without contrast performed in December 2008 showed a broad-based disc bulge at L1-L2, L2-L3, L3-L4 and L4-L5. At L5-S1, in addition to the broad-based disc bulge, there was also an osteophyte complex and evidence of flavum hypertrophy without canal stenosis. There was severe bilateral neural foraminal stenosis at L5-S1 and moderate neural foraminal stenosis at L1-L4. An echocardiogram was performed in November 2008 and showed mild left atrial enlargement, normal left ventricular systolic function, mild concentric left

ventricular hypertrophy, scleral degenerative changes in the aortic and mitral apparatus, mild mitral regurgitation, mild tricuspid regurgitation and mild to moderate aortic regurgitation.,DIAGNOSTIC IMPRESSION: ,The patient presents with a severe neuropathy with marked large fiber sensory as well as motor findings. He is diffusely weak as well as atrophic in all muscle groups both in his upper and lower extremities, although he is disproportionately weak in his lower extremities. His proprioceptive and vibratory loss is severe in both the distal upper and lower extremities, signifying that he either has a severe sensory neuropathy or has involvement of the dorsal root ganglia. According to the history, which was carefully checked, the initial onset of these symptoms goes back one and a half years, although there has only been significant progression in his condition since July 2008. As indicated below, further diagnostic studies including a detailed nerve conduction and EMG test today showed evidence of a severe sensory, motor, and axonal neuropathy and in addition there was evidence of a diffuse polyradiculopathy. There was no involvement of the tongue on EMG. The laboratory testing as indicated below failed to show a specific cause for the neuropathy. We are still, however, waiting for the paraneoplastic antibodies, which were send out lab to the Mayo Clinic. This type of very severe sensorimotor neuropathy with significant proprioceptive loss may be seen in several conditions including peripheral nerve vasculitis due to a variety of disorders such as SLE, Sjogren's, rheumatoid arthritis, and mixed connective tissue disease. In addition, it may also be seen with certain toxins, particularly chemotherapeutic agents. The patient did not receive any of these. It may also be seen as part of a paraneoplastic syndrome. Although the patient does not have any specific clinical symptoms of a cancer, it is noted that he has had an 18-pound weight loss in the past month and does have a remote history of smoking. We have requested that he obtain a CT of his chest, abdomen and pelvis while he is in Acute Rehabilitation. The verbal reports of these possibly did not show any evidence of a cancer. We did also request that he obtain a gallium scan to see if there was any evidence of an unsuspected neoplasm. The patient did undergo a nerve and muscle biopsy, this was a radial nerve and biceps muscle biopsy from the left arm. This showed evidence of severe axonal loss. There was no evidence of a vasculitis. The vessels did show some mild intimal changes that would be consistent with atherosclerosis. There were a few perivascular changes; however, there was no clear evidence of a necrotizing vasculitis even on multiple sections. The muscle biopsy showed severe muscle fiber atrophy, with evidence of fiber grouping. Again, there was no evidence of inflammation or vasculitis. Evaluation so far has also shown no evidence of an amyloid neuropathy, no evidence of a monoclonal gammopathy, of sarcoidosis, and again there is no past history of a significant toxin or infective cause for the neuropathy. Specifically, there is no history of HIV exposure. We would await the results of the gallium scan and of the paraneoplastic antibodies to see if these are helpful in making a diagnosis. At this point, because of the severity and the axonal nature of the neuropathy, there is no specific therapy that will reverse the course of the illness, unless we find a specific etiology that can be stopped or reversed. I have discussed these issues at length with the patient and with his son. We also addressed whether or not there might be a previously undiagnosed inherited neuropathy. I think this is unlikely given the short history and the rapid progression of the disorder.,There is also no family history that we can detect a neuropathy, and the patient does not have the typical phenotype for a chronic inherited neuropathy such as Charcot-Marie-Tooth disease type 2. However, since I have only seen the patient on one occasion and do not know what his previous examination showed two years ago, I cannot be certain that there may not have been the presence of a neuropathy preceding this.,PLAN:,1. Nerve conduction and EMG will be performed today. The results were indicated above.,2. The following laboratory studies were requested including electrolytes, CBC, thyroid function tests, B12, ANA, C-reactive protein, complement, cryoglobulins, double-stranded DNA antibodies, folate level, hemoglobin A1c, immunofixation electrophoresis, P-ANCA, C-ANCA, protein electrophoresis, rheumatoid factor, paraneoplastic antibody studies requested from the Mayo Clinic, B12. These studies showed minor changes, which included a low sodium level of 129 as previously noted, a low creatinine of 0.74, low calcium of 8.6, low total protein of 5.7. The B12 was greater than 2000. The immunoelectrophoresis, ANA, double-stranded DNA, ANCA, hemoglobin A1c, folate, cryoglobulins, complement, C-reactive protein were all normal or negative. The B12 level was greater than 2000. Liver function tests were normal. The glucose was 90. ESR was 10. Hemoglobin A1c was 5.5.,3. A left radial sensory and left biceps biopsy

were requested and have been performed and interpreted as indicated above.,4. CT of chest, abdomen and pelvis.,5. Whole body gallium scan for evidence of an underlying neoplasm.,6. The patient will go to the Rehabilitation Facility for Acute Rehabilitation and Training.,7. We have not made any changes to his medication. He does have some mild orthostatic changes; however, he is adequately controlled with midodrine at a dose of 2.5 mg three times a day as needed up to 5 mg four times a day. Usually, he uses a lower dose of 2.5 three times a day to 5 mg three times a day.,8. Followup will be as determined by the family.