NEUROLOGY CASE 4

Chief Complaint: A 30-year-old right-handed woman presents to your primary care clinic for one week of double vision.

History of Present Illness:
One week prior to presentation, the patient developed diplopia. During the course of one day, she noted mild blurry vision every time she looked to the left. This became more severe the next day. The day after that, she noted that instead of blurry vision, she now saw horizontal double images when she looked to the left. This occurs reproducibly every time she looks to the left and has not changed subsequently except for one day when she sat in a sauna and the double vision worsened noticeably with more distance between the images. This returned to baseline after several hours.

She denies previous diplopia, but she does recall an episode of blurry vision in her right eye a few years prior that gradually developed over several days. She is sure it was only in her right eye because she alternately covered each eye and the vision in her left eye was normal but the entire visual field of her right eye was blurry. She also had a dull discomfort behind the right eye, particularly when she moved the eye. All of this gradually resolved over several weeks.

For several months when she flexes her neck, she has noted an "electrical" sensation that quickly radiates down her back.

Past Medical History:
Multiple admissions for recurrent chest pain and palpitations with unrevealing evaluations. Multiple admissions for recurrent abdominal pain and nausea with unrevealing evaluations.

Medications: None

Allergies: No known drug allergies.

Social History: She works at a coffee shop but states that he will have to leave soon because working aggravates her symptoms. She is single and lives alone. She has never smoked, has about two drinks per week, and denies recreational drug use.

Family History: Negative.

Review of Systems: In addition to the above, she responds affirmatively when asked if she has shortness of breath, headaches, back pain, joint pains, muscle aches, pain with intercourse and menses, constipation, heartburn, dysuria, cough, fatigue, generalized weakness, insomnia, limb swelling and dizziness. She is vague on details and declines to elaborate further for any of these symptoms, stating "I've had all of this for years and none of the other doctors could figure out what was wrong with me, so why bother telling you?".

General Physical Examination: The vital signs and general examination are normal except for diffuse abdominal tenderness, without guarding or rebound, and diffuse tenderness with palpitation of most joints and muscles without deformity or signs of synovitis.

Mental Status: She is alert and oriented to person, place and time with normal attention to language and memory. Psychomotor activity is normal, affect alternates multiple times from bright to tearful when discussing her symptoms, mood is "upset". Thought process is normal except for perseveration about her symptoms. Thought content is normal. Insight and judgment are good.

Cranial Nerves: There is mild dysarthria. Visual fields and acuity are normal. There is a right afferent pupillary defect. Eye movements are full and conjugate in all directions except for left gaze where there is adduction weakness of the right eye, which does not cross the midline, and horizontal nystagmus of the left eye when full abducted. Facial sensation and contraction of the muscles of mastication are normal. Facial movement is normal. Hearing is normal. The palate elevates symmetrically. Shoulder shrug and head rotation are full strength. The tongue has normal bulk and movement.

Motor: Bulk and tone are normal throughout, and there are no fasciculations or involuntary movements. The neck is supple. Strength is 5/5 bilaterally in the tested proximal and distal muscles of all four limbs.

Somatosensation: Normal throughout to touch, pin, temperature and position sense. Romberg's sign is absent.

Coordination: There is mild ataxia of the left arm and leg with finger-to-nose and heel-to-shin testing as well as mild dysdiadochokinesia of the left arm and leg with rapid alternating movements.

Reflexes (right/left): Brachioradialis 3/2, biceps 3/2, triceps 3/2, patellar 3/2, Achilles 3/2. The plantar response is flexor on the left and extensor on the right.

Gait: Her base is wide and she is unsteady during stride. Heel and toe walking are normal, but she is unable to do tandem gait.

Subsequent Course:
You diagnosed somatization disorder based on her symptoms involving multiple systems without an apparent underlying medical condition, but you also recognized that some of her symptoms are due to a neurological disorder. You recognized right internuclear ophthalmoplegia as the cause of her current diplopia, and you recognized multiple other distinct central nervous systems lesions including previous optic neuritis, at least one likely cerebellar lesion, and a unilateral corticospinal tract lesion. As she has evidence of multiple focal central nervous system lesions of subacute onset which are "separated in time and space", as well as the presence of Lhermitte's symptom and Uhthoff's phenomenon, you diagnosed multiple sclerosis. This was confirmed by finding additional characteristic white matter lesions with magnetic resonance imaging of the brain and cervical spinal cord and the finding of oligoclonal bands in her cerebrospinal fluid. You prescribed an eye patch to relieve the diplopia until it resolves, corticosteroids to speed recovery, and an immunomodulatory drug to decrease the frequency of attacks.

Key Concepts:
Understand how to locate dysfunction of the nervous system based on symptoms and signs, and how to combine this with the syndrome time course and risk factors to determine the causal pathophysiology and diagnosis.

Learning Objectives:

1. Describe the anatomy of dysconjugate gaze deficits and abnormalities of the pupils and eyelids.
2. Describe and distinguish between the anatomy of monocular versus binocular visual loss.
3. Describe and distinguish between the anatomy of limb versus gait ataxia.
4. Define Lhermitte's symptom and Uhthoff's phenomenon and appreciate their significance.
5. List common causes of focal central nervous system dysfunction of subacute onset.
6. Describe somatization disorder and appreciate the difficulty in recognizing the development of a neurological disorder in patients with somatization disorder.

GUIDE TO NEUROLOGY CASE FOUR

Diagnosis:

1. Multiple sclerosis.
2. Somatization disorder.

Part One

Chief Complaint:

Diplopia may be caused by dysfunction of the extra-ocular muscles, the cranial nerves or nuclei of eye movements or their connections.

History of Present Illness:

When the eyes become dysconjugate by only a few degrees, the vision becomes blurry and when the dysconjugacy progresses by more than a few degrees, the images become doubled. Horizontal diplopia on lateral gaze is from either abduction weakness of the abducting eye or adduction weakness of the adducting eye.

Uhthoff's phenomenon refers to elevated body temperature transiently worsening the deficits of multiple sclerosis; the mechanism of this is unclear. The most common syndrome of multiple sclerosis is optic neuritis which is autoimmune demyelination of one optic nerve. This causes the subacute onset of monocular visual loss usually accompanied by retro-orbital pain, particularly with eye movement. Deficits of multiple sclerosis lesions usually have a subacute onset over days and resolve completely or incompletely over several weeks. Lhermitte's symptom is common with, but not specific to, multiple sclerosis where a lesion in the posterior columns of the cervical spinal cord causes an electric-like sensation that quickly radiates down the back with neck flexion.

Past Medical History and Review of Systems:

She has somatization disorder with multiple chronic complaints of unexplained physical symptoms despite appropriate evaluations including pain of multiple areas, gastrointestinal symptoms, and sexual or menstrual symptoms, all of which started earlier than 30 years of age.

Part Two

General Physical Examination and Mental Status:

Tenderness without objective abnormalities, labile affect, and perseveration about symptoms may be seen with somatization disorder.

Cranial Nerves:
A deficit of enunciation (dysarthria) may be seen with dysfunction of the muscles of speech, their cranial nerves or nuclei, or the upper motor neurons innervating these nuclei. Dysarthria may be caused by many disorders that cause focal dysfunction of these structures, such as multiple sclerosis in this case, or by many disorders that cause diffuse nervous system dysfunction, such as alcohol intoxication.

Any disorder of the eye, retina, or optic nerve that leads to less light information reaching the brain will decreased the direct (same eye) and consensual (other eye) pupillary light reflex when light is shown in the affected eye. Many patients with prior optic neuritis will continue to have the sign of afferent pupillary defect despite resolution of the symptom of monocular visual loss.

She has a lesion of the right medial longitudinal fasciculus in the central pons causing internuclear ophthalmoplegia. When she tries to look left, her right frontal eye fields project to the left pontine horizontal gaze center, which projects to the left abducens nucleus and nerve, causing the left eye to abduct, but the projection from the left pontine horizontal gaze center to the right oculomotor nucleus and nerve, to cause the right eye to abduct, is blocked by the lesion of the right medial longitudinal fasciculus. On left gaze, therefore, the left eye abducts but the right eye does not adduct. The horizontal nystagmus of the normal eye that occurs with internuclear ophthalmoplegia may be a reflexive compensation for diplopia.

Coordination:
Unilateral limb ataxia is usually caused by dysfunction of the ipsilateral cerebellar hemisphere or its connections and is usually accompanied by prominent tremor as the finger nears its target (intention tremor) on finger-to-nose testing and diminished ability to do rapid alternating movements (dysdiadochokinesia).

Reflexes:
The right-sided hyper-reflexia and extensor plantar response suggest dysfunction of the corticospinal tract which may be seen without weakness or spasticity with mild lesions.

Gait:
Gait ataxia is usually caused by dysfunction of the midline cerebellum. The narrowed base of the tandem gait improves the ability to detect subtle gait ataxia. It is typical to not be able to do tandem gait if there is obvious ataxia with casual gait.

Part Three

Subsequent Course:
Few disorders cause recurrent episodes of focal central nervous system dysfunction of subacute onset separated in time and space, and multiple sclerosis is by far the most common of these. Corticosteroids may speed recovery from an attack but also may not be needed in this case as the deficits are likely not disabling.