NEUROLOGY CASE ONE

Chief Complaint: An 80-year-old right-handed man presents to your primary care clinic for one day of difficulty typing and continued difficulty sleeping.

History of the Present Illness:
Yesterday evening, the patient noted difficulty typing an email to his daughter. He knew what he wanted to write, but he could not type some words, and when typing other words, he would type a similar but incorrect word, such as "truck" instead of "car". Some of the words he typed were nonsense. He saved the email as a draft and did not try typing again until this morning when he opened it. Reading the part he wrote yesterday, he could clearly see which words were wrong and he was able to edit some of them correctly. When he tried to fix other words, he only replaced the existing incorrect words with different incorrect words, at which point he decided to come in to see you.

He wrote an email to his son in the morning yesterday without difficulty. He has not been out of his house or communicated with anyone else since then.

He thinks the fingers of one of his hands might have been a little clumsy while typing, but he is not sure which hand it was.

The only other thing he noticed is that since yesterday, he has difficulty looking at anything on his right side and has to turn his head toward whatever he wants to look at to be able to look at it directly.

His chronic difficulty sleeping has been gradually worsening for several months. He is able to fall asleep easily but then wakes early in the morning and cannot get back to sleep. He is tired during the day and has low energy. Other symptoms have also developed in the last few months. He is eating less, which he attributes to not enjoying food as much as he used to. He used to look forward to his daily walk around town but has not felt up to this recently. He wonders if the difficulty typing is a sign telling him to stop bothering his children, as he feels that he is not much use to them anymore. He is concerned that he is becoming a burden to them.

Past Medical History: Hypertension, coronary artery disease and coronary artery bypass grafting 10 years prior.

Medications: Aspirin, atenolol and simvastatin.

Allergies: No known drug allergies.

Social History: He is a retired engineer. His wife died five years ago after 55 years of marriage. He has two adult children and several grandchildren that live far away, so he only sees them on holidays. He and his wife socialized with several other married couples when she was alive, but he has not kept in contact with them after her death. He smoked one pack of cigarettes daily for 50 years but quit 10 years ago. He denies drinking alcohol or using recreational drugs.

Family History: Negative.

Review of Systems: Otherwise negative.

General Physical Examination: The vital signs are normal. His weight is 160 pounds, down from 170 pounds at his last visit three months prior. There is an old midline chest incision scar and a left carotid bruit; the rest of the general examination is normal.

Mental Status: He is alert and oriented to person, place and time. He has normal attention and memory. There is a mild deficit of fluency with reduced total output and occasional paraphasias with both speaking and handwriting. Naming and repetition are mildly impaired. Verbal comprehension and reading are normal. He is aware of the fluency impairment and becomes frustrated when he cannot express himself properly. There is psychomotor slowing and restricted affect, his mood is "down". Thought process is bradyphrenic with increased speech latency, and ruminative about bothering his family with his problems. Thought content is without delusion or hallucinations. Insight and judgment are good.

Cranial Nerves: Visual fields and acuity are normal. Pupils are equal, round and reactive to light and accommodation. The eyes are conjugate but there is a mild left gaze preference and mild limitation of right gaze, which can be overcome with the oculocephalic maneuver. Facial sensation and contraction of the muscles of mastication are normal. Facial movement is normal except for flattening of the right nasolabial fold during smile. Hearing is normal for age. The palate elevates symmetrically. Shoulder shrug and head rotation are full strength. The tongue has normal bulk and movement.

Motor: Bulk is normal throughout, and there are no fasciculations or involuntary movements. There is mild resistance to passive movement of the joint of the right arm that increases with increasing velocity. The neck is supple. There is mild pronator drift and slowed fine finger movements of the right hand. 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. There is mildly diminished vibration sensation of the toes bilaterally. Romberg's sign is absent.

Coordination: Finger-to-nose, heel-to-shin and rapid alternating movement testing are normal.

Reflexes (right/left): Brachioradialis 3/2, biceps 3/2, triceps 3/2, patellar 2/2, Achilles 2/2. The plantar responses are flexor.

Gait: Normal base and stride with casual, heel, toe and tandem gait.

Subsequent Course:
You diagnosed the patient as likely having had a left frontal lobe ischemic stroke in the territory of the middle cerebral artery which was confirmed with a computed tomographic scan of the head. You also diagnosed a major depressive episode. You admitted him to the hospital for frequent neurological checks by nursing staff in case of deterioration from recurrent ischemic stroke, or edema or hemorrhagic transformation of the cerebral infarct. You had him take nothing by mouth until formal dysphagia screening could be performed because you know that the risk of aspiration pneumonia is high after stroke.

You suspected that the cause of his ischemic stroke was atheroembolism from a stenosing plaque at the left carotid bifurcation which was confirmed with ultrasound showing 90% stenosis of the origin of the left internal carotid artery. Cardiac rhythm monitoring overnight did not find paroxysms of atrial

fibrillation. You did not suspect cerebral subcortical small artery degenerative disease as the cause of his stroke syndrome because it was located in the territory of a large cerebral artery. You consulted surgery and carotid endarterectomy was performed the following day without complication. You prescribed physical, occupational and speech therapy, and he made an excellent recovery over the subsequent weeks, although he continues to make rare paraphasic errors.

He denied suicidal ideation when you specifically asked him about it during your initial interview, but over the subsequent weeks, his depression worsened despite having started on antidepressant medication after surgery and he was voluntarily admitted to the psychiatry service. His depression improved and he transitioned to continued outpatient care.

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 and distinguish between the anatomy of aphasia versus other abnormalities of communication such as dysarthria, dysphonia and confusion.
2. Describe and distinguish between the anatomy of conjugate gaze deficits versus dysconjugate gaze deficits.
3. Describe the anatomy of the upper motor neurons and distinguish upper from lower motor neuron signs as well as cortical versus subcortical motor syndromes.
4. Describe and distinguish the anatomy of central versus peripheral facial weakness.
5. List common causes of ischemic stroke and how they relate to the vascular risk factors.
6. List common secondary complications of stroke such as aspiration pneumonia, venous thromboembolism and pressure sores.
7. Appreciate that there is a short-term increased risk for recurrent ischemic stroke after ischemic stroke and transient ischemic attack.
8. Appreciate the increased risk of depression following stroke.

GUIDE TO NEUROLOGY CASE ONE:

Diagnosis:
1. Left frontal lobe ischemic stroke in the middle cerebral artery territory caused by atheroembolism from a stenosing plaque of the left carotid bifurcation.
2. Major depressive episode and disorder.

PART ONE

Chief Complaint:
Difficulty typing could be a motor problem of one or both hands or a cognitive problem such as aphasia or apraxia. Insomnia is common in the elderly with many potential causes, one of which is depression.

History of the Present Illness:
He is describing a mild expressive aphasia which he is noticing as difficulty typing because that is the only language he attempts to produce during this time. He has both word-finding difficulty and word errors (paraphasias) but no difficulty reading so his aphasia is expressive and likely secondary to dysfunction of Broca's area. Broca's area is on the inferior aspect of the lateral frontal cortex of the dominant hemisphere which is the left in most people.

Several things could cause clumsiness of one hand, but the most common would be unilateral dysfunction of the upper motor neurons in the corticospinal tract. Upper motor neuron lesions usually produce weakness, but mild lesions may only produce clumsiness of fine finger movements such as typing. The upper motor neuron cell bodies are in the primary motor cortex which is the posterior cortex of the frontal lobe on the precentral gyrus. Dysfunction of the ipsilateral cerebellar hemisphere could also produce hand clumsiness, but that is usually more obvious with reaching activities than fine finger movements.

He could be describing a gaze paresis where his eyes remain conjugate but cannot fully move in one direction. When this is horizontal, the lesion is usually in the contralateral frontal eye fields of the superior aspect of the lateral frontal cortex. Lesions of the extraocular muscles, cranial nerves, nuclei or their connections in the brainstem would usually cause a dysconjugate gaze producing diplopia. Lesions of the visual pathways produce visual loss.

His neurological symptoms can all occur with dysfunction of the left lateral frontal lobe which is in the territory of a single artery, the middle cerebral artery. His syndrome had a sudden or rapid onset during the course of one day. The term stroke refers to the syndrome of focal central nervous system deficits of sudden onset and presumed vascular cause. Focal cerebral ischemia is the more common cause of a stroke syndrome than intracranial hemorrhage, and he lacks headache which is usually seen with hemorrhagic stroke. He has the vascular risk factors of advanced age, hypertension, prior cigarette smoking and known atherosclerosis in his coronary arteries so he is, therefore, a person at increased risk for ischemic stroke.

He has more than five symptoms of depression for more than two weeks including dysphoria, anhedonia, feelings of worthlessness, guilt and changes in sleep and appetite. With one episode of major depression and no features pointing to another diagnosis, he appears to have major depressive disorder.

Past Medical History:
He has several common vascular risk factors which are associated with atherosclerosis and degenerative disease of the cerebral subcortical small arteries.

Medications:
He is taking common medications for coronary artery disease or atherosclerosis in any arterial bed. Aspirin prevents atherothrombosis, atenolol treats hypertension and simvastatin treats dyslipidemia.

Social History:
Social isolation is both a risk factor for developing depression and a risk factor for suicide from depression. Prior cigarette smoking remains a vascular risk factor because atherosclerotic progression was accelerated during the smoking period, although the risk is lower than with continued smoking.

PART TWO

General Physical Examination:
Unintentional weight loss has many potential causes including depression. The chest scar is from his coronary artery bypass graft surgery. The carotid bruit is likely from a stenosing atherosclerotic plaque at the bifurcation which, if confirmed, becomes the most likely cause of his syndrome of left middle cerebral artery ischemic stroke. Atheroembolism from the carotid bifurcation tends to go to either the ophthalmic artery or the middle cerebral artery which is a direct continuation of the internal carotid artery.

Mental Status:
He has an expressive aphasia with anomia (deficit naming), reduced amount of language production, paraphasias and a deficit of repetition, all of which is common with dysfunction of Broca's area. There appears to be no dysfunction of Wernicke's area because his comprehension is normal. Frustration is common with expressive aphasia. A key concept is that aphasia is distinct from other communication abnormalities such as dysarthria (deficit of enunciation), dysphonia (abnormal voice quality) and confusion (a global abnormality of cognition with or without aphasia as part of the syndrome). Aphasia is specifically a deficit of language production or comprehension which may be lost in isolation and is usually caused by dysfunction of the language cortices. He has many features suggestive of depression including dysphoric mood, restricted affect, psychomotor slowing, bradyphrenia, long speech latency and excessive guilt.

Cranial nerves:
His right horizontal conjugate gaze paresis is most likely from dysfunction of the contralateral frontal eye fields. This is often accompanied by a gaze preference (the eyes at rest are not in primary position) toward a destructive lesion such as a frontal lobe infarct. Overcoming this with oculocephalic maneuver confirms that the lesion is supranuclear rather than in the brainstem because the pathways are intact for the vestibular nuclei to project to the eye movement nuclei via the medial longitudinal fasciculus (the vestibule-ocular reflex). He has subtle right lower facial weakness which suggests a lesion of the corticobulbar tract to the facial nucleus. With central facial weakness, the upper face is spared because those lower motor neurons receive bilateral supranuclear innervation while those to the lower face receive only contralateral supranuclear innervation.

Motor:
Weakness can localize to several areas, but the upper motor neuron signs include spasticity (hypertonia that increases with increased velocity of passive joint movement), pronator drift, hyper-reflexia and the extensor plantar response. With mild lesions of the corticospinal tract, these features may be present without detectable weakness on manual muscle testing, and there may be slowed or clumsy fine finger movements which are primarily mediated by the corticospinal tract. Sensorimotor loss of the unilateral face and arm more than the leg is often seen with lesions of the lateral hemispheric cortex (most commonly from infarction in the territory of the contralateral middle cerebral artery), because the face and arm portions of the primary motor and somatosensory cortices are lateral while the leg portions are medial which is in the territory of the anterior cerebral artery.

Somatosensation:
A mild and symmetric loss of vibratory sensation in bilateral toes is a normal change of aging and may represent a mild degenerative polyneuropathy that is not part of a specific disorder. Romberg's sign, when present, indicates substantial proprioceptive loss of the legs.

Reflexes:
The right-sided hyper-reflexia suggests pathology of the corticospinal tract which, in this case, is likely at the level of the motor cortex because the other deficits localize to the neighboring cortical areas of the Broca's area and the frontal eye fields.

PART THREE

Subsequent Course:
There is a short-term increased risk of ischemic stroke after ischemic stroke or transient ischemic attack so admission for frequent neurological assessments is appropriate in addition to expediting evaluation of the cause of the ischemic stroke and to effective prevention of secondary complications. Many stroke patients are at increased risk of aspiration from dysphagia and should receive nothing by mouth until cleared by formal dysphagia screening. Venous thromboembolism and pressure sores are both common after stroke due to immobility. Most stroke patients improve, but the speed and completeness is highly variable. Depression is very common after stroke, even with minimal or no residual deficits, suggesting a biological effect of the brain injury. This likely explains his worsening, although it is possible he could have worsened without the stroke as well. He developed suicidality and was appropriately admitted for safety and treatment.