NEUROLOGY CASE 2

Chief Complaint: a 40-year-old right-handed woman presents to your primary care clinic for multiple types of "spells" occurring over different periods of time.

History of Present Illness:
Ever since she was a 20-year-old college student, the patient has had episodes of sudden onset of heart racing, chest pressure, shortness of breath, sweating, tingling of bilateral fingertips and lips, and a feeling of terror, as if she might die. These would last about 10 minutes and occur several times per year. Sometimes the spells would be triggered by a busy crowded place, but often they would occur without an obvious cause. She is always anxious about the possibility of these spells occurring and tends to avoid crowded places. She last had one of these events several months earlier.

Over the last month, she developed different episodes of a tingling sensation of the left side of her body. The tingling starts in her hand and face and then spreads up her arm, down her trunk and into her leg over about one minute before gradually fading away over the next minute. She is coming to see you today because she had another one of these spells yesterday that lasted several minutes and during which she also experienced a few seconds of rhythmic twitching of her left face and hand.

She has no other complaints, but when asked if she has headaches, she says yes. Since she was young, she has had moderate to severe headaches almost every month with menses that last hours to a full day, are unilateral and throbbing and improve if she lies down in a dark, quiet room.

Over the last two months, she has developed a continuous headache that was initially mild but has progressed to moderate intensity. It is holocephalic, worse when she wakes up, and worsens instead of improved if she lies down. It becomes severe if she coughs, strains or bends over.

When asked if she has other symptoms, she says no but mentions that a friend told her that she has seen the patient run into doorways with her left side multiple times. The patient has not noticed this occurring.

Past Medical History: None.

Medications: None.

Allergies: No known drug allergies.

Social History: She is an accountant and does all her work from home. She is not married and has no children. She has a few close friends and they tend to meet at each other's houses instead of public places. She never smoked, she drinks about two drinks a week, and she denies recreational drug use.

Family History: Negative.

Review of Systems: She describes herself as a nervous person who is excessively worried about something out of proportion to the situation most of the time which is often very bothersome to her. Otherwise negative.
General Physical Examination: The vital signs and general examination are normal.

Mental Status: She is alert and oriented to person, place and time with normal language and memory. Sustained attention is normal with subtraction of serial sevens. She will converse with the examiner on both her right and left sides, but there is extinction of visual and tactile stimuli on the left side with bilateral simultaneous stimuli. There is normal psychomotor activity. Affect is restricted, but her mood is "good". There is aprosodia. Thought process, content, insight and judgment appear normal.

Cranial Nerves: Visual fields and acuity are normal. Pupils are equal, round and reactive to light and accommodation. There is bilateral papilledema. Eye movements are normal and without nystagmus. Facial sensation is mildly diminished to all modalities on the left. Contraction of the muscles of mastication is normal. The face is symmetric at rest and with movement. 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, involuntary movements or pronator drift. The neck is supple. Strength is 5/5 bilaterally in the tested proximal and distal muscles of all four limbs.

Somatosensation: Mildly diminished throughout the left side to touch, pin, temperature, and position sense. Stereognosis and graphesthesia are normal on the right but diminished on the left. Romberg's sign is absent.

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

Reflexes (right/left): Brachioradialis 2/2, biceps 2/2, triceps 2/2, 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:
While it appeared that the patient has a history of migraines without aura and panic disorder, you were concerned that her new spells were simple focal seizures of the primary sensory cortex of the right parietal lobe with the last one also involving the adjacent primary motor cortex of the frontal lobe. You also considered migraine aura as a cause of these spells, but you felt that the associated rhythmic motor activity of the last spell made seizures more likely. These new focal seizures, combined with her new progressive headache, symptoms of intracranial hypertension, papilledema and symptoms and signs of focal brain dysfunction made you concerned that she may have a cerebral neoplasm in her right parietal lobe. This was confirmed on magnetic resonance imaging of the brain that you had her do the same day. The imaging finding of a single tumor in the white matter of the parietal lobe suggested a glioma, which was confirmed as a low-grade astrocytoma by the pathologist during her open biopsy that week.

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 abnormal Somatosensation and distinguish cortical from subcortical syndromes.
2. Describe the anatomy of hemineglect and extinction to bilateral simultaneous stimuli.
3. Recognize the symptoms and signs of seizures including amnesia, automatisms, spread and positive symptoms.
4. Describe and distinguish between seizures that are focal versus generalized and simple versus complex.
5. List common causes of seizures.
6. Describe and distinguish between migraine with and without aura.
7. Describe the symptoms, signs, time course and risk factors for intracranial hypertension.
8. Describe the symptoms, signs, time course and risk factors for intracranial neoplasia.
9. Contrast the syndrome of a panic attack with that of a seizure.

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Diagnosis:
1. Simple focal seizures, focal deficits of attention and somatosensation, and the syndrome of intracranial hypertension, all caused by a right parietal astrocytoma.
2. Migraine without aura.
3. Panic disorder.

Part One

Chief Complaint:
Neurologists use the noncommittal term spell for any episode of neurological symptoms or signs. A spell may represent many disorders including syncope, transient ischemic attack, migraine with aura and, in this case, seizures and panic attacks.

History of the Present Illness:
She has panic disorder with recurrent unexpected attacks that last minutes to hours of the sudden onset of at least four symptoms related to intense fear, and her attacks are followed by at least one month of persistent fear of having another attack. She has mild agoraphobia because of this. Numbness or tingling of the bilateral lips and fingertips often causes diagnostic confusion about spells of neurological versus psychological origin.

She has simple focal seizures of unilateral paresthesias. The positive aspect of the symptom and the phenomenon of spread of the symptom are both suggestive of seizure or migraine aura as opposed to a vascular cause. The last episode also included unilateral rhythmic twitching (a positive motor symptom) of her hand and face which indicates that the seizure spread from the sensory cortex to also involve the motor cortex. If the seizure had spread to involve bilateral cerebral cortex, it would be termed a focal seizure with secondary generalization, which would include an alteration in awareness, so that it would be complex instead of simple.

She has a typical description of migraine without aura in her past, but her new headaches are different and include multiple symptoms suggestive of intracranial hypertension. These include worsening with recumbency, coughing, straining and bending over, all of which increase intracranial pressure.

Repeatedly bumping into things with her left side without noticing is suggestive of hemineglect which is usually seen with right parietal cortical dysfunction.

Part Two

Mental Status:
Moderate or severe hemineglect is usually obvious during the interview or examination because responses to stimuli, such as questions, are diminished on the affected side. Mild hemineglect may only be detected by presenting simultaneous bilateral visual or tactile stimuli, to which the patient may not perceive the stimulus on the affected side, which is termed extinction. This cannot be tested unless there is some residual vision or somatosensation on that side to unilateral stimuli. Hemineglect is a deficit of attention on one side, almost always the left side from right parietal cortical dysfunction, and only rarely seen on the right side from the left parietal cortical dysfunction.

Many disorders cause a restriction of affect, such as depression, which should also be accompanied by dysphoric mood. Right parietal cortical dysfunction often causes a restriction of affect without depressed mood, as well as diminished ability to produce and perceive prosody (inflection, the emotional content of speech). Because of these changes, these patients often appear depressed but are not.

Cranial Nerves:
Bilateral papilledema suggests intracranial hypertension but has been present for at least several days.

Somatosensation:
Unilateral diminished perception of all primary somatosensory modalities suggests dysfunction of the contralateral brainstem or hemisphere, because pain and temperature ascend on the opposite side of the spinal cord from vibration and position sense. Diminished perception of objects (astereognosis) or letters/numbers (agraphesthesia) by touch alone suggests dysfunction of the sensory cortices in the parietal lobe instead of just the subcortical sensory tracts because this type of higher-level sensory processing occurs in the cortex.

Part Three

Subsequent Course:
New seizures in an adult without a clear cause, such as alcohol withdrawal, are always concerning for a potential new structural brain lesion such as a tumor. In this patient with new focal seizures, new focal persistent deficits, new progressive headache and other symptoms and signs of intracranial hypertension over a timeframe of weeks to months, the most likely diagnosis would be a cerebral neoplasm. In adults, cerebral metastases are more common than primary brain tumors, but here there is a single tumor in the hemispheric white matter and no symptoms or signs of systemic malignancy, so a primary brain tumor is likely, the most common of which is astrocytoma.