CHAPTER 25
Case History Problem Solving: Part III
Cerebral Cortex: Cortical Localization

These cases represent the gamut of disease affecting the cerebral cortex. The pathology represented may be tumor, infarction, or hemorrhage. Some of the lesions are intrinsic; others are extrinsic. The nature of the pathology may be uncertain; the location of the pathology, however, should be evident to you.

CASE 25-1A: Three months prior to admission, this 39-year-old, right-handed, white, male mechanic had the first of repetitive episodes characterized by "vertical wavy lines" in his left visual field. The initial episode lasted 30 minutes; subsequent episodes, 1 to 2 minutes. The patient had been seen by an ophthalmologist during the first of these episodes and a left visual field defect, which would disappear at the end of the episode, was detected. An electroencephalogram revealed no definite abnormality. Approximately 1 month later, the left visual field defect remained as a permanent deficit. At this time the patient also noted the onset of numbness in his left hand. Two months later the patient noted the onset of bitemporal and bifrontal headaches precipitated by any motion of the head. One week prior to admission there was an onset of vomiting and these symptoms prompted his admission.

Past History: Not remarkable.

General Physical Examination: Not remarkable.

NEUROLOGICAL EXAMINATION:

1. *Mental status:* This was intact except for some vagueness in the chronology for events in the present illness. The patient was oriented as to time, place and person. Delayed recall was intact. No aphasia was present; reading and writing were intact.
2. *Cranial nerves:*
   a. A complete left homonymous hemianopsia was present.
   b. Fundoscopic examination indicated bilateral papilledema with venous engorgement, arteriovenous nicking, and a recent hemorrhage in relation to the right disc. Visual acuity, however, was well preserved.
   c. Both pupils demonstrated a sluggish response to light.
   d. A left central (supranuclear) type of facial weakness was present with a droop of the left corner of the mouth.
3. *Motor system:* Strength was intact and cerebellar tests were negative. However the patient tended to lean slightly to the left when walking.
4. *Reflexes:*
   a. Deep tendon reflexes were asymmetrical, being slightly more active on the left than on the right.
   b. Plantar responses were flexor. Abdominal reflexes were present bilaterally.
   c. No release of grasp and suck reflexes had occurred.
5. *Sensory system:*
   a. Pain, touch, and vibration were intact.
   b. Position sense for fine amplitude movements at the fingers and toes was decreased on the left.
   c. On simultaneous tactile stimulation of the left and right side, extinction of stimuli occurred on the left.
   d. Errors were made in object identification in the left hand.
   e. Occasional errors were made in the identification of numbers drawn on the fingers of the left hand.

LABORATORY DATA:

The *electroencephalogram and imaging studies* now suggested a single lesion.

QUESTIONS:

1. Localize the initial symptoms? In your con-

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sideration as the location of the lesion, you will wish to consider the localizing significance of the following: the initial episodes of "vertical wavy lines" b. the homonymous hemianopsia. What is the nature of these initial symptoms?

2. Which areas were subsequently involved by the lesion? Take into account the pattern of findings on sensory examination.
3. What is the pathological nature of the lesion? Take into account the short duration of symptoms and the age of the patient.
4. Which diagnostic study would you perform?
5. How would you treat the seizures?
6. How would you manage the basic lesion?

CASE 25-2: This 35-year-old, right-handed, white male had the onset of a sudden pins-and-needles sensation which began in the left foot and then spread within a few seconds to involve the entire left side of the body including the arm and the face. The total episode lasted for 8 minutes. Similar episodes then recurred once or twice weekly beginning most frequently on the left side of the face or tongue and less often on the left foot or hand. Four months later the patient had a similar episode but fell to the floor with a loss of consciousness of several minutes duration during which observers reported some clonic movements of the left arm and leg. Neurological examination, lumbar puncture, and electroencephalogram at that time were reported as normal. The episodes were temporarily controlled with anticonvulsant medication, phenytoin (Dilantin), but then recurred with increasing frequency. At the time of hospital admission, 19 months after onset of symptoms, these episodes of numbness were occurring 3 to 4 times per day.

Past History: There was no history of significant head trauma. There was no family history of seizures.

NEUROLOGICAL EXAMINATION:

1. *Mental status:* Orientation and memory were normal.
2. *Cranial nerves:* No abnormalities were present.
3. *Motor system:* No relevant findings were present. The patient had fallen during the secondarily generalized seizure, sustaining a fracture of the left humerus and dislocation at the left shoulder. Since that time he had a poor swing of the left arm due to limitation of left shoulder movement. A slight atrophy of the hypothenar eminence with some weakness in the ulnar distribution had also been present since that injury.
4. *Reflexes:* Deep tendon reflexes were symmetrical; plantar responses were flexor.
5. *Sensory system:* All modalities were intact except for errors in graphesthesia in the left hand.

LABORATORY DATA:

1. *Skull x-rays:* The calcified pineal was shifted 4 to 5 mm compared to a shift of 2 mm. in films obtained 1 year previously.
2. *Electroencephalogram:* Focal slow wave activity of 4 to 6 Hz was present, consistent with focal damage.
3. *Cerebrospinal fluid:* Normal pressure and protein (21 mg./100 ml.) were present.
4. The *imaging studies* were consistent with a single focal lesion.

QUESTIONS:

1. Where is the lesion in terms of the origin of the seizure activity?
2. What are the possible pathological diagnoses? In answering this question you should keep in mind that the patient is of middle age (35), the seizures are clearly focal, and there is a 19-month history. These facts alone, even without the confirmatory laboratory data, should lead to the most likely diagnosis.
3. Which diagnostic study would you perform?
4. How would you treat the seizure?
5. How would you treat the basic lesion?

CASE 25-3: A 66-year-old, right-handed, white widow developed changes in affect and lack of interest in her surroundings over a 1-month period. Although previously she had been alert, caring for herself, doing her own shopping, and interested in her relatives, she now took to her bed and became disheveled in appearance. She would talk little if at all. She complained of a vague weakness in her legs, was unwilling to walk, but was able to walk to the bathroom without assistance.

Past History: A myocardial infarct had occurred 10 months previously. There was a 10-year history of Paget's disease involving the bones of the lower extremities.

NEUROLOGICAL EXAMINATION:

1. *Mental status:*
   a. The patient was apathetic with a dull appearance.
   b. The patient was disoriented for person and time. She knew that she was in a hospital in the state of Massachusetts but did not know the city.
   c. She was unable to recall her birth date or to name any president. She could give no account of her illness.
   d. She was unable to add 5 and 5 and could not do multiplication or subtractions.
   e. She repeated 4 digits forward and none in reverse.
   f. Spontaneous speech was scanty but without a definite dysarthria or aphasia. She could name 3-out-of-6 objects, could repeat 6-out-of-6, and could read aloud 4-out-of-6. She was able to perform slowly a two-step command. She was apparently unable to write.
2. *Cranial nerves:* II-XII were intact except that the left disc margin was blurred on funduscopic examination.
3. *Motor system:*
   a. No focal weakness was present.
   b. There was a variable resistance in both lower extremities described as "gegenhalten."
   c. The patient was very hesitant and very fearful of sitting, standing, or walking. There was some retropulsion (a tendency to fall backward when attempting to sit or walk). When she walked her right foot appeared to be glued to the floor.
4. *Reflexes:*
   a. Deep tendon reflexes were symmetrical and active.
   b. An equivocal plantar response was present on the right; that on the left was flexor.
   c. Strong grasp reflexes were released at hands and feet. Visual and tactile suck reflexes were also released.
5. *Sensory system:*
   a. Pain, touch, and position sense were intact.
   b. There was a minor decrease in vibratory sensation at the toes, which may well have related to the patient's general nutritional status.

LABORATORY DATA:

1. *Sedimentation rate* elevated to 98 mm. in one hour.
2. *Skull x-rays:* An osteolytic lesion (area of bone destruction) was present.
3. *Lumbar spine x-ray:* A suspicious, probably osteolytic lesion was seen in the body of the L1 vertebra.
4. *Stool Guaiac:* A small amount of occult blood was present. Hematocrit and hemoglobin were normal. Rectal and proctoscopic examination revealed a firm mass at approximately 9 cm. above the anus.
5. *Electroencephalogram* indicated a focal lesion having the quality of damage (continuous focal 2 to 4 Hz slow waves).
6. *Imaging studies* were consistent with a single focal lesion.
7. A *lumbar puncture* 3 weeks prior to admission had revealed pressure increased to 220

mm. of CSF and protein elevated to 59-mg./100 ml.

QUESTIONS:

1. Where is the lesion? (Assume a single central nervous system lesion).
2. What is the most likely pathology?
3. How would you manage this problem as regards more specific diagnosis and therapy? Which neuroimaging studies would you perform?

In considering these questions you will wish to keep in mind the following points:

1. As regards the location of the lesion:
   a. The personality change, accompanied by impairment of all areas of mental function,
   b. The release of strong grasp reflexes in both upper and lower extremities, in addition to suck reflexes,
   c. The apparent inconsistent findings as regards resistance to passive motion,
   d. The disturbance of sitting posture and of gait.
2. As regards the type of pathology:
   a. The elevation of sedimentation rate,
   b. The apparently rapid onset of symptoms,
   c. The presence of occult blood in the stool,
   d. The presence of osteolytic lesions,
   e. The mass noted on examination of the rectum.
3. Which diagnostic studies would you perform?

CASE 25-4: On the evening prior to admission, his wife noted that this 62-year-old right-handed, white, male civil engineer had the relatively sudden onset of slurred speech associated with a drooping of the left side of the face. The patient denied that these symptoms were present. The following morning he was examined by his brother, a physician, who confirmed the presence of slurred speech and weakness of the lower one-half of the left side of the face but also noted a weakness of the left arm and leg.

Past History: Gout had been present since age 20. He had sustained an apparent coronary artery occlusion in the past.

NEUROLOGICAL EXAMINATION:

1. *General:* blood pressure: 130/80; pulse 85 and regular.
2. *Mental status:*
   a. The patient was oriented for time, place, and person.
   b. Memory for recent and remote events was intact. Immediate and delayed recall was intact.
   c. The patient could do simple calculations but had difficulty with more complex problems.
   d. These results may have been influenced by the fact that the patient demonstrated a marked impersistence in following commands. The patient was somewhat apathetic and yawned frequently during the examination. There was a flattening of mood.
   e. The patient denied that he had any particular problems requiring hospitalization. He denied illness and stated he was as healthy now as he had been one week prior to hospitalization.
   f. Abstract reasoning was intact, as tested in proverb interpretation (used abstract concepts).
   g. No aphasia was present.
   h. A distinct deficit in the construction of three-dimensional drawings was present.
3. *Cranial nerves:* All were intact except for:
   a. A significant left facial weakness of supranuclear type.
   b. An inconstant neglect of left field on simultaneous stimuli to left and right visual fields.
4. *Motor system:* Strength and gait were intact and cerebellar tests were negative.
5. *Reflexes:* Deep tendon reflexes were symmetrical, plantar responses were flexor and no pathological grasp reflex was present.

6. *Sensory system*
   a. Pain and touch were intact.
   b. Stereognosis, two-point discrimination, and tactile localization were normal. However, position sense was decreased at toes and fingers on the left. There was a tendency to ignore tactile stimuli on the left face, arm, and leg when simultaneous left and right stimulation was carried out. Graphesthesia was intact in the right hand, but defective on the left.

LABORATORY DATA:

1. Blood *serological test* for syphilis was negative.
2. *Skull x-rays* were normal.
3. *Electroencephalogram*: abnormal because of focal 2-5 Hz slow waves.
4. *Brain scan (radioactive Hg 197):* A diffuse focal uptake was present.

SUBSEQUENT COURSE:

Over the next 3 months, significant improvement occurred. The patient was able to do complex calculations. He had no difficulty in drawing or in locating points on a map. Sensory examination was normal. The patient still had difficulty in maintaining his concentration. Psychological testing now indicated verbal IQ of 123, performance IQ of 104, with full scale IQ of 116, compared to much lower scores of 111, 87, and 101 respectively at the time of his acute illness.

QUESTIONS:

1. Where is the lesion (be specific)?
2. What is the nature of the lesion? If vascular discuss the type of lesion and the vascular territory
3. Which studies should be performed (in terms of neuroimaging and other studies)?

CASE 25-5: This 52-year-old housewife was referred for evaluation of episodic "twitching of the left thumb and forefinger" beginning 2 weeks prior to admission. The patient had noted the sudden onset of a numbness of the left thumb, followed in seconds by twitching of the left eyelid and then almost immediately by a twitching of the distal segment of the left thumb. This soon spread to the left forefinger, then to the middle finger, and then involved the hand in repetitive clonic movements. According to observers, the clonic movements then spread into the left arm. The twitching about the eyelid spread to the entire face. This initial episode lasted a total of 15 minutes; there was, however, a residual numbness of the left thumb and index finger: "as though they had fallen asleep." There was also a transient weakness of those areas first involved. Since the first episode, the patient had experienced minor recurrences of "tingling" of the left thumb and index finger.

Past History: Six years prior to admission the patient underwent a right radical mastectomy for carcinoma of the breast. Regional lymph nodes were reported as negative. The patient's mother and sister also had carcinoma of the breast.

NEUROLOGICAL EXAMINATION:

1. *Mental status:* intact with no evidence of aphasia. No deficits were noted in drawing.
2. *Cranial nerves:* A minimal left supranuclear central facial weakness was present.
3. *Motor system:* A mild weakness was present in the left upper extremity and rapid alternating movements of the left hand were slowly performed
4. *Reflexes:*
   a. Deep tendon stretch reflexes were increased on the left.
   b. An equivocal plantar response was present on the left; that on the right was flexor.
   c. No grasp reflex was present.
5. *Sensory System:*
   a. Pain, light touch, position, and vibration were normal.
   b. There was a slight disturbance in graphesthesia over the left hand and face. There was minor impairment of two-point discrimination over the left thumb and forefinger.

QUESTIONS:

1. Indicate the location of the lesion in this case.
2. Indicate the most likely pathology.
3. (a) Attach a label to the twitching which begin in the left thumb, then spread to the other fingers, then to left hand, arm, and face.
   (b) Attach a label to the transient weakness, which followed these episodes of twitching.
4. Which diagnostic studies would you perform?
5. Discuss management of this patient.

CASE 25-6: This 19-year-old, right-handed, white, male pharmacy student was referred for evaluation of frequent minor seizures. At age 18 months, the patient had fallen striking his head against a concrete floor and within hours had lapsed into coma. Later that day he required emergency neurosurgical treatment at the Children's Hospital for a "skull fracture and a bleeding vessel which the neurosurgeon had to tie." The patient apparently made an excellent recovery. However, from 3 years of age until 5 years of age he experienced "large convulsions" (apparently generalized convulsive seizures). During this time he also experienced minor episodes characterized by the sensation that objects were becoming smaller or larger. The patient recalled that at times, he had the sensation that bright geometric patterns were present and shrinking in size. Almost invariably the patient experienced the sensation of fear with these episodes. The patient had no seizures from age 5 years until age 16 years. Since age 16, the patient had frequent minor episodes of several types occurring as frequently as 15 to 30 times a day.

One type of episode was accompanied by dreamy sensations lasting a few seconds; at times things looked unreal. At the same time the patient would feel that the whole episode was somehow very familiar, that objects looked very familiar in a manner that could not be described. There was an accompanying sensation of fright. During such an episode the patient would be able to continue driving or walking.

A second type of episode, lasting 30 to 120 seconds, was characterized by an inability to talk properly, a babbling of speech, and an inability to understand speech. During this time a sensation of vague familiarity and fatigue and at times a sense of reincarnation would exist.

A third variety was characterized by the visual sensation of looking down a long dark corridor with a light at the other end. There was no concept of time: at times the corridor seemed to move from right to left. At the end of such an episode things would not look as they should; objects appeared distorted, and color altered. Objects were seen as black and white. The whole scene would seem unworldly. Sounds would be perceived but would fail to be registered and would not make sense.

NEUROLOGICAL EXAMINATION:

Mental status, cranial nerves, motor-system reflexes, and sensory system were all intact except for a minimal lag of the left side of the face in smiling. During the examination, the patient had a minor episode; his head dropped and he appeared out of contact for 20 to 30 seconds. He did not answer questions directly but replied by saying, "What was then, what was that then, what was that then?" He was confused for several minutes, after the episode. He was subsequently amnestic for the events of the episode.

QUESTIONS:

1. The electroencephalogram indicated a stable focus of discharge. Indicate the location of that focus.
2. Classify the seizure according to the International Classification of Seizures.
3. What is the most likely explanation for the acute events, which occurred at age 18 months?
4. Why did seizures recur at age 16 years?

CASE 25-7: This 20-year-old white male (Mr. K.B.) had been admitted to Boston City Hospital at age 9 months with acute right hemiplegia and acute right-sided hemi convulsions associated with a fever. A right hemiplegia was present for several days and he continued to have a mild right hemiparesis. Several additional seizure episodes occurred between ages 1-3 years. No additional episodes occurred until age 17 when the patient had a focal seizure characterized by tonic posture of the right arm – abducted at the shoulder and flexed at the elbow – followed by turning of the head and eyes to the right. Consciousness was impaired but not completely lost. For several minutes the patient was unable to speak. Upon regaining speech, he was able to follow commands.

Treatment with anticonvulsants (Eskabarb – long-release form of phenobarbital) was relatively effective in preventing seizure recurrence. Once a month the patient did experience a few seconds of "weakness or limpness" of the right hand with a possible small jerk of the right hand. Approximately two weeks before his initial office visit in January 1968, the patient discontinued his medications. Four days prior to this visit, at a time of sleep deprivation, the patient had a focal seizure with tonic movement of the right arm and deviation of the head and eyes to the right followed by a generalized convulsive seizure. Since age 9 months, residual weakness of the right arm had been present. However language functions had developed and he had done well in school

NEUROLOGICAL EXAMINATION:

1. *General:* The patient was left-handed with long-standing smallness of the right side of the face, arm, and leg.
2. *Mental status:* All areas were intact with no aphasia.
3. *Cranial nerves:* All were intact except for slight lag on the right in smiling.
4. *Motor system:*
   a. There was a mild weakness of the right upper extremity, most marked distally and particularly marked in the thumb abductor.
   b. Gait: A minimal circumduction of the right leg was noted.
5. *Reflexes:*
   a. Deep tendon reflexes were increased on the right.
   b. Plantar response was equivocal on the right.
6. *Sensory system:* Minor deficits were present in the right upper extremity characterized by errors in fine-movement perception of the right fingers and errors in graphesthesia of the right hand. Pain and touch were intact.

SUBSEQUENT COURSE:

With adjustment of anticonvulsant medication, excellent seizure control was attained. Only rare limited focal (partial) seizures occurred over the next 30 years.

QUESTIONS:

1. Locate the lesion.
2. What is the origin of focal (partial) seizures characterized by tonic posture of the right arm with deviation of the head and eyes to the right and with arrest of speech?
3. The patient has a right hemiplegia and yet he does not have aphasia. Explain.
4. In view of the acute onset of the hemiplegia at age 9 months and the subsequent lack of progression, speculate

Concerning the etiology.

5. Why was the weakness most prominent in the distal muscles of the upper extremity (particularly the thumb)?
6. Why did seizures recur?
7. Why has the smallness of the right side occurred?
8. What is the nature of the sensory deficit?
9. How would you manage the seizure problem?
10. Does the recurrence of seizures at age 17, after no seizures since age 3, indicate that a progressive pathology is present?

CASE 25-8: This 23-year-old, right-handed, white male was admitted with chief complaints of frequent and increasing headaches and "seizures". The problem apparently began 15 months prior to initial evaluation, when several months after their marriage, the patient's wife noted a change in his personality. In particular, the patient no longer had any desire for sexual intercourse. He became more irritable, at times with only minimal provocation and he was verbally and physically abusive.

Eight months later, (7 months prior to initial evaluation), he began to awake from sleep with numb feeling of right side of the lips which quickly spread to the right side of face; would last a few minutes and resolve. When the patient awoke during such episodes, he found himself sitting upright. These would occur weekly and only as the patient was going to sleep or after he had fallen asleep.

After a few weeks the seizures changed in character. While sleeping, the patient would suddenly be thrown toward his right side, his head would arch back and he would have difficulty breathing. He would be aware during these episodes and could understand what people said, but could not speak. Sometimes the patient would not be thrown to the side, but would simply have tonic stiffening and claw-like formation of the right hand, lasting 20 to 30 seconds.

Two months later, (5 months prior to evaluation) he underwent a right inguinal herniorrhaphy and had several attacks in the recovery room. Since that time, the patient has experienced one or two seizures per day, some of which occurred while awake.

Because of these episodes, he was admitted to a local hospital for evaluation. EEG and LP were reported to be normal. Following the lumbar puncture he developed a bifrontal headache. While sitting in a wheelchair, he began to complain of headaches. A nurse placed her hand on his forehead and his wife told the nurse to get her hand off. According to other observers, he ran down the corridor pushing the nurses aside and ran into a wall. He had to be restrained by several people, and was given some injections. Psychiatric treatment was begun with no significant improvement. Two months prior to evaluation, the patient's wife inquired of him whether the psychiatrist had been able to tell him anything about the cause of his episodes. He threw a jar of mustard at her, then the plates off of the dinner table. He rushed at her. As she retreated, he kept coming after her. He seemed unaware of his actions, and did not seem to recognize her. He placed his hands about her throat and began to choke her. With difficulty, she managed to break free. She noted that he appeared dazed and sleepy. At this point, fearing for her life, the wife decided upon a divorce. The patient denied any memory for this episode. Intermittent bitemporal and bifrontal headaches began to increase in frequency. Occasionally, he had incontinence of urine and tongue biting during his severe nocturnal seizures, which now were occurring, on a daily basis.

Fifteen days prior to admission, the patient became angry at a dog that was barking outside the house. He ran outside to chase the dog, but could not catch him and he kicked the fence gate. This is the last he remembers. His girlfriend relates that the patient began punching the brick wall of the house and then began to bang his head forcibly against the wall, stopped, and fell on his back staring, whereupon his girlfriend helped him walk into the house. The patient next recollects the trip to the hospital where lacerations of his hands were sutured and he was released.

NEUROLOGICAL EXAMINATION:

On admission evaluation, this was entirely normal. Bilateral paresthesias of the hands were reproduced by hyperventilation.

SUBSEQUENT COURSE:

8 months later, minor findings were present:

1. *Mental status:* He was anxious but affect was indifferent and passive.

2. *Cranial nerves:* A slight right central facial weakness was present
3. *Reflexes:* Deep tendon reflexes were slightly increased at right biceps, patellar and Achilles. The right plantar response was equivocal.

QUESTIONS

1. What is the most likely explanation and anatomical basis for the episodes of aggression in this case?
2. How often does directed aggression occur as an ictal event as opposed to non-directed aggressive behavior during ictal or post ictal confusional states?
3. From the anatomical standpoint what is the explanation for the change in personality?
4. What is the localization for the episodes of seizures while sleeping characterized by the following description "the patient would suddenly be thrown toward his right side, his head would arch back and he would note difficulty in breathing"?
5. What is the anatomical; localization for the episodes of tonic stiffening and claw-like formation of the right hand? What is the explanation for the initial episodes of tingling of the right side of face and lips?
6. How can you tie all of this together into a comprehensive diagnosis?
7. Discuss the possible underlying pathology and your diagnostic and therapeutic management.