CHAPTER 31
Case History Problem Solving: Part V
General

Case 31-1: This 59-year-old, white male, married plumbing contractor, approximately 7 years prior to admission, had the onset of a tremor of the right hand occurring mainly at rest. One year prior to admission, the patient had noted progression. The tremor was now present at the wrist and shoulder in addition to the fingers. During this time, the patient had also noted a stiffness of the right leg when walking. For one year prior to admission, since an alleged back injury, he had been unable to straighten up when walking and had had to walk in a very stooped position. During the 6 months prior to admission, the patient had experienced short episodes of subjective vertigo, resulting in a tendency to fall forward.

Past History:

1. Poliomyelitis at age 2, with residual atrophy of left gastrocnemius muscle.
2. Influenza at age 18.

NEUROLOGICAL EXAMINATION:

1. *Mental status:*

a. Patient was depressed and anxious.

b. There was mild impairment of digit span and of delayed recall, and an inability to retell stories. Some evidence of confabulation was present.

2. *Cranial nerves:* A fixed facies with minor facial asymmetry was present.
3. *Motor system:*

a. Strength was intact, except for plantar flexion at left ankle, with associated atrophy of gastrocnemius and shortening of the left Achilles tendon.

b. Gait: The patient walked stooped over, with slow short steps. He turned *en bloc.* There was a lack of associated movements of the arms, with greater impairment on the right side.

c. There was a resting tremor of the right upper extremity at the shoulder, wrist, and fingers. The tremor was rhythmical at 3 to 4/sec. and pill rolling in type.

d. There was a plastic resistance to passive motion with an intermittent cogwheel component in the right upper extremity at the shoulder, elbow, and wrist. To a lesser degree, rigidity was present in the right lower extremity.

e. Alternating movements were impaired bilaterally (right more so than left).

4. *Reflexes:*

a. Deep Tendon Reflexes: Biceps and triceps were more active on the left; patellar and Achilles were more active on the right.

b. Plantar responses were extensor bilaterally.

5. *Sensory system:* Intact.

LABORATORY DATA:

1. Thyroid function normal.
2. Cerebrospinal Fluid: Normal pressure, cells, protein, and Hinton.
3. EEG: Normal

QUESTIONS:

1. Indicate the diagnosis: provide a differential diagnosis.
2. Where is the lesion?
3. What significance do you attach to the bilateral Babinski signs?
4. What significance do you attach to the changes in mental status?
5. Indicate the pathophysiology and etiology.
6. Outline current concepts of diagnostic and therapeutic management.
7. Indicate the prognosis.

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Case 31-2: This 42 year-old woman reported to the doctor with a complaint of "trouble with my eyes". At around the age of 20, her menses ceased and never returned. The patient stated she had had an underactive thyroid for 20 years. Her menstrual symptoms did not respond to thyroid medication. During the past few years, the patient had had some automobile accidents and had noted that these accidents occurred when there were objects to her right. During the month before entry, the patient felt excessively tired and was sleeping more than usual but was able to go to work. Her skin had become dry; she had gradually put on weight over the years; her hair was scant with loss of underarm hair and scant pubic hair. A bifrontal dull headache had been present for one week.

PHYSICAL EXAMINATION:

There was puffiness of hands and face and dryness of the skin. Blood pressure was 108/72, pulse 82 and regular. Skin and mucous membranes were pale, and there was bilateral pallor of the optic discs.

NEUROLOGICAL EXAMINATION:

1. *Mental status:* Intact.
2. *Cranial nerves:* Intact except the visual fields showed a bitemporal upper quadrantic field defect that was almost complete in the right eye and less marked on the left. There was also bilateral pallor of the optic discs. Pupils and ocular movements were normal.
3. *Motor system:* No abnormalities noted.
4. *Reflexes:* Within normal limits.
5. *Sensory System:* Normal.

SUBSEQUENT COURSE:

Patient returned the next day, having had an emotional upset at work with the onset of a severe pressure headache. She complained of sudden worsening of vision in the left eye and a film over the temporal fields. On examination, she was pale and sweating. Blood pressure was 100/70; pulse was 100. There was more marked cutting of visual fields, particularly on the left, with a true hemianoptic defect (both eyes now demonstrated a full temporal hemianopia). The patient was drowsy but could provide concise answers. Neurologic examination was otherwise normal. The patient was immediately admitted to the hospital. Laboratory studies in the hospital confirmed the clinical impression of hypothyroidism. In addition, there was no FSH detected on an assay of a 24 -hour urine specimen. Plain skull films were positive and bilateral carotid arteriograms were performed. Patient was started on endocrine therapy, and an operation was performed. Three days following surgery, it was noted that the patient was complaining of constant and excessive thirst and was consuming huge quantities of fluids.

QUESTIONS:

1. In this case, neurological evaluation many years previously would have established the diagnosis. Indicate the location of this lesion. Discuss the differences between this lesion at onset and the lesion at the present time.
2. Indicate the most likely pathology. Speculate as to the other possible causes of this syndrome.
3. What happened the day after the initial office evaluation?
4. Today, which neurodiagnostic study would have best demonstrate the location and nature of the pathology?
5. This patient had 24-hour urine FSH levels performed. Which additional endocrine blood test would be performed today? What are the causes of abnormalities in that additional test?
6. What is the localizing significance of the thyroid dysfunction?
7. What is the localizing significance of the ovarian dysfunction?
8. Why did the patient have excessive thirst and excessive consumption of fluids (polydipsia) after surgery?
9. Why was the patient sleeping more than usual and why was she markedly drowsy at

the time of admission? (Several explanations are possible in this case.)

Case 31-3: Three weeks prior to admission, this 60 year-old right-handed white male had the acute onset of clumsiness of his right hand. Soon thereafter, he noticed weakness and numbness involving the right hand. He then noticed that his right leg was weak, and he fell to the floor. The weakness and numbness of the hand and leg cleared in a matter of minutes. The patient, however, continued to have clumsiness of the right hand. On two subsequent occasions, he had a recurrence of weakness of the right hand.

The patient's history was also of significance in that over a 2-month period, he had had 3 episodes, each 3 minutes in duration, of a monocular blindness of the left eye. Diabetes mellitus had been present for approximately 20 years. Six months before admission, the patient had had a sudden onset of weakness and coldness of the right lower extremity, due to sudden occlusion of the right femoral artery. Symptoms improved when a right femoral endarterectomy with bypass was performed. The patient had had 5 to 10 minute episodes of chest pain (angina pectoris) for a number of years.

Family history was significant in that the patient's mother, father, daughter and son had all experienced severe diabetes mellitus.

GENERAL PHYSICAL EXAMINATION:

1. Blood pressure was 130/80 in both the right and left arms.
2. Light pressure on the left eyeball led to a blackout of vision; similar symptoms were not produced on the right side.

NEUROLOGIC EXAMINATION:

1. *Mental status:* The patient was oriented for time, place, and person.
2. *Cranial nerves:* No significant findings.
3. *Motor system:* Strength was intact, except for a minimal drift downward of the outstretched right arm when both arms were outstretched. Cerebellar tests and gait were intact.
4. *Reflexes:* Deep tendon reflexes were slightly increased in the right arm compared to the left. The Achilles' tendon reflexes were absent bilaterally.
4. *Sensory System:*

a. Vibration sensation was decreased bilaterally at the toes.

b. Occasional errors were made in letter and number recognition in the right hand

QUESTIONS:

1. What is the diagnosis? Be specific as to the vessel and area involved, if vascular in nature.
2. Outline your diagnostic approach to this problem.
3. Indicate possible therapeutic measures.
4. What is the explanation for the absent Achilles' deep tendon reflexes and the decreased perception of vibration at toes?

Case 31-4: The patient was a 20 year-old, right-handed white housewife who was seen initially in the 23rd week of her third pregnancy. The patient presented with a history of generalized convulsive (tonic clonic) seizures beginning at age 7. These apparently would occur once a month without warning. She was begun on treatment with phenytoin and phenobarbital about age 8. She had had recurrences of seizures two years prior to the visit and one year prior to the visit. On each occasion, these seizures related to her omission of medication for several days. Her medications at the time of the initial visit were phenytoin, 100 mg a day and phenobarbital, 15 mg a day.

FAMILY HISTORY:

A maternal grandfather had convulsions and staring spells. Two cousins also had a seizure disorder, manifested by staring spells.

NEUROLOGICAL EXAMINATION:

In detail, the entire examination was normal.

LABORATORY DATA:

1. E*lectroencephalogram:* The patient had bilat-

eral, relatively synchronous bursts of poly spikes -slow waves complexes most prominent in the frontal recording areas. At times, the bursts would continue for 2 to 3 seconds. No clinical phenomena were noted during the discharges.

2. *Blood Levels:* Dilantin blood level was less than 2.5 uG/ml.; Phenobarbital was 5 uG/ml.

SUBSEQUENT COURSE:

The patient agreed to increase her intake of Dilantin to 200 mg a day and to increase her intake of Phenobarbital to 15 mgs. two times a day. The blood levels were checked again approximately 3.5 weeks later. Her phonation level was still low at 2.8 ug/ml., her phenobarbital was 4.5 ug/ml. The patient did not return again for follow up until 17 months later. In the interim, she had had no additional seizures of either a major or minor type. Her infant and previous offspring were normal.

1. **Classify these seizures. If possible be specific.**
2. **What other types of seizures might be expected in this syndrome?**
3. **What would be the most appropriate anticonvulsant?**
4. **What would neuro imaging studies demonstrate?**
5. **Discuss the relationship between epilepsy and pregnancy.**
6. **Discuss the relationship between anticonvulsants and pregnancy.**
7. **Discuss the role of anticonvulsants and related drugs in teratogenesis.**
8. **Discuss the interaction of anticonvulsants and birth control pills.**

Case 31-5: This 23 year-old white right-handed, army recruit private, on the day prior to admission, was evaluated on sick call for an apparent upper respiratory infection. The patient was given a day of barracks rest. Early on the morning of admission, the patient apparently vomited. At reveille, the patient was found in his bed, unresponsive and he was found to have been incontinent of urine and feces.

GENERAL PHYSICAL EXAMINATION:

1. The patient was febrile with a temperature of 102 to 103∞F. Blood pressure was 130/70; pulse was 96.
2. There was a marked degree of nuchal rigidity.
3. Examination of the skin revealed a diffuse petechial rash with a number of ecchymotic areas on the extremities. Some of these areas had apparent necrotic centers.

NEUROLOGICAL EXAMINATION:

1. *Mental status:* The patient was in a coma; he responded to painful stimuli by withdrawing his extremities.

2. *Cranial nerves:*

a. The pupils were midposition and responded very poorly to light.

b. Funduscopic examination was not remarkable.

3. *Motor system* There was no definite lateralized weakness. Cerebellar system and gait could not be tested.

4. *Reflexes:*

a. Deep tendon reflexes were equal but hyperactive throughout.

b. An extensor plantar response was present on the left side.

5. *Sensory System:* Could not be adequately tested.

QUESTIONS:

1. What is your diagnosis?
2. Which studies must be performed and when must they be performed?
3. When must treatment be started? Which treatment would be most appropriate?

LABORATORY DATA:

1. *CBC:* White blood count was markedly elevated to 42,000, with a differential count of

64% neutrophils, 31% bands, and 5% lymphocytes. Platelets were decreased to 95,000 per mm3. Blood serology was negative.

2. *Spinal fluid examination* revealed a marked increase in pressure (>600mm of CSF) The fluid was grossly cloudy. There were 7250 white blood cells per cubic mm; 100% of these were polymorphonuclear leukocytes. Spinal fluid sugar was 6-mg/100 ml (blood sugar was within normal range); spinal fluid protein was increased to 314-mg/100 ml.
4. *Smears and eventually cultures* of spinal fluid and blood were consistent with the clinical diagnosis.

SUBSEQUENT COURSE:

The patient was begun on specific therapy. As the patient's level of consciousness began to improve, on the second hospital day it was apparent that a left facial weakness was present. On the fourth hospital day, as the patient began to respond to questions by opening his eyes and moving his right hand, it became apparent that he had a left facial weakness, a weakness of the left arm and leg, and a left Babinski response. On the sixth hospital day, even though the patient was continuing to improve, he had the onset of focal seizures beginning on the left side of the body. Some remained localized to the left side; some became generalized. Following readjustment of therapy, seizures disappeared. With physiotherapy, a significant improvement in function of the left arm and left leg occurred.

QUESTIONS:

4. Do you wish to modify your diagnosis and proposed treatment based on the CSF findings? What do you expect the cultures of blood and CSF to indicate?
5. Indicate the pathophysiology involved in the complications that developed during the patient's hospitalization.

Case 31-6: This 62 year-old white divorcee was referred for evaluation of memory subsequently of personality change. The patient had a high school education and had been described in the past as a bright, alert and intelligent lady. The patient was unable to provide a history. She showed little insight into her problem and seemed unaware of any reason for her neurological evaluation. The patient's son indicated that she had begun to show some decline in intellectual capacity as long as 3 or 4 years prior to admission. During the preceding 12 months, a more marked change had occurred, particularly with regard to memory. In more recent months, a lack of concern for her personal appearance and a lack of spontaneity had developed. She had lost interest in her home, friends, and activities.

Past history was unremarkable as regards cardiovascular disease, diabetes or alcohol or drug and ingestion.

GENERAL PHYSICAL EXAMINATION:

Blood pressure was 140/96; cardiac status was normal.

NEUROLOGICAL EXAMINATION:

1. *Mental status:*

a. The patient was pleasant and well mannered.

b. She was disoriented for the day of the week, for the day of the month, for the month and for the year.

c. She was unable to remember any of four test objects after a 5-minute period.

d. The patient was unable to remember the year she was born.

e. She was unable to recall any president but Kennedy.

f. She was able to follow very simple directions, but was unable to remember any instructions beyond these.

g. She was unable to comprehend subtraction of serial 7s and could not do even simple additions. She was unable to repeat numbers in reverse.

h. She was unable to draw a triangle but did recognize a circle.

i. There was a severe dysnomia for even

common objects.

2. *Cranial nerves:*

a. Pupillary reactions were normal.

b. At times, the patient had extinction of simultaneously presented visual objects in the left or right visual field.

3. *Motor system:*

a. Strength and gait were intact.

b. Minor tremulousness was present in the performance of hand and arm movements.

c. Minor variable resistance on passive motion existed and was described as Gegenhalten.

4. *Reflexes:*

a. Deep tendon reflexes were symmetric and physiologic.

b. Plantar responses were flexor bilaterally.

c. Grasp was absent.

5. *Sensory system:* Normal.

QUESTIONS:

1. Does this patient have focal disease or a more generalized disease? If the latter, which areas are predominantly affected? What is the significance of the dysnomia in this case? Which diagnosis would you assign to this case?
2. Describe the most likely pathology in this case. Present a differential diagnosis.
3. What treatable causes of this progressive syndrome much be considered? Discuss in terms of:

a. Infections

b. Nutritional diseases

c. Metabolic diseases

d. Intoxications

e. Other treatable etiologies For each entity indicate the appropriate diagnostic tests.

4. How would you manage this patient as regards diagnosis and treatment?
5. What would an electroencephalogram demonstrate?
6. What would a CT scan demonstrate?
7. What cerebrospinal fluid findings are to be expected?
8. What is the natural history and prognosis?
9. What more restricted mental status changes might have been present early in the disease course? What would be a more likely diagnosis if personality changes had been the primary early symptoms? If basal ganglia symptoms had been present early in the course of the disease, which diagnoses might have been considered.

Case 31-7 (Patient of Doctor John Sullivan and Doctor Huntington Porter): This 67-year-old white right-handed retried schoolteacher awoke one morning with numbness over the entire right side of her body. The right corner of her mouth drooped. She was able to go to a family physician but was unsteady on her feet. She had no diplopia or aphasia. Numbness on the right side disappeared gradually over a two-week period, but then appeared in the left foot. The left hand became restless and would more in a jerky up-and-down movement. The hand and arm would take on abnormal postures and tend to trail behind her. One week later, this involuntary movement began to affect the left foot, which became restless. Its constant motion seriously interfered with her ability to walk. Over the next several weeks, the movement disorder in the left hand disappeared.

Past History was not remarkable except for menopause at age 30. Family history indicated that her mother died at age 63 of a "heart attack". Father died at age 72 of a "shock".

GENERAL PHYSICAL EXAMINATION:

Blood pressure was 160/100, with a normal cardiac examination.

NEUROLOGICAL EXAMINATION:

1. *Mental Status:* All areas were intact, with no evidence of aphasia.

2. *Cranial Nerves:*

a. A slight droop of the left corner of the mouth was present.

b. There was an unsustained nystagmus on far lateral gaze.

3. *Motor System:*

a. Strength was intact, with good retention of skilled movements of the hands.

b. In the left leg, there was an almost constant play of movement, smooth and rhythmical, consisting of eversion/inversion of foot and ankle with flexion/extension movements of the toes. In the left arm, the same type of movement was present. This movement was not synchronized with that in the foot and was often absent for periods of time. As the patient used the right arm, the movement disorder became apparent in the left hand. When the patient performed repetitive movements of the left fingers, mirrored movements of the right fingers occurred.

c. Gait: The body was twisted to the left. The left hand trailed behind, with the index finger pointing down and the other fingers clenched. The gait was unsteady. Walking did not dampen the movement in the left leg. At times, the left leg would suddenly buckle.

4. *Reflexes:*

a. Deep tendon reflexes were symmetric and physiologic.

b. Plantar responses were flexor bilaterally.

c. There was no release of the grasp reflex and no repellent apraxia.

5. *Sensory System:* Pain, touch, position, and other discriminative modalities were intact. There was a slight decrease in vibration sensation at the toes.

LABORATORY DATA:

1. *Fasting blood sugar* was elevated to 250-mg/100 ml.
2. *Cerebrospinal fluid:* Opening pressure was 60; no cells; protein was 45-mg/100 ml.
3. *Chest and skull X-rays* were negative, except for calcification of the carotid siphon.
4. *Sedimentation rate* was 17 mm in one hour.

SUBSEQUENT COURSE:

The patient did well for approximately one month. Then she began to have sudden "jerky movements" of the left hand, causing the arm to strike her in the face. The left arm also began to posture in a position of relative flexion at the elbow, wrist, and fingers. The patient also reported occasional violent flinging movements of the arm. To some extent, numbness had returned to the right upper extremity.

When she was re-examined 4 months after the initial episode, certain changes had occurred in the movement disorder. A distal fluid, flowing 2 to 4 Hz movement was still noted at the fingers, toes, hand, and foot, alternating between flexion/supination and extension/pronation. This movement had now appeared at more proximal joints, and some spread of movement into the face was reported. In addition, there were sudden pronation and posterior rotatory movements at the shoulder. At times, this was described as a wild swinging or flinging movement of the left upper extremity. At the time of a visit 5 months later, significant improvement had begun to occur. By the time of a follow-up 2 months later, approximately one year after onset of symptoms, there was little evidence of any movement disorder. Occasional distal movements occurred only when the patient was emotionally excited.

QUESTIONS:

1. Provide a diagnostic label for the abnormality of movement noted on the initial neurologic examination.
2. Provide a diagnostic label for the movement disorder that subsequently evolved.
3. Discuss the neuroanatomic basis of these disorders.
4. Discuss diagnostic and management approaches.

Case 31-8: One week prior to admission, this 51-year-old white housewife, developed episodic posterior headaches beginning in the neck and radiating to the vertex of the head. The headaches were usually related to and were definitely exacerbated by straining or

coughing. At the same time, the patient noted clumsiness of the left hand, and a tendency to fall to the left side. During the week prior to admission, a progressive gait ataxia also had developed.

PAST HISTORY:

Eight months previously, the patient had been admitted to the gynecology service before with a 10-week history of postmenopausal vaginal bleeding. She was found to have an infiltrative anaplastic carcinoma of the uterine cervix. The patient was treated with radiotherapy (4000R to the pelvis), followed by vaginal insertion of radium. Re-evaluation one month prior to the present admission had revealed that this anaplastic epidermoid carcinoma of the cervix had spread to the anterior vaginal wall. In addition, metastatic nodules were now present in the lung.

NEUROLOGIC EXAMINATION:

1. *Mental status:* The patient was an anxious white female who was cooperative, alert and well oriented.

2. *Cranial nerves:*

a. There was coarse nystagmus on gaze to the left.

b. There was a minor dysarthria for lingual and guttural sounds.

3. *Motor system:*

a. Strength was intact.

b. Gait was unsteady, with tendency to fall to the left, especially when turning to the left. On a narrow base, the patient tended to fall backwards.

c. Cerebellar tests revealed no definite truncal ataxia. There was a marked Appendicular ataxia on the finger-to-nose test in the left upper extremity and the heel-to-shin test in the left lower extremity.

4. *Reflexes:*

a. Deep tendon reflexes were active bilaterally. This, however, was felt to be consistent with the patient's degree of anxiety.

b. Plantar responses were flexor.

5. *Sensory system:* All modalities were intact, except for a minimal decrease in vibratory sensation at the toes.

LABORATORY DATA:

The results of specialized studies were consistent with the clinical diagnosis.

QUESTIONS:

1. Where is the lesion? Be specific.
2. What pathological process is to be expected?
3. Outline your diagnostic and therapeutic approach for this case.
4. Which is the appropriate neuroimaging study?
5. Should a lumbar puncture be performed? Indicate why or why not.

Case 31-9: A 21-year-old white right-handed wife of an air force enlisted man had been married for two months and had just moved from her home in the south to an air force base in New Jersey. She had been complaining of increasingly severe right frontal temporal or bifrontal headaches for three to four weeks. During the 10 days prior to admission, she had been treated on several occasions at the base dispensary for these headaches, which were relatively constant and were now accompanied by blurring of vision and frequent vomiting. Various medications had been prescribed for an upper respiratory infection or sinusitis. In the 48 hours prior to admission, the patient had become increasingly confused and ataxic. Her past history was otherwise negative as regards head injury or pulmonary and ear infection.

GENERAL PHYSICAL EXAMINATION:

1. The patient was dehydrated and somewhat lethargic.
2. She was complaining of severe headache, particularly on head movement.
3. There was a moderate degree of hyperventilation.
4. No otitis media was present.

NEUROLOGICAL EXAMINATION:

1. *Mental status:* The patient was disoriented for time and place. She was unable to provide a history and was unable to cooperate for the examination. She was impersistent in fixing gaze or in maintaining eyes in open position.
2. *Cranial nerves:*

a. On funduscopy, bilateral papilledema was present, with 2 to 3 diopters of disc elevation, accompanied by fresh hemorrhages and venous engorgement.

b. The right pupil was dilated and fixed in response to light. Extraocular movements were otherwise intact.

c. A left central facial weakness was present to a marked degree.

3. *Motor system:*

a. All limbs were moved spontaneously, but there was a downward drift of the outstretched left arm.

b. The patient was ataxic in a sitting position and in attempting to stand, even on a broad base.

c. There was a tremor of the outstretched hands, increasing to a minor degree on movement and intention.

4. *Reflexes:*

a. Deep tendon reflexes were increased on the left in the upper and lower extremities.

b. Plantar response was extensor on the left, equivocal on the right.

c. Grasp reflex was present bilaterally.

5. *Sensory system:* Pain sensation was intact; modalities could not be tested.
6. *Neck:* Minor resistance to passive motion.

LABORATORY DATA:

1. *Skull and chest X-rays* were negative.
3. *EEG* demonstrated almost continuous focal 1 to 2 Hz slow-wave activity.

QUESTIONS:

1. This patient was seen relatively late in her disease course. You should be able to localize the primary location of the pathology.
2. Indicate what secondary complications have occurred.
3. The nature of the pathology may be somewhat uncertain to you. However, you should be able to present a differential diagnosis and then to indicate the most likely diagnosis. Keep in mind that the headaches had been present for only 3-4 weeks and the additional symptoms for only 2-10 days.
4. Was the papilledema long-standing?
5. What is the significance of the pupillary findings?
6. Assuming a cerebral hemisphere lesion, why was the patient ataxic in sitting and standing when examined late in her course?
7. Which diagnostic studies would you perform in this case, and when would you perform these?
8. Which diagnostic studies would you not perform? State reasons.
9. How would you manage this problem from a therapeutic standpoint? Indicate when you would institute this therapy.

Case 31-10: This 55-year-old white widow, 6 to 8 years prior to admission, had the onset of a peculiar reeling gait. At the same time, she was noted to be "very nervous", having many peculiar restless movements. At other times, she was described as constantly fidgeting. The patient's children felt that in recent years, she had not been thinking as clearly as she once did. A personality change had also occurred.

FAMILY HISTORY:

The patient's mother died in her sixties with a disorder that had been labeled as Parkinsonism. From the description of relatives, however, it was evident that the mother had restless movements that were similar to the patient's.

NEUROLOGICAL EXAMINATION

1. *Mental status:* The patient was oriented and alert with an intact general store of information. Object recall was three out of four in 10 minutes. Digit span was six forward and

three in reverse

2. *Cranial nerves:* Intact.

3. *Motor system:*

a. Strength was intact with no significant spasticity or rigidity.

b. Gait showed a swooping quality of variable degree; there was also a decrease in associated arm movements.

c. As the patient sat, she would move her head, neck, and feet almost constantly .

d. The patient was able to walk a tandem gait, and there were no abnormalities of cerebellar function.

4. *Reflexes:* Deep tendon reflexes were symmetric. Plantar responses were flexor. No grasp reflex was present.

5. *Sensory system:* Intact.

LABORATORY DATA:

1. Routine laboratory studies were not remarkable.

2. *Specialized neuroradiology studies* were consistent with the clinical diagnosis.

QUESTIONS:

1. Present a differential diagnosis of this problem and indicate the most likely diagnosis.

2. Outline your diagnostic and therapeutic approach to this problem.

3. What did the specialized neuroradiology studies reveal?

4. Which neurodiagnostic study would be most cost-effective and specific in establishing the diagnosis today?

5. Discuss the underlying molecular basis of this disorder.

Case 31-11 (patient of Dr.Thomas Sabin): A 50-year-old white man was admitted to the hospital because of drowsiness and confusion of 5 days duration. His wife stated that he was in good health until that time. For 4 days prior to admission, he merely slept more than usual and napped during the day, but while awake, was lucid. The day before admission, he was confused and this confusion increased on the day of admission.

NEUROLOGIC EXAMINATION:

This was a thin man with normal vital signs who answered questions quickly but often incorrectly. In addition to the confusion, drowsiness and problems in memory, pertinent findings included the following: Extraocular movements were limited in all directions, and there was horizontal and vertical nystagmus. The gag reflex was diminished bilaterally. Finger-to-nose and heel-to-shin tests showed a mild deficit. The patient had an unsteady, broad-based gait. No other motor findings were present

QUESTIONS:

1. Do you require more historical information? If so, indicate what is required.

2. Present a differential diagnosis.

3. What is the most likely diagnosis?

4. In a general sense what is the localization of lesions in this disease. What is the localization for each specific symptom or sign (confusion and impairment of new learning, lethargy, impairment of extraocular movement and ataxia?

6. How could your suspected diagnosis be confirmed?

7. How and when would you treat this patient? Which findings would rapidly resolve over the next 1-2 days?

8. You administer specific treatment but the difficulties with memory fail to clear and one year later the patient is still confused and confabulating. How do you modify your diagnosis?

9. You administer specific treatment but one year later, the patient is still ataxic. How do you modify your diagnosis?

Case 31-12: This 60-year-old right-handed retired priest was referred for evaluation of problems in walking. The history was not precise. Two and a half years previously, he had been admitted to his local hospital, for severe

diabetic coma. Although he recovered, he had some difficulty with memory thereafter. He began to fall. Problems in walking had progressed, particularly in the last 6 to 8 months, so that he was no longer able to walk without assistance. His memory problems had progressed over the last year. He denied numbness in his extremities but was aware of "twitching" of his extremities. He denied any significant family history. He indicated he had been a very heavy drinker and had been hospitalized once in the previous year for treatment of alcoholism. Admitted intake was at least half a liter (500 cc) of gin per day.

NEUROLOGIC EXAMINATION:

1. *Mental status:* He was very vague about the date. Delayed recall was limited to two out of five objects in five minutes.
2. *Cranial nerves:* All were intact, except for a tremulous voice.
3. *Motor system:*

a. The patient stood on a broad base and was very unsteady standing on a narrow base, requiring support. This unsteadiness slightly worsened with eye closure.

b. He had been brought into the office in a wheelchair. He walked with assistance with small steps but was very ataxic.

c. There was no impairment on finger-to-nose testing but there was dysmetria on heel-to-shin test.

4. *Reflexes:*

a. Deep tendon reflexes were everywhere absent.

b. Plantar responses were not responsive.

5. *Sensory system:*

a. Pain sensation was absent to the level of the knees.

b. Position sense was intact.

c. Vibration sensation was markedly decreased at the toes and moderately decreased at the ankles.

LABORATORY DATA:

1. *Blood studies:* Folic acid, B12 levels, serum and urine immuno-electrophoresis, ESR, and ANA were normal. Serological tests (RPR and FTA) were negative. Liver function studies had been previously elevated but now, except for alkaline phosphatase, were normal.
2. *Electromyogram and nerve conduction* studies indicated a severe sensory-motor peripheral neuropathy, primarily axonal in type, as well as left median nerve (carpal tunnel) and right ulnar nerve (olecranon groove) entrapment neuropathies.
6. *MRI* demonstrated:

a. Prominence of cerebellar folia, particularly of the superior vermis;

b. Moderate cerebral cortical atrophy;

c. Periventricular white matter involvement (leukoariosis).

SUBSEQUENT COURSE:

An alcoholism program with discontinuation of all alcohol intake and administration of multiple B vitamins was recommended. When last seen in follow-up, the patient was consuming at least 500 cc of vodka per day. His neurologic examination was unchanged except that pain sensation was now intact. He refused additional neurologic followup.

QUESTIONS:

This patient had several neurologic problems: At least five can be specified.

Discuss in terms of:

a. Ataxia

b. Memory changes

c. Peripheral nerve disorders

d. Toxic and metabolic diseases