

CHAPTER 14

Problem Solving II: Brain Stem and Cranial Nerves

LESION DIAGRAMS: for each of the following diagrams indicate the structures involved by the cross hatched lesion(s). For each structure involved, indicate the expected clinical signs or symptoms with appropriate lateralization i.e. left or right (ipsilateral or contralateral). Where appropriate, indicate the vascular territory involved or the designation of the type of pathology and or the various names of the syndrome.

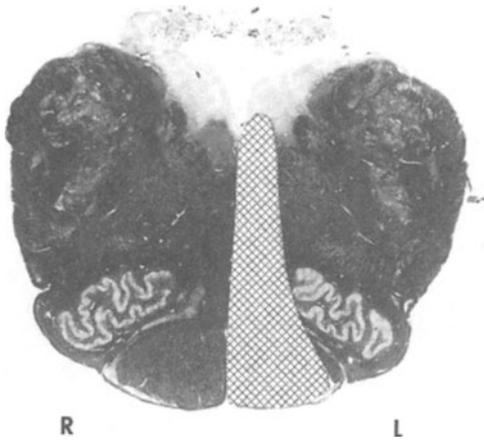


Figure 14-1.

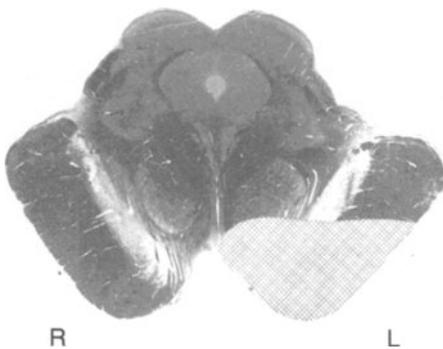


Figure 14-2.

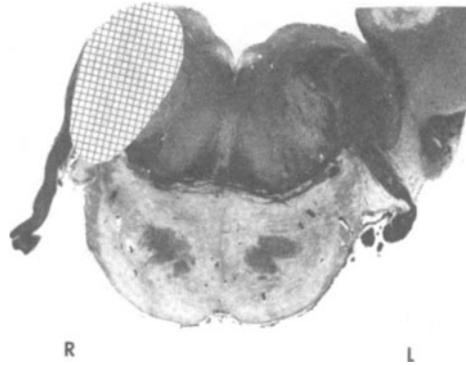


Figure 14-3.



Figure 14-4.

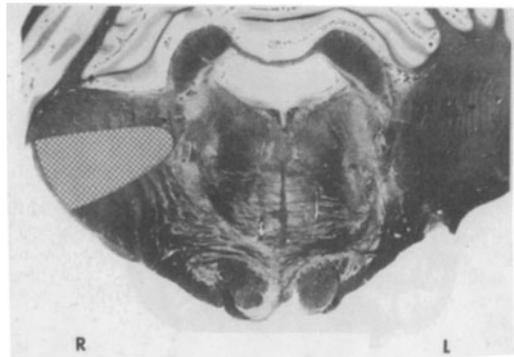


Figure 14-5.

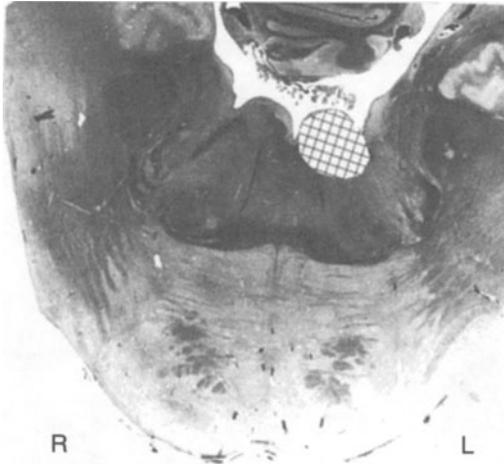


Figure 14-6.

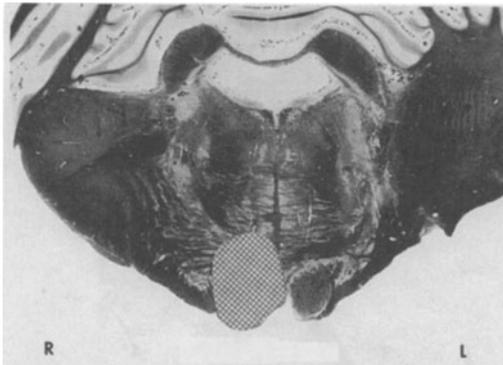


Figure 14-7.

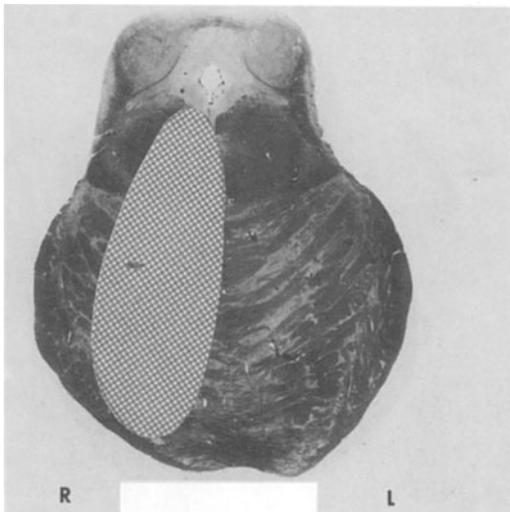


Figure 14-8.

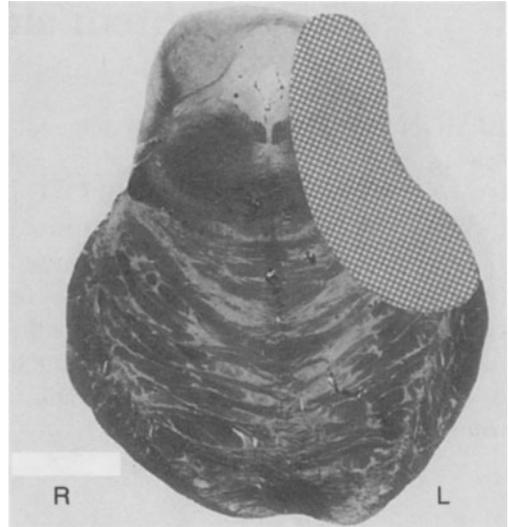


Figure 14-9.

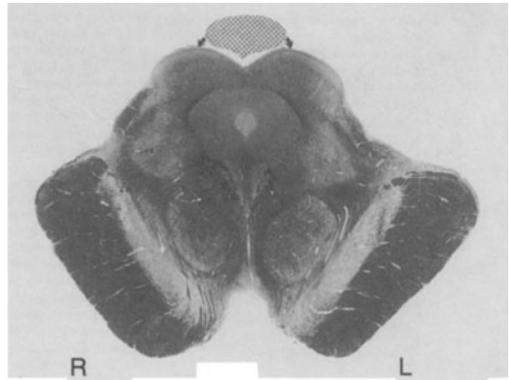


Figure 14-10.

**CASE HISTORY PROBLEM SOLVING
PART II - BRAIN STEM**

Each of the following case histories deals with disease at the level of the brain stem or of the cranial nerves. In some cases, it will be evident that the disease process involves the spinal cord as well as the brain stem. Some of the cases deal with intrinsic disease, some with extrinsic disease. For each case indicate and diagram the location of the lesion and indicate the nature of the pathological process. Where appropriate, indicate the name(s) of the syndrome, and or the vascular territory.

Case 14-1: (Patient of Doctor John Sullivan): A 31-year-old white policeman entered the hospital with symptoms of progressive difficul-

ty in speech, in swallowing, and with weakness of his grip in both hands. Fifteen months before admission, he first noted hoarseness, fatigability of his voice and faulty articulation. This has been slowly, steadily progressive, such that now speech was barely intelligible. Three months after onset of symptoms, he noted difficulty in swallowing both solids and liquids. He had a tendency to regurgitate liquids through his nose. Finally, 3 months before entry, the patient began to note increasing weakness of handgrip. He had no complaints referable to his legs. He denied any sensory symptoms. Six months before admission he noted the presence of muscle twitching with a diffuse distribution, but particularly in the arms and shoulders. He had lost ten pounds in weight and was easily fatigued. System review was otherwise entirely negative and general physical examination was well within normal limits.

NEUROLOGIC EXAMINATION:

Mental Status: Normal.

Cranial Nerves: Positive findings in cranial nerve function were:

- a) Stiffness and weakness of jaw muscles, with jaw clonus.
- b) Weakness of facial musculature, including eye closure, incomplete retraction of the corners of the mouth.
- c) Fasciculations were seen in the facial muscles.
- d) Gag reflex was very brisk, but palate moved weakly.
- e). Tongue would not be protruded; it was atrophic with fibrillations seen beneath the mucous membrane.
- f) Speech, as noted, was slurred, slow and strained.

Motor System:

- a) Weakness of handgrips.
- b) Atrophy was present in the thenar eminence and dorsal interosseus spaces. The proximal muscles were strong, without atrophy; leg muscles were strong and revealed no atrophy.
- c) Widespread muscle fasciculations were seen in both upper and lower extremities.

Reflexes:

- a) Deep tendon reflexes were increased symmetrically in the lower extremities.
- b) Plantar responses were extensor bilaterally (positive Babinski sign).

Sensory System: Normal.

QUESTIONS

1. Does this patient have a level lesion or a system disease?
2. Indicate the significance of the hyperactive jaw jerk and of the hyperactive gag reflex.
3. Indicate the significance of the fasciculations in the facial muscles, the atrophy of the tongue and fibrillations seen in the tongue.
4. Indicate the significance of the atrophy in the hands and the wide spread fasciculations.
5. Indicate the significance of the hyperactive deep tendon reflexes in the lower extremities and the bilateral Babinski signs
6. Where is the pathology?
7. What is the pathology?
8. Which diagnostic laboratory studies would assist in the diagnosis?
9. What would a muscle biopsy reveal?
10. Which neuroimaging studies, if any, are indicated in this case? What results are expected?
11. What is the prognosis? Discuss in terms of this patient in particular and then in terms of patients in general with this disease.

Case 14-2: [Patient of Dr. John Sullivan]

This is a 63 year old woman, who on the day before admission was suddenly seized by a sensation as though a weight had descended upon her head. She felt dizzy, staggered to a chair and called for help. It was noted that her speech was slurred and indistinct. As she looked at an object, it seemed to her to be indistinct. Since then she has had great difficulty in swallowing liquids because of a tendency to regurgitate through her nose. Solid foods did not seem to pass down. She also noted clumsiness of her right hand and of the right leg with staggering to the right side.

GENERAL EXAMINATION:

Blood pressure was 210/100 and she was obese with cardiomegaly.

NEUROLOGICAL EXAMINATION:

Mental Status: Brief mental status examination revealed her to be alert and cooperative, exactly oriented and rather apprehensive; there was no evidence of organic intellectual deficit.

Cranial Nerves:

a) examination of her fundi showed marked tortuosity of the retinal vessels; discs were normal; there are no hemorrhages or exudates

b) visual fields were full

c) Left pupil was 7 mm in size; the right 4 mm; both reacting briskly to light and accommodation; there was a partial ptosis on the right and slight enophthalmos on the right

d) There were no ocular palsies, but there were fairly well sustained quick nystagmoid jerks on gaze to either side

e) Right corneal reflex was absent; pain and temperature sensation on the right side of the face were lost, but touch sensation was intact

f) Hearing was acute bilaterally

g) Voice was somewhat hoarse; speech was slightly slurred and scanning. The right vocal cord was paralyzed; gag reflex was not elicited on the right, present on the left; uvula pulled to the left; 11th and 12th cranial nerve function were normal

Motor Examination:

a) There was instability of posture of the right arm with dyssynergia and intention tremor, which was also present in the right leg.

b) Strength of arms and legs was approximately normal

Reflexes:

a) Deep tendon reflexes in upper and lower extremities were equal.

b) Right and left plantar responses were flexor.

Sensory system:

a) There was a loss of pain and temperature sensibility throughout the left half of the body

b) Position and vibration sensation were intact

QUESTIONS:

1. Diagram the lesion using anatomical diagrams--be specific. Mark this case 1. Be certain to indicate laterality. Prepare a list of symptoms and signs in one column-anatomical structures involved in the opposite column.

2. What is the nature of the pathological process?

3. If vascular, indicate the vascular territory in terms of specific vessel.

4. Which labels or names are attached to this syndrome? Are other syndromes also present?

CASE 14-3: This 54 year old right-handed, obese white female was referred for evaluation of diplopia and ataxia. The patient had a 19-year history of diabetes mellitus initially treated with insulin and more recently with diet alone. The patient also had experienced significant pain in both lower extremities related to intermittent claudication, initially occurring on exercise but more recently occurring also at rest. Evaluation by the vascular surgery service had indicated bilateral carotid bruits.

On the night prior to admission, the patient had the acute onset of a diplopia. At the same time, she noted that she was no longer able to move her eyes upward and that in order to look up she had to turn her head back. At the same time, she developed a sense of unsteadiness. This morning she had a persistence of symptoms.

NEUROLOGICAL EXAMINATION:

Mental Status: Intact with marked anxiety

Cranial Nerves:

a) She had significant bilateral impairment of upward gaze

b) As she attempted to gaze upward, she had significant lid retraction.

c) In addition there was an indication of a weakness of right medial rectus.

d) The pupils were equal and responded to light

e) There was now no evidence of ptosis. There was no definite fatigue of the lids by repetitive movement.

Motor System:

- a) strength intact
- b) on examination of gait, patient walked on a broad base. She tended to fall to the left. She was unable to walk a tandem gait. She had a minor tremor of outstretched hands but no definite appendicular cerebellar findings.

Reflexes:

- a) deep tendon reflexes were 2+ except Achilles which were absent (probably) related to diabetes mellitus
- b) plantar responses flexor.

Sensory system:

Intact except for a decrease in vibration of toes (consistent with diabetes mellitus).

Hospital Course:

The patient had no additional progression. By day five, she had shown improvement in right medial rectus function and had improvement in her ability to look up.

QUESTIONS:

- 1) The bilateral lid retraction and the bilateral impairment of conjugate upward gaze in this case probably reflect involvement of _____?
- 2) The blood supply of this area is derived from branches of the _____ artery.
- 3) In this case impairment of upward gaze was due to an ischemic event, however, impairment of conjugate upward gaze is also commonly noted in relation to other pathological processes. Specify _____

CASE 14-4 [Patient of Dr. John Sullivan]:

This 65-year-old man entered another hospital complaining of headaches, severe vertigo, nausea and vomiting. The diagnosis of labyrinthitis was made and the patient seemed to recover after a few days. He remained well, however, only a few days when he again had severe vertigo and vomiting. The latter was so persistent and severe that he became dehydrated. On the morning of transfer to this hospital, at about 3:00 AM, the patient noted the sudden onset of weakness and numbness of his right thigh.

GENERAL PHYSICAL EXAMINATION:

The patient had a blood pressure of 110/80. There was a cold, clammy perspiration over his entire body. No other gross physical abnormalities were discovered.

NEUROLOGICAL EXAMINATION:

Mental Status: The patient was extremely restless and confused

Cranial Nerves:

- a) He had a paralysis of left external rectus muscle
- b) There was a coarse, irregular nystagmus on gaze to either side
- c) His fundi were quite normal
- d) The pupils and pupillary reflexes were normal
- e) there was a selective diminution of pain sensation on the left side of the face. Left corneal reflex was diminished.
- f) He had a left facial paralysis of peripheral type
- g) Hearing was markedly diminished on the left
- h) Gag reflex on the left was diminished; uvula pulled to the right; patient's speech was hoarse and he had a left vocal cord paralysis; He was unable to swallow without choking
- i) His tongue protruded slightly to the right

Motor System:

- a) On examination of his limbs, there was an intention tremor on the left involving both arm and leg
- b) His arms and legs were strong and there seemed no increased resistance to passive movement

Reflexes:

- a) Deep tendon reflexes were increased on the right side throughout
- b) Both plantar responses were extensor (positive Babinski Signs)

Sensory system:

- a) There was a loss of pain and temperature sensibility throughout the entire right side
- b) Other forms of sensation could not be adequately tested because of patient's inability to cooperate

QUESTIONS:

1. Considering in isolation the left lateral rectus palsy, the left peripheral facial weakness, the decrease in hearing in the left ear, the severe vertigo and the decrease in pain sensation on the left side of the face and right side of body. Diagram this specific lesion at the proper level. Label this Case #4.

2. What specific vessel supplies the area you have outlined above?

3. Now take into account those additional findings in this case that were not included in the consideration of Question 1. Indicate vessel responsible for the entire episode (there are 2 possibilities).

CASE 14-5: This right-handed, 43-year-old white housewife had the onset of deafness in the right ear. Rapid progression of deafness was noted; some tinnitus (sensation of ringing) was also noted. Caloric testing of labyrinthine function at that time indicated no response to cold or hot water on the right side. In the following month, the onset of a minor unsteadiness of gait was noted. Approximately five months later the patient noted defects in coordination of the right hand, particularly in typing, with a progression of unsteadiness of gait. At the same time a "numbness" sensation of pins-and-needles -- "like Novocain given by a dentist" -- was noted over the entire right side of the face. Relatively continuous pain was noted extending from the right side of the neck to the sub occipital area and right post auricular area. One month later difficulty in swallowing solids and liquids was noted.

PAST HISTORY:

Episodes of vertigo 5 to 6 years prior to admission. Right frontal headache since age 33.

NEUROLOGICAL EXAMINATION:

Mental Status: Intact.

Cranial Nerves:

a) Pain and touch sensation were decreased over all divisions of the trigeminal nerve on the right side, including the face, cornea, and the right side of the tongueb)

b) There was minimal flattening of the

right naso-labial fold and a poorer degree of eye closure on the right side than the left (orbicularis oculi).

c) There was no perception of voice in the right ear.

d) No vestibular response was present to ice water caloric testing in the right ear.

e) Minimal rotatory nystagmus was present on horizontal gaze with a minor degree of vertical nystagmus on upward gaze.

f) A minimal degree of dysarthria was apparent as regards guttural sounds.

Motor System:

a) Strength and tone were intact.

b) Cerebellar tests revealed a slight clumsiness in fine finger movements of the right hand.

c) Gait was slightly ataxic when performed on a narrow base with eyes open.

Reflexes:

a) Deep tendon reflexes were symmetrical and physiologic.

b) Plantar responses were flexor.

Sensory system: All modalities were intact.

LABORATORY DATA:

Cerebrospinal fluid protein was slightly elevated to 50 mg./100 ml. Skull x-rays were negative.

QUESTIONS:

1. This patient presents a typical example of classic neurological syndrome. Locate the lesion.

2. What is the most likely pathology to be found by the neurosurgeon in this location? Which pathological processes are also possible but less likely?

3. The initial involvement of the functions of cranial nerve VIII prior to involvement of other cranial nerves should provide a clue as to the structure from which this lesion arises.

4. Does the pattern of sensory disturbance over the right side of the face indicate primary involvement of the trigeminal nerve extrinsic to the brain stem or of the descending spinal tract and associated nucleus of the trigeminal nerve within the brain stem?

5. Were long sensory and motor tracts

within the brain stem involved by this lesion?

6. Predict the clinical picture that would have occurred if the lesion had progressed.

7. During the course of surgery, aimed at total resection in these cases, the facial nerve must often be sacrificed or damaged. Based on your understanding of the anatomical considerations in these cases, indicate why this occurs. Which studies may be performed during surgery in early cases to preserve residual hearing or facial nerve function?

8. Which critical diagnostic studies would you perform?

9. When should these studies be performed?

CASE 14-6: This 57-year-old housewife one day prior to admission suddenly developed double vision and a drooping of the right eyelid. In addition, she had difficulty walking. Examination of the patient in the emergency room revealed weakness of adduction of the right eye, ptosis of the right eye and a left extensor plantar response. She was admitted to the neurology service.

PAST HISTORY:

1. Moderate hypertension had been present for many years.

2. Four years previously, the patient had begun to have sudden 30-minute episodes of right-handed weakness and dysarthria. Twenty-six months prior to admission, she had a 15-20 minute episode of bilateral blurred vision, unsteadiness of gait and tingling paresthesias of the right hand and face. Twenty-two months prior to admission, she had episode of paresthesias involving either the left leg and arm or the right arm and leg. Examination at that point demonstrated increase deep tendon reflexes on the right side. Eighteen months prior to admission, she experienced 10-minute episodes of numbness of the left hand and face accompanied by renewed numbness of the right hand. Three days prior to admission she had a brief episode of right hand weakness.

3. The patient had been receiving anticoagulant therapy (Coumadin) for phlebitis for

several months prior to admission.

PHYSICAL EXAMINATION: The patient was obese and anxious with blood pressure elevated to 160/100.

NEUROLOGICAL EXAMINATION:

Mental status: the patient was alert and oriented. Recent memory was poor and delayed recall was limited to 2/4 objects. (All similar to 18 months previously)

Cranial nerves:

a) Ptosis of the right eyelid was present, but pupillary responses were intact.

b) At rest, the right eye was deviated out to the right and down. No medial or upward movement of the right eye was possible. (Note change).

c) A mild left central (supranuclear) facial weakness was now present.

Motor system: Strength was intact.

Reflexes: There were changes compared to examinations 22 and 18 months previously.

a) Deep tendon reflexes were now increased in the left lower extremity.

b) A left Babinski sign was now present with an absent left abdominal reflex.

Sensory system: All modalities were intact.

Carotid pulses: Strong bilaterally.

LABORATORY DATA:

1. Skull x rays demonstrated calcifications in the cavernous carotid arteries.

2. Electrocardiogram was normal.

3. Electroencephalogram demonstrated scattered multifocal slow waves.

4. Blood studies: Complete blood counts, serology, blood sugar and total cholesterol were normal.

5. Prothrombin time was 68% of normal.

6. The patient refused lumbar puncture and angiography.

QUESTIONS:

1. This is clearly a more complex case. However the new symptoms of diplopia, ptosis of the right eyelid plus the new findings of a paralysis of medial and upward movement of the right eye plus increased deep tendon reflexes in the left lower extremity plus a left

Babinski sign and a left central facial weakness should allow for the diagnosis and localization of at least one syndrome. What name do you assign to this syndrome? Does this syndrome involve a specific vascular territory?

2. How do you explain the earlier episodes?

a) Episodes of right hand weakness and dysarthria

b) Bilateral blurring of vision, plus unsteadiness plus tingling of the right hand and face.

c) Paresthesias left leg and arm followed by paresthesias of the right arm and leg.

3. Are there any additional questions, which you might pose for this patient?

4. If this patient had a severe headache at the time of admission, why might a lumbar puncture have been considered?

5. Why was angiography considered?

6 The patient refused both lumbar puncture and angiography. Would you have requested these studies or would you have requested other studies? If so present your plan of workup justifying each study.

7 Present your working diagnosis and final diagnosis. (There are several possible ways to tie together all of the episodes).

CASE 14-7: This 69-year-old white male was seen for outpatient neurological re-evaluation. His neurological problems began four years previously. The patient had a history of intermittent ptosis of the left lid. In addition, at times significant ptosis of both lids had been noted. Over the years, he had intermittent episodes of diplopia. He had also noted weakness of his jaw in chewing and weakness in shoulders or in his neck on exercise of these muscle groups.

NEUROLOGICAL EXAMINATION:

Mental Status: intact

Cranial Nerves:

a. The patient demonstrated a significant bilateral ptosis that was variable, it was significantly increased by exercise; it was more marked in the left eye.

b. The patient showed a significant bilater-

al defect in upward movement and of lateral movement of the right eye. The defect in lateral and upward movement was bilateral but was more marked on exercise.

c. The patient was able to smile for 8 seconds and then his smile began to evaporate bilaterally.

d. The patient had weakness in jaw muscles on repetitive opening and closure of jaw.

Motor System.

The patient had weakness in shoulder abduction, which developed after 10 repetitive movements. He had weakness in forward head movement which developed after 4 repetitive movements. No definite atrophy was present, no definite weakness was otherwise present in the extremities.

Reflexes: Deep tendon reflexes were intact, plantar responses were flexor.

Sensory System: intact.

QUESTIONS:

1. Indicate the diagnosis. Be specific.

2. Where is the defect located in this disease? Be specific!!

3. Discuss the underlying pathophysiology.

4. Which tests would confirm the diagnosis and produce temporary improvement?

5. Outline therapeutic approaches.

CASE 14-8: This 56-year-old white housewife was admitted for evaluation of episodes of stupor and cyanosis associated with severe laryngeal stridor (high pitched and harsh respiratory sounds) and stertorous breathing. Laryngeal stridor had been present for 20 years and had grown worse during the last 5 to 6 years. An episode of anoxia during a Cesarean section 18 years prior to admission may have been a complication of this problem. Sixteen years prior to admission numbness and weakness of the right leg had been noted. Progression had occurred in the last 2 years and episodic unsteadiness of gait had been noted. During this same period, weakness of the right hand had developed. During the one year prior to admission the patient had three hospital admissions related to episodes of

coma and cyanosis. Each had followed a several-week period in which there was increased stridor and increased accumulation of tracheal-bronchial secretions with frequent periods of daytime sleepiness.

NEUROLOGICAL EXAMINATION:

Mental Status: Intact.

Cranial Nerves:

a. Pain sensation was selectively decreased over all three divisions of the right side of the face with decreased right corneal reflex.

b. Laryngoscopy revealed paralysis with atrophy of the left vocal cord. The right cord moved but was inhibited on abduction, indicating partial paresis.

c. Horizontal nystagmus was present on lateral gaze with minimal vertical nystagmus on upward gaze.

Motor System:

a. Atrophy of the right upper extremity was present including the shoulder, arm, and hand.

b. Weakness was present in the right upper extremity -- approximately 50 per cent of normal strength at shoulder, elbow, wrist, and fingers. Weakness without atrophy was present in the right lower extremity and to a lesser degree in the left lower extremity.

c. Spasticity was present on passive movement at the right knee and ankle and to a lesser degree at the left knee.

d. Gait: There was circumduction of the right leg with unsteadiness on rapid turns.

e. Cerebellar tests were negative.

Reflexes:

a. Deep tendon:

Biceps: right, 0; left, 0

Triceps: right, 0; left, 0

Radial: right, 0; left, 0

Patellar: right, 4+; left, 3+

Achilles: right, 4+; left, 3+

b. Superficial reflexes:

Plantar: extensor on the right and possibly extensor on the left

Abdominal: right, 0; left, 0

Sensory system:

a. Position, vibration, and touch were intact.

b. Pain and temperature were selectively decreased in a cape-like distribution over the shoulders.

LABORATORY DATA:

1. Cerebrospinal fluid: normal (pressure 150, cell count 0, protein 45 mg./100 ml.)

2. Skull x-rays were negative.

3. Cervical spine x-rays revealed minor non-significant degenerative changes at C5-C7.

SUBSEQUENT COURSE:

The patient was readmitted to the hospital 10 weeks later in a semi comatose and cyanotic condition. She required an emergency tracheostomy and 48 hours of respiratory assistance. Subsequent neurological examination was unchanged from that recorded earlier.

QUESTIONS:

1. Where is the lesion? Does this lesion involve a single localized segment or are several segments involved?

2. Is the pathology limited to the brain stem or to the spinal cord?

3. Indicate what structures are involved to produce:

a. laryngeal paralysis,

b. cape-like deficit in pain and temperature but sparing touch and vibration,

c. atrophy of all muscle groups in right upper extremity,

d. a bsence of deep tendon reflexes in both upper extremities, and

e. defective pain sensation on the right side of the face.

4. Indicate the most likely pathology and the probable prognosis.

5. What diagnostic tests should be undertaken to establish the diagnosis?

6. Why did the patient have episodes of coma and excessive daytime sleepiness? (There are several explanations)