# Case #1

ed by: Rosa E. Fiol, M.D.
School of Medicine
University of Puerto Rico
San Juan, Puerto Rico 99036

Reference: No. A-632-74

A sixty-two year old Puerto Rican male had a history of urinary tract infection and arterial hypertension. He lived somewhere in Pennsylvania but was born and raised in a Puerto Rican town surrounded by brooks. He developed low back pain and weakness in the lower extremities. He refused to have a myelogram and returned to Puerto Rico. Neurological exam revealed the complete loss of sensation at the $T_8$ level anteriorly and $T_6$ posteriorly including pain, touch, vibration and position. He became comatose and died four days after admission to the hospital.

Necroscopy revealed bilateral bronchopneumonia, multiple granulomas of liver and a cortical adenoma of the right kidney. The spinal cord at $T_6$ and below showed no differentiation between grey and white matter. The cut surface was soft, mottled and grey-red.

Microscopic Pathology: Hematoxylin-eosin stained section of the spinal cord.

Points for Discussion:

1. Diagnosis
2. Importance of history of brook bathing in Puerto Rico
3. Differential diagnosis of myelitis.

# Case #2

Submitted by: Cheng-Mai Shaw, M.D.
University of Washington
Seattle, Washington 98195

Reference: (NP 4362, University of Washington Medical School)

This 14 year old boy was said to have Batten's disease since about age 10. He was first evaluated at age 11 in November, 1971 for intellectual deterioration and incoordination. A rectal biopsy at that time revealed storage material in the ganglion cells. In April, 1972, examination showed an obese, dysarthric and sluggish boy who walked stooped over, bent at the knees and hips. He had limited upward gaze, questionable non-macular vision in the left, abnormal ocular fundi with pigmentary degenerated macula, hyperactive DTR's, sustained ankle clonus, upgoing toes, moderate spasticity (worse in the legs than in the arms), ataxia and Romberg's sign. There was a small right subcapsular cataract. EEG was abnormally slow and mildly epileptiform. Urine screen for mucopolysaccharides was negative. The diagnosis was Batten-Spielmeyer-Vogt disease (or juvenile cerebromacular degeneration). The patient also had skin lesions suggesting epidermolysis bulbosa in the extremities and trunk for 3 years, but a skin biopsy in May, 1972 showed only subacute dermatitis.

The patient was reevaluated in January, 1974 for a decubitus ulcer of the right hip. He had not walked for 6 months, his extremities were weak and wasted. He responded to pain and the simplest commands but was unable to speak, only uttering sounds. He had difficulty swallowing and choked frequently. Seizures, which had been only occasional in May 1972, had increased to one per day and included jerking of the mouth and upper limbs. Neurological examination in March, 1974 showed frequent yawning and chewing movements, positive snout reflex and glabellar rugae. He was able to seen and follow an object but did not respond to hand clapping. Gag reflex was absent. Muscle tone was increased with contractures of both elbows and knees. DTR's were 4+ and Babinski's sign was present bilaterally. He was on Dilantin, 200 mgs. per day, which seemed to control the myoclonic jerks. He was seen again in September 1974, essentially unchanged.

The patient died in October, 1974 of bronchopneumonia following a cold for several days.

General autopsy: Bronchopneumonia

Gross neuropathology: Mild to moderate degree of ventricular dilatation, granular appearance of cerebellar cut surface. Whole brain weight: 1030 grams.

Microscopic slides: Frontal cortex: One stained with LFB-PAS-H, one unstained.

Points for Discussion:

1. Diagnosis.

## Case #3

Submitted by: John Pearson, M.D.
New York University Medical Center
550 First Avenue
New York, N. Y. 10016

Reference: A-3-74

A 27-year-old male, who had never grown facial hair, collapsed suddenly at work. He gradually recovered after intramuscular caffeine injection. Temperature was 96°F. The tongue was bitten. Pupils were equal and reacted sluggishly to light. There was horizontal nystagmus. Tendon reflexes were depressed. Kernig's sign was positive. Spinal tap was refused. Serum chloride was 120 MEq./L. Two days later, transitory apathy was noted. Blood pressure was 80/60 and pulse 56/min. Four days thereafter, he suddenly became unconscious and had a systolic blood pressure of 80 mm. Hg. Later there was an episode of projectile vomiting. He was found dead the following day.

General Necropsy Findings: The testes and thyroid were small. The right adrenal gland was small, the left contained a cortical adenoma.

Adjacent to the right adrenal gland was a 4 x 3 x 1 cm. smooth-surfaced para-aortic ganglioneuroblastoma; the tumor was histologically typical and did not resemble the nodules described below.

CNS: At the base of the brain, a smooth, sessile, firm, pale-pink mass mottled with flecks of yellow protruded from the tuber cinereum. After fixation, the focally necrotic tissue was found to infiltrate and destroy parts of the hypothalamus bilaterally. Sixteen similar well demarcated, roughly spheroidal nodules, 0.5 to 1.6 cm. in maximum dimensions, were found widely dispersed in the brain. They could not readily be separated from adjacent tissue. Nodules were found in the cerebral cortex, amygdala, right caudate head, left caudate tail, posterior parts of the right hippocampus, adjacent thalamic pulvinar, left cerebellar cortex and postero-lateral aspect of the right medulla. Other nodules were entirely within cerebral and cerebellar white matter.

The ventricular surfaces were normal. There were no tubers on the cortical surfaces. No abnormality was found in the spinal cord and attached nerve roots. Microscopically, in addition to the nodules, many ectopic neurons were found in white matter.

Microscopic Pathology: Hematoxylin and Eosin stained section plus one unstained slide.

Points for Discussion:

1. Are these nodules neoplastic? Yes

2. If not, what are they?

3. If neoplastic, what is their nature? Glial

## Case #4

Submitted by: William I. Rosenblum, M.D.
Medical College of Virginia
Richmond, Virginia 23298

Reference: A-71-75

The patient was a 55 year old White lady with a history of rheumatic heart disease. She was said to have had a left middle cerebral artery occlusion several years before death, and she had a residual hemiparesis. She fell 25 days antemortem and then complained of stiff neck and headache. A lumbar puncture revealed xanthochromia. Congestive heart failure, oliguria, hypotension and deep coma developed before death.

Autopsy revealed massive, recent bilateral pulmonary emboli and idiopathic, hypertrophic, subaortic stenosis. There was recent subarachnoid hemorrhage, thought to be caused by a leaking vascular malformation in the medulla. There was fresh infarction of portions of the right thalamus and cerebral cortex. No infarct was found in the left hemisphere. There was "granular atrophy" of much of the cortical surface, bilaterally.

Of interest are the small blood vessels found in virtually all the sections of cerebrum.

Microscopic Pathology: Slides stained with Azo-carmine.

Points for Discussion:

1. What do you call this lesion of vessels? ? cap. clusters
2. Is it the cause or the result of ischemia? ? might be more of a incidental finding
3. Once formed, does it, in turn, produce symptoms of neurologic and/or psychiatric nature?

# Case #5

Submitted by: Margaret L. Grunnet, M.D.
Department of Pathology
University of Utah
Salt Lake City, Utah 84132

Reference: A 4-74

The patient was a 33 year old White male with a history of chronic, progressive renal failure since 1960. He had received a cadaver transplant in 1970, following which, he had been treated with Prednisone and Imuran. In November, 1973, he developed a flu-like syndrome associated with fever, headache and nausea. On physical examination, he had papilledema, and a lumbar puncture showed an opening pressure of 324 mm. $H_2O$. Chest x-ray showed nodular, confluent densities which increased in size rapidly over the next few days. A pleural effusion developed. He became progressively hypoxic and obtunded without focal neurological signs. Repeated taps of the pleural effusion were ineffective. A lung biopsy was performed, which was said to show pleomorphic lymphoma. He died following a six-week course in the hospital.

At autopsy, he had a necrotizing "lympho-proliferative" disease involving the lungs. Lymph nodes and spleen were spared. He also had end-stage kidneys and acute pyelonephritis of the transplanted kidney. Gross inspection of the brain revealed mild edema with opaque leptomeninges. Multiple coronal sections revealed many hemorrhagic grey lesions of cerebral cortex, which measured up to 1-1/2 cms. in diameter.

Microscopic Pathology: Slide is stained with hematoxylin and eosin.

Points for Discussion:

1. What is the disease process? Reticulum cell sarcoma (some variant of lymphoid granulomatosis)
2. How often does it affect the central nervous system?
3. What are possible etiologic factors?

# Case #6

Submitted by: M. Meyer, H. Powell, P. Lampert
University of California, San Diego
La Jolla, California 92093

Reference: UCSD A-59-75

This 56 year old White male with known cauda equina syndrome and atherosclerotic coronary vascular disease was admitted to the San Diego VA Hospital because of a possible myocardial infarct. No definite clinical or laboratory evidence of an acute infarction was found. He was treated for congestive heart failure, seemed to stabilize but died of a cardiorespiratory arrest.

His past neurological history was involved. In October, 1941, he was in an airplane accident and sustained a lumbar spine injury. Over the next few months, he began to experience pain in the hips and both legs. Physical examination several months after the accident showed atrophy of the left thigh and calf muscles. Spine films showed only calcareous changes of $L_4$ and $L_5$ and myelography failed to reveal any additional defect. In July, 1942, a spinal fusion was attempted. The incision drained for seven months before it finally healed. The patient did well until 1945 when back pains worsened. Skull and spine films showed retained contrast medium and the diagnosis of arachnoiditis was made. In 1948, the pain became so severe that surgery was again necessary. One procedure was performed to remove the dense lumbar scar tissue and a second operation to fuse the spine. Symptoms persisted and in 1950, the spine was re-explored with removal of the $L_{4-5}$ disc.

Over the next 10 years, he suffered intermittent back pain, leg pain and "migraine" headaches with gradual loss of bowel and bladder function. In 1958, he developed a left facial palsy. Four years later, he suddenly lost the hearing in his right ear and began to notice hearing impairment in the left. A workup included a pneumoencephalogram which failed to reveal and cerebellopontine angle abnormality. In 1972, he again suffered severe spasms and was admitted for a second cordotomy. His deafness was now complete. Skull x-rays showed numerous intracranial densities which were thought to be intracortical. A CPA myelogram showed findings consistent with a left acoustic neuroma. Over the ensuing years, the patient had to be placed in a nursing home because of his deafness, heart disease and complete incontinence of bowel and bladder.

Autopsy Findings: 1. Acute congestive failure. 2. Severe atherosclerotic disease. 3. Acoustic neuroma. 4. Meningioma.

Microscopic sections of spinal cord are stained with hematoxylin and eosin.

Points for Discussion:

1. Nature of deposits? Thrombosis ?
2. Effect of myelography on meninges? Fibrosis
3. Induction of tumors by contrast media? Thrombosis

# Case #7

Submitted by: Julio H. Garcia, M.D.
University of Maryland Hospital
Baltimore, Maryland 21201

Reference: SP74-7943

A previously healthy 15-year-old Black female was seen in October, 1974 because of difficulty moving her right leg and right arm for the past two days. This was not preceded by seizures, syncope or sensory disturbances. She had suffered from chronic headaches since the age of 11 and for the first time, she had begun to have projectile vomiting. She complained of no difficulties with vision or other related symptoms.

On admission, she was described as an obese, well-oriented, pleasant patient with obvious weakness of the right limbs, no cranial nerve disturbances and no sensory deficits. There was no evidence of papilledema.

Brain scan showed a positive uptake in the left parasagittal-parietal region and the arteriogram showed an increased vascularity in the same area. The EEG disclosed a left temporal focus of "spike activity".

Other laboratory data and the remainder of the physical examination were within normal limits.

At craniotomy, a sample of an intraparenchymal mass was obtained.

The slides distributed are stained with hematoxylin and eosin.

Points for Discussion:

1. Diagnosis of this tumor: Glial? Mesodermal? Mesench. Chondrosarcoma or Myxofibrosarc
2. Prognosis ?
3. Mode of therapy Resection

# Case #8

Submitted by: J. G. Chi, M.D. and N.K. Blank, M.D.
Children's Hospital Medical Center
Boston, Massachusetts 02115

Reference: S75-5497

This white baby boy was born to a 24 year old $G_2P_1$ mother after a full-term, uncomplicated pregnancy by vaginal delivery. At birth, the infant was pink and cried spontaneously. At four hours of age, he was noted to be having apneic episodes which were alleviated by face mask $O_2$. Physical examination disclosed an enlarged head with wide open fontanelles and sutures. At twelve hours of age, severe tachypnea and grunting were noted, followed by generalized tonic-clonic seizure movement. The routine laboratory analyses were within normal limits save for a hematocrit of 38%.

The infant was transferred to Children's Hospital Medical Center on the second day of life. Tachycardia and tachypnea were present. He was alert, had a good Moro response, excellent suck, and normal muscle tone. The deep tendon reflexes were symmetrically active, bilateral ankle clonus was elicited, and a questionable left Babinski was present. On the next day, a CAT scan showed a 5.5 x 5.5 cm. mass within the lateral ventricle. An angiogram showed a vascular mass in the area of the left trigone which was supplied by the left vertebral and anterior choroidal arteries.

A left craniotomy with total excision of an 80 gram tumor was removed from the left trigone region of the ventricle on the fifth day. He had a cardiac arrest post operatively from which he was successfully resuscitated but he remained comatose and died at one week of age.

Necropsy examination disclosed the presence of massive hydrocephalus, extensive subarachnoid hemorrhage, a hematoma in the right lateral ventricle. There was no residual tumor. Perinatal telencephalic leukoencephalopathy was found on microscopic study.

Microscopic Pathology: Hematoxylin and Eosin section of the tumor.

Points for Discussion:

1. Diagnosis. Malig. choroid plexus papilloma (carcinoma)
2. Criteria for differentiating choroid plexus papilloma from ependymoma.
3. Criteria of malignancy in congenital tumor.

## Case #9

Submitted by: J. T. Hughes
The Radcliffe Infirmary
Oxford, England 0X2 6HE

Reference: 993

A 34-year-old man was the driver of a car involved in a "head-on" collision. He sustained a non-penetrating 'steering wheel' chest injury which caused a rupture of the aorta 2 cm. distal to the left subclavian artery. Neurological examination was normal.

At a cardio-thoracic surgical unit, through a left thoracotomy at the level of the 4th rib, the damaged part of the aorta (see diagram) was resected and replaced by a 3.5 cm. long Dacron crimped tubular graft, whilst a left atrium to left femoral bypass carried on the aortic blood flow for the required 70 minutes. The blood pressure in the upper aorta was monitored through a cannula in the right radial artery. At the end of the operation, the normal anatomy of the aorta was restored and bi-lateral femoral pulses were recorded.

On recovery from the anesthetic, the patient was unable to move his legs and the first post-operative examination indicated a severe para-paresis with accompanying sensory loss. By the following day, it was clear that he had complete areflexic paralysis of both lower limbs with complete loss of sensation below the T9 - T10 sensory level. Bladder and bowel function were also paralysed. This neurological state remained static until his death five months later due to mediastinal hemorrhage from a leak of the Dacron graft anastomosis.

The two diagrams provided give the gross findings at necropsy in the aorta, and the histological findings in the spinal cord, every segment of which was sectioned.

The slide of three transversely cut sections of spinal cord (L1, L2 and L3) is stained by hematoxylin and eosin.

Points for Discussion:

1. The mechanism by which the spinal cord was damaged.
2. Pertinent features in clinical history, operative procedure and necropsy findings as related to Point #1.