

Case #1

Submitted by: David F. Dean, M.D., University of South Alabama,
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Armed Forces Institute of Pathology, Washington, D.C.

Reference: AFIP No. 1474870

This 19-year-old caucasian male sustained a severe comminuted open left fronto-temporal-maxillary-orbital-basilar skull fracture in an auto accident in Mississippi. The same day, he had a left frontal craniotomy, removal of epidural and subdural hematomas, debridement of brain lacerations, repair of dural tears, reapproximation of bone fragments, instillation of a Silastic orbital floor stint, suture of lacerations and a reapproximation of his left ear. Following surgery, he was placed on intravenous ampicillin and IM Kantrex as well as IM dexamethasone. Seven days after surgery, the patient began having spiking fever to 101° and his left para-orbital tissues became indurated, red and tender. CSF examination disclosed no bacteria and bacterial cultures were negative. The patient became more toxic. The sutures were removed from the wounds on the ninth post-operative day and drains were instilled which did not drain much. On the tenth post-operative day, the Silastic stint was removed and seropurulent material was sent for culture. On the 12th day, his left eye was enucleated and the multiple bone fragments removed. His wound did not bleed, appeared dark and necrotic and relatively avascular. Brain tissue appeared to be herniating into the orbit.

This patient responded to surgical debridement and therapy with a total of 3.2 grams of amphotericin B. He is alive and doing well.

Microscopic Pathology: The eyeball showed panophthalmitis and ischemic necrosis of the retina secondary to the orbital infarctions. Portions of cerebral cortex within the biopsy showed similar inflammation and necrosis. The H & E slide submitted was prepared from the orbital tissue.

Point for Discussion:

1. Diagnosis.

Case #2

Submitted by: John T. McGrath, V.M.D. and Edgar Fernandez, V.M.D.
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Philadelphia, Pa. 19104

Reference: No. 74-470

This 6-year-old male cocker spaniel dog was presented with a history of head tilt to the left, progressive ataxia with occasional falling and circling movements. At presentation, there were head tilting to the left, right sided palsy of the 5th and 6th cranial nerves and exaggerated spinal reflexes. The cerebrospinal fluid contained no cells and protein value was normal. Skull radiographs and angiography showed no abnormality. The animal was killed in lateral recumbancy.

Necropsy revealed a granular pink tumor mass occupying the right lateral region of the fourth ventricle and infiltrating the adjacent brain parenchyma. Ventrally, the leptomeninges in the region of the pons and right trigeminal nerve appeared infiltrated.

Microscopic Pathology: Hematoxylin-eosin stained section of the cerebellum-medulla with the gross tumor. Also note neoplastic tissue in the adjacent cerebellar roof nuclear area and tumor tissue in the left medulla oblongata especially involving the left sensory 5th and vestibular areas.

Points for Discussion:

1. Diagnosis.
2. Is this a single mixed tumor or more than one tumor?
3. Are there intranuclear inclusions in that area simulating a gemistocytic astrocytoma.
4. Is this an example of neoplastic "field" transformation and multicentricity?

Case #3

Submitted by: Angeline R. Mastri, M.D. and Joo Ho Sung, M.D.
University of Minnesota Medical School
Minneapolis, Minnesota 55455

Case A (Ref. No. 74-288)

This 37-year-old housewife with von Recklinghausen's disease had a neurofibrosarcoma excised from the left thigh in February, 1973. She subsequently developed multiple pulmonary metastases which were treated with a number of cancer chemotherapeutic agents with little response. Her final admission in July, 1974 was because of increasing respiratory distress and fever. Physical findings included a large head with frontal bossing. Pupils were meiotic. There were many cafe au lait spots over the entire body and numerous cutaneous nodules over the trunk. No neurological findings were noted. The patient was thought to have pneumonitis superimposed on pulmonary metastases and was treated with antibiotics and chemotherapy. She received a low dose of methotrexate followed by citrovorum factor following which she developed severe symptoms of methotrexate toxicity including renal and hepatic failure, stomatitis and pancytopenia. She became progressively obtunded, developed bilateral Babinski signs and supraventricular tachycardia and died on 7-22-74. Autopsy findings: Metastatic sarcoma in both lungs, and benign neurofibromas of the skin. Neuropathological findings: Striking brown discoloration of the globus pallidus and substantia nigra, bilaterally with numerous spheroids and abundant pigment.

Case B (Ref. No. 74-435)

This 52-year-old white man was admitted to University Hospitals because of dyspnea on exertion and shortness of breath. He had had two episodes of myocardial infarction and intermittent chest pain for approximately three years. He had known von Recklinghausen's syndrome with numerous cutaneous fibromata. In 1955, a pheochromocytoma was removed at the Mayo Clinic. Physical examination was not remarkable except for a grade II systolic heart murmur and hepatomegaly. On the third day of hospitalization, the patient had the onset of a slow irregular heart rate leading to ventricular fibrillation from which he could not be resuscitated. Autopsy findings: Old and possibly recent myocardial infarction; severe atherosclerosis with complete occlusion of right and left coronary arteries; congestion of the lungs and spleen. Neuropathological findings: Mild atherosclerosis of the cerebral vessels, prominent pigmentation and degeneration of the reticular zone of the substantia nigra, bilaterally, with numerous spheroids and abundant pigment deposition. Similar but milder changes were noted in the globus pallidus on histological examination. Heterotopic glial tissue was noted in the cerebellar leptomeninges.

Slides Submitted: Section of midbrain from case A (H & E)
Section of substantia nigra from case B (unstained)

Points for Discussion:

1. Nature of the pigment
2. Relation of this degeneration to von Recklinghausen's disease
3. Relation to Hallervorden-Spatz disease, if any

Case #4

Submitted by: J. G. Chi, M.D. and E. C. Dooling, M.D.
The Children's Hospital Medical Center
Boston, Massachusetts

Reference: 74-152 (J.S.)

This 5-1/2 year old white boy was admitted because of headache and high fever of one day's duration. One week earlier, he had travelled through an endemic eastern equine encephalitis area in Rhode Island. On the day prior to admission, he was given 2 doses of ampicillin. The child continued to be febrile (40° C.), and complained of headache, stiff neck and dizziness. He vomited several times and had episodes during which his eyes were said to "roll back." On examination, he was confused and disoriented but he could follow simple commands. There were twitches of the eyelids and mouth and occasional jerks of the extremities. Petechiae were present on the neck, the throat was hyperemic and there was nuchal rigidity. Except for altered sensorium and equivocal plantar responses, the neurological findings were unremarkable. A lumbar puncture yielded clear fluid under an elevated pressure (300 mm H₂O). It contained 290 white blood cells (50% polymorphs and 50% monocytes), no red blood cells, 245 mg% protein and 95 mg% sugar. The peripheral white blood cell count was 19,400 with a shift to the left; platelets, hemoglobin, prothrombin time and electrolytes were normal.

Eight hours after admission, the child was stuporous but still responded to his name. Twelve hours after admission, he had a generalized seizure and subsequently became decerebrate and unresponsive. The fundi showed blurred disc margins, flame hemorrhages, and absence of venous pulsations. He was treated with Mannitol and Glycerol. Within six hours of the seizure, the pupils were dilated and fixed. He required assisted ventilation two hours later, and when carotid arteriography was performed, there was no significant passage of dye into the brain. Two electroencephalograms within the next 24 hours were isoelectric, and he was declared dead approximately 43 hours after admission and 3-1/2 days after first becoming ill.

Past history was unremarkable except for DPT vaccination two months earlier, two oral polio vaccinations 2 months and one week earlier, rubella and measles vaccinations six weeks earlier, and mumps vaccination one week earlier, in readiness for school.

All bacterial cultures, including blood and cerebrospinal fluid, were negative. Viral titers were not significantly elevated.

Visceral Pathology: General postmortem examination revealed no significant lesions in any organs, including the spleen, kidneys and lymph nodes.

CNS Pathology: The lesions exemplified by those in the submitted slide were scattered throughout the neuraxis without topographic specificity. The slide submitted is stained with Luxol fast blue - Hematoxylin and Eosin.

Post for Discussion:

What is the disease ?

Submitted by: Raymond A. Clasen, M.D., Presbyterian -
St. Luke's Hospital, Chicago, Illinois 60612

Reference: 575-6848

A 58 year old Negro female was admitted to hospital on 7/3/74 complaining of headache of six months duration, which had become progressively more severe following an apparent seizure three weeks prior to admission. At the time of admission, she was lethargic and disoriented. The right optic disc was elevated and she showed a left-sided weakness with a Babinski sign. A right carotid angiogram showed a right frontal tumor mass. This was resected on 7/8/74. The neurosurgeon thought that the tumor was a typical meningioma but he had some difficulty removing the tumor from the underlying brain.

Postoperatively, the patient was given radiation therapy. One month after surgery, she showed signs of accumulation of fluid below the craniotomy flap and purulent material was aspirated. On 8/15/74, she underwent craniotomy and debridement. An abscess was found at the site of surgery. The patient responded to treatment and was discharged on 9/18/74. At the time of discharge, her wound was well healed and neurologic examination was within normal limits. At the time that this protocol was written, the patient's condition had not changed.

Microscopic Pathology: Two slides are submitted from the surgical specimen; one is stained with Hematoxylin - Eosin and the other by the Gomori trichrome method.

Point for discussion:

1. Diagnosis.

Case #6

Submitted by: Minoru Suzuki, M.D., Department of Pathology,
Baylor College of Medicine, Houston, Texas

Reference: 68382

This 48 year old black lady had been on stilbesterol medication. She was admitted to hospital because of headache. She was delusional and was having visual hallucinations. Physical examination revealed sparse pubic hair and atrophic dermatitis. The pupils and eye grounds were normal. The sensations of touch, pain, vibration and position were intact. The clinical impression at that time was paranoid schizophrenia.

Hematocrit 31%; WBC 4,500 (PMN 46%, juv 2, lymph 42, mono 4, eos 6); glucose 104 mg%; electrolytes Na 115 mEq/L, k 3.8, Cl 90, CO₂ 21; arterial gases pH 7.51, pCO₂ 21.5 mmHg, pO₂ 74, O₂ sat 92%; CPK 22 units (normal 0-35); VDRL nonreactive. The CSF examination showed 4 lymph per mm³, 55 mg% protein, 74 mg% glucose, 114 mEq/L chloride, nonreactive VDRL, and colloidal gold 00 00 00 00 00. Hormonal assays of blood or urine were not done. T-3 test was 20% (normal 25-35) and T-4 9.8 micro gm% (normal 5-14).

The patient began having seizures, became febrile and stuporous and was placed on chlorpromazine. She expired shortly after a seizure.

General Pathology: The adrenals were atrophic; each weighed 2.5 gm. The thyroid weighed 23 gm. and show no abnormalities. The ovaries were normal in size. The pituitary was firm, gray and weighed 1.5 gm.

Microscopic Pathology: An H & E stained section and an unstained section of pituitary gland are submitted.

Points for Discussion:

1. Diagnosis.
2. Etiology.

Case #7

Submitted by: Bruce Horten, M.D., Stanford University Medical Center
Stanford, California 94305 and B. A. Van Ketel, M.D.
Academisch Ziekenhuis, Utrecht, The Netherlands

Reference: C-1771

This 11 year old boy was well until December, 1973, when he developed episodes of headache interspersed with vomiting. These "migraine-like" attacks occurred approximately once each week. By Spring of 1974, though performing well at school, the child suffered an increase in the frequency and severity of his headaches and so entered the Utrecht University Clinic for thorough neurological evaluation.

Physical examination revealed bilateral papilledema and right hyper-reflexia. No extremity weakness was demonstrated. Diagnostic studies included an EEG and a brain scan, both of which suggested a mass lesion within the left parieto-occipital lobe.

At craniotomy, three interconnected, firm, well-demarcated tumor masses, each 5 cm. in diameter, were resected from the left parieto-occipital region. Post operatively, the patient developed right hemiparesis and aphasia, which have since considerably cleared.

Microscopic Pathology: Tissue sections stained with hematoxylin-van Gieson and untuned Bielschowsky.

Points for Discussion:

1. Diagnosis.
2. Gross and microscopic features of such a tumor.
3. Significance of the dense stromal tissue.
4. Value of silver impregnation in the diagnosis of this tumor.

Case #8

Submitted by: Hernando Mena, M.D., Julio H. Garcia, M.D. and
J. E. Viloria, M.D., University of Maryland Hospital,
Baltimore, Maryland 21201

Reference: A73-107 (University of Maryland)

A 31 year old black male had a diagnosis of chronic glomerulonephritis at age 27. He was dialysed for 8 months prior to November 9, 1972- at which time, he had a unilateral cadaver-kidney transplant. Immunosuppressive therapy included: (Imuran), antilymphocytic globulins, prednisone, prednisolone and hydrocortisone. Dialysis was continued since the amount of urine filtered was small.

While being dialyzed one week after transplantation, he convulsed and became abruptly unresponsive. He responded to painful stimuli by opening the eyes slightly; there were no spontaneous movements of extremities. Eyes wandered aimlessly, pupils were symmetrical, 3-4 mm. in diameter and responded to light normally; involuntary eye movements were normal; ciliospinal reflex was present, neck was supple, extremities were flaccid and deep reflexes were preserved. There were weak withdrawal attempts after painful stimulation. After regaining "consciousness", he showed neurologic recovery during the next few weeks although he remained dysphasic. He could communicate appropriately in writing. Serum creatinine levels which had been falling began to rise; this was attributed to a rejection crisis, and he received increased doses of immunosuppressive agents. After November 29, 1972, dialysis was not necessary and renal function was nearly normal. His neurologic status was much improved but he was unable to talk at the time of his discharge on December 21, 1972.

He was readmitted to hospital five months later because of rising serum creatinine, increased blood pressure and decreased urine filtration. He had several successive hypotensive and resuscitative episodes before death in May, 1973.

General Pathology: 1) "End-stage nephropathy", 2) chronic rejection (transplanted kidney), 3) occlusion of right renal vein and artery, and 4) left ventricular hypertrophy.

CNS Pathology: The brain shows two cavitory lesions measuring up to 2 cm. in diameter in the centrum semiovale. The slide submitted is representative of these lesions.

Microscopic Pathology: An H & E stained slide is submitted.

Points for Discussion:

1. Diagnosis.
2. Etiology.

Case #9

Submitted by: Samuel K. Ludwin, M.D.
Stanford University Medical Center
Stanford, California 94305

Reference: C-1395

This white female infant was born at 39 weeks gestation, the first child of a 17 year old mother. The pregnancy was unremarkable apart from a brief febrile illness during the 4th month and the onset of albuminuria a few days prior to delivery. The child was born as a double footling breech after a 12 hour labor and was noted to be cyanotic. Convulsions developed soon after birth, which improved after suctioning and administration of oxygen. Blood sugar levels of 3 mg% and 9 mg% were recorded, and the infant was treated with intravenous dextrose and intramuscular Glucagon.

On physical examination, the patient was found to be markedly floppy, and the striking hypertonia permitted easy palpation of the abdominal viscera. Multiple congenital anomalies were found; notably, a prominently rotund face, low set ears, bilateral epicanthic folds, corneal clouding, bilateral simian creases, clubbed feet, and a protruding upper lip. There was moderate hepatosplenomegaly and the kidneys were apparently of normal size.

The clinical course over the next 5 days was marked by transient hypoglycemia, and an inability to feed, requiring gavage feeding. On the 4th day of life, the patient developed an episode of gastrointestinal bleeding and a platelet count of 87,000 with a normal prothrombin time was found. This failed to respond to therapeutic measures and the patient died after five days of life.

General Pathology: The foramen ovale was patent, and the pulmonary artery gave rise to the descending aorta. The ascending aorta continued as the innominate artery and terminated as the left subclavian artery. The liver showed some patchy fibrosis, and subcapsular and medullary cysts were seen in the kidney.

CNS Pathology: The brain weighed 370 grams and was externally unremarkable except for some slight thickening of some of the temporal gyri. Coronal sections showed focal blurring of the normal cortico-medullary junction. A small sub-ependymal cyst measuring 4 mm. in diameter was present in the left caudate nucleus.

Microscopic Pathology: Two slides, stained with Hematoxylin-eosin and Oil Red O.

Points for Discussion:

1. Diagnosis.
2. Pathological features of the disease.
3. Etiology and pathogenesis.