

Case #4

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This 6-year old girl had been in good health until the age of 2 years at which time, she developed back pain accompanied by stooping and guarding with limitation of motion of the spine. She had "spasms" which caused her to walk in a peculiar position and held her head turned to the right. These were thought to be seizures and she was treated with phenobarbital with some improvement. At age 4-1/2, she became irritable, complained of a sore back, walked with a stiff posture and had two episodes of urinary incontinence. There were no focal neurological findings. Spine films showed straightening of the lumbar spine with no bony abnormality; myelography showed a block at T-5 extending to T-2. A laminectomy was performed. The spinal cord was enlarged and a dorsal myelotomy was performed. A second stage operation was performed ten days after at which time, the tumor was found herniating through the myelotomy incision with a definite cleavage plane between the tumor tissue and normal cord structures. Biopsies taken at both operations appeared to be astrocytoma. The patient did well for the next 1-1/2 years except for slight ataxia and a left Babinski sign.

She complained of knee pains, headaches and occasional vomiting. These symptoms increased in intensity and she was readmitted to the hospital.

At that time, she was alert, with normal motor function decreased touch sensation in the left leg and an equivocal left Babinski sign. Lumbar spinal puncture showed normal pressure, CSF protein of 390 mg.% and no cells. Myelography was said to be normal except for a possible defect at the caudal end of the dural sac. Pneumoencephalography showed moderate ventricular enlargement. Several hours following pneumoencephalography, the patient developed severe headache, nausea and vomiting followed by confusion, opisthotonic posturing and decerebrate rigidity. The patient was treated with Decadron, mannitol and ventricular drainage but died the following day.

Autopsy findings: Acute tubular necrosis of kidneys and bilateral patchy emphysema of the lungs.

Neuropathological findings: The brain was swollen and weighed 1460 Gm. The unci had herniated into the incisura of the tentorium, bilaterally. The leptomeninges at the base of the brain were infiltrated by soft, gray tumor which was particularly abundant in the region of the cisterna magna, the interpeduncular fossa and about the midbrain and pons. No tumor was found in the parenchyma of the brain. The spinal cord showed some opacification of the leptomeninges overlying the spinal cord segments from approximately C-8 to T-5. Transverse sections of the spinal cord at these levels showed much of the tumor to be firm, white and rubbery; a portion of the tumor, dorsally, was soft and gelatinous. Sections of the upper thoracic spinal cord and hypothalamic region (H & E stains) are submitted.

Points for Discussion:

Nature of the neoplasm.