CASE 4

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A previously healthy three-year-old male showed no neurologic signs when first examined 3 months after the onset of progressively severe headaches, vomiting and personality changes. The skull x-ray showed separation of sutures and a right retrobrachial arteriogram and a ventriculogram revealed slight hydrocephalus. The lumbar spinal fluid was xanthochromic; it contained 400 mg.% protein, and 50-90 cells (80% lymphs 20% polys). Symptoms continued despite insertion of a ventriculo-peritoneal shunt. Six months after onset of symptoms myelography showed an almost complete block at T-12. At laminectomy tumor was found widely infiltrating the subarachnoid space around the spinal cord and cauda equina. The tumor biopsy was composed of many small cells with scanty cytoplasm which were of uniform appearance and randomly distributed. The patient's neurological condition deteriorated. He developed lower cranial nerve palsies, required tube feeding and tracheostomy and died eight months after the onset of the illness.

At autopsy no tumor was found grossly or microscopically outside the central nervous system. The eyes were not examined.

Gross examination of the brain and cord showed a tumor infiltrating the subarachnoid space around the cord and along the ventral surface of the brain-stem. There were patches of grey-black discoloration at the tips of both temporal horns, along the medial surfaces of both occipital lobes, in the striate pons, and outlining both dentate nuclei. All four ventricles were dilated.

The brown pigment within the tumor cells is bleached by permanganate, stains positively with Lillie's ferrous iron technique and with Bodian's silver protargol, and stains negatively with the Prussian blue reaction.

Diagnosis: Primary melanomatosis of leptomeninges with parenchymatous involvement.

Submitted are: Two 2 x 2 transparencies, 1 H&E stained section of medulla, and 1 unstained section of rostral pons.

Points of discussion:

Origin of tumor

- Primary vs. metastatic
- Multifocal or unicentric within CNS