CASE 3

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The patient, an 8 year old girl, was well until 15 months before death, when she developed a left foot drop. Over the next several months she complained of headache and vomiting, developed diplopia, slurred speech and right arm weakness. Her first admission was precipitated by a decreasing level of consciousness. Clinical examination and diagnostic studies suggested a fourth ventricle tumor, but surgical exploration of the posterior fossa failed to yield a diagnosis. Subsequently a ventriculo-atrial shunt was carried out for developing hydrocephalus. Although some neurologic deficit persisted, she showed no progression until two months before death, when her level of consciousness again deteriorated. At that time, the cerebrospinal fluid (CSF) protein was 1700 milligrams per cent, but the CSF pressure was normal. There were two large areas of increased uptake on the Technetium 99 brain scan. One focus in the left frontal lobe and another in the right temporal region. An attempt at ventricular puncture yielded 45 cc. of "oily" fluid. She continued to deteriorate and died.

The general autopsy showed only bronchopneumonia. No visceral neoplasm was found. Examination of the brain revealed a diffuse leptomeningeal neoplasm over both hemispheres, at the base, over the cerebellum and surrounding almost the entire spinal cord. In some areas the tumor was densely adherent to the brain and invaded it. Coronal sections through the brain showed the tumor in the leptomeninges to be white and quite firm. The parenchymal nodules were multiloculated in appearance, soft, gray, and appeared grossly to be mucinous. Nodules were found in all areas of the brain and spinal cord. No single area could be identified as a primary site.

Submitted are 2 gross Kodachrome transparencies, 1 slide stained with H and E, and 1 unstained slide.

The section submitted is representative of both the parenchymal and meningeal areas of the tumor.

Point for discussion: What is the histogenetic nature of this neoplasm? Where did it arise?