

CASE 3

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This 2-1/2 year old male was first seen at the age of 16 months with a three week history of irritability, lethargy and unsteadiness in walking. Examination disclosed a right VI, VII and IX cranial nerve palsy, limb and truncal ataxia, left hemiparesis and bilateral Babinski signs. He was given 3400 r for a presumed intra-axial glioma, with considerable improvement. A year later, however, the head was enlarged and there was early papilledema and mild left facial and left leg weakness. Plantar responses were flexor and no cerebellar signs were elicited. A right ventriculopleural shunt was performed. The downhill course was aggravated by renal, skin and lung infections. The patient died 13 months after the onset of symptoms.

At autopsy an irregularly lobulated, pale grayish mass measuring about 6.0 x 7.0 x 8.0 cm., was found ventral to the pons and upper medulla and in the right cerebellopontine angle. It was embedded in the right cerebellar hemisphere; pons and medulla which were greatly compressed, deformed and displaced, as was the IVth ventricle. The mass was sharply demarcated from the brain stem but was not separable from the cerebellar hemisphere. The tentorium was elevated. On section portions of the tumor were smooth and pale grayish, while other parts were more granular, mottled, softer and tan. There were several cysts, ranging up to 1.5 cm., which contained light greenish-gray, gelatinous material. Many small nodules of cream colored tissue were present in the leptomeninges over the dorsal surface of the spinal cord.