

## INFANTILE SPINAL CORD TUMOR PRESENTING AS HYDROCEPHALUS

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Although relatively rare, the association of spinal cord tumors with hydrocephalus and increased intracranial pressure, normal pressure hydrocephalus and benign intracranial hypertension is a well-recognized phenomenon.<sup>1</sup> Usually, the hydrocephalus becomes apparent or is discovered after the diagnosis of an intraspinal tumor.<sup>1,2</sup> In rarer circumstances, hydrocephalus is the presenting feature and evidence of spinal cord dysfunction appears subsequently.<sup>3-6</sup> In this report, we present the case of an infant who was treated for communicating hydrocephalus of undetermined etiology. Eleven months later, a malignant astrocytoma of the cervical spinal cord was diagnosed.

### Case Report

A one-year-old male infant presented with drowsiness and a progressive strabismus of one month's duration. Upon examination, severe bilateral papilloedema and bilateral 6th nerve palsies were noted. The child was unable to walk without help, although he had been able to do so one month earlier. Motor power and deep tendon reflexes were normal. CT scan of the brain showed hydrocephalus involving the whole ventricular system. The region of the foramen magnum and the craniovertebral junction were normal. Thus, since there was no evidence of obstruction within the intracranial cerebral spinal fluid (CSF) pathways, a diagnosis of idiopathic communicating hydrocephalus was made.

A ventriculoperitoneal shunt was inserted, after which the patient's clinical condition rapidly improved. The CSF was clear and colorless, and had no abnormalities on laboratory examination. The drowsiness and strabismus were immediately relieved and the patient was discharged within several days. He remained well at follow-up, and

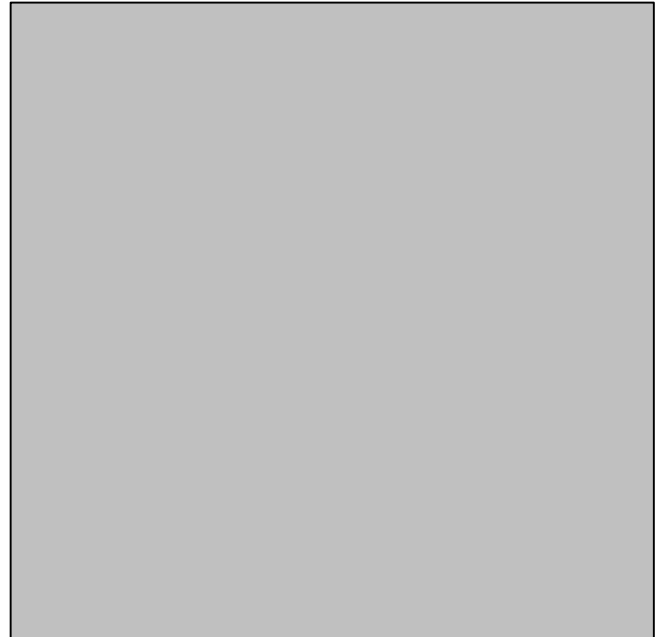


FIGURE 1. Sagittal gadolinium-enhanced T1-weighted MRI of the cervical spine showing an intramedullary tumor extending from the medulla to C6.

when examined at the age of 18 months, he was walking independently without neurological abnormality. At the age of 23 months, his parents noted clumsiness of his right upper limb.

Examination revealed no clinical evidence of increased intracranial pressure. The right deltoid and bicep muscles were weak and atrophic. In addition, the right triceps and the small muscles of the right hand were also weak, though to a much lesser degree. The right biceps and brachioradialus reflexes were absent. Tone in the lower limbs was increased, as were the deep tendon reflexes. Bilateral extensor plantar responses were present. A CT scan of the brain showed normal-sized ventricles with the shunt catheter in place. Magnetic resonance imaging (MRI) of the cervical spinal cord showed an intramedullary tumor, which extended from the lower medulla to the C6 region (Figure 1).

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## Discussion

In a review of 87 reported cases published in 1992, Bland and McDonald emphasized the rarity of the occurrence of hydrocephalus in association with spinal cord tumors.<sup>1</sup> They found that the tumor was intramedullary in 60 reported cases, and that in 27 of these (45%), the tumor was malignant. They also noted that the association of hydrocephalus with benign extramedullary tumors was even more uncommon, with only 27 documented cases.

The hydrocephalus is usually discovered after the spinal cord tumor has been recognized.<sup>3-6</sup> In a single institution series of 171 patients with intramedullary spinal cord tumors, Rifkinson-Mann et al. found 25 subjects with symptomatic hydrocephalus.<sup>7</sup> In five of these, hydrocephalus was the presenting feature, and shunt procedures were carried out. In four, the spinal cord tumor was diagnosed one month later, and in the other patient, after six months. However, it should be noted that most of the patients in this series had varying degrees of tumor resection as well as radiotherapy. Also, 62% had radiological or autopsy evidence of leptomeningeal metastatic disease.

The case reported by Caveness et al.<sup>8</sup> is similar to ours. Their patient was a child who presented at four months of age with hydrocephalus of undetermined etiology. A shunt was inserted and the symptoms resolved. However, signs of spinal cord dysfunction appeared at age nine months, and a spinal cord astrocytoma was diagnosed. These authors suggest that hydrocephalus in infants with spinal cord tumors may occur because of a disruption of cerebrospinal fluid flow during development of the neural tube. This is a similar process whereby hydrocephalus develops in infants with myelomeningocele.<sup>9</sup>

The pathogenesis of increased intracranial pressure or hydrocephalus that occurs in association with spinal cord tumors remains speculative. A variety of mechanisms have been suggested, the principal one being the impaired absorption of CSF, the causes of which include elevated protein,<sup>5</sup> basal arachnoiditis due to tumor infiltration into the basal cisterns,<sup>7</sup> and compression of the venous spinal plexus by tumor.<sup>1</sup> Kudo et al.<sup>5</sup> reviewed the mechanisms whereby elevation of CSF protein may lead to hydrocephalus. They pointed out that an increase in protein content causes an increase in CSF viscosity, thereby decreasing its absorption. In addition, the protein molecules may cause mechanical clogging of the semi-permeable membrane through which the protein and water of the CSF are absorbed. However, they also noted that in other reported cases, the CSF protein content was not increased. Moreover, most cases of spinal cord tumor with increased

CSF protein do not have hydrocephalus.<sup>8</sup> Similar arguments and counterarguments exist regarding the presence or absence of tumor cells in the CSF.<sup>7</sup> Caveness et al.<sup>8</sup> called attention to the association of hydrocephalus with benign spinal cord tumors. In such cases, an increased CSF cell count or tumor seeding into the basal cisterns are unlikely to be the cause of CSF obstruction. Obviously, when tumors of the cervical spinal cord extend in a rostral direction to the cervicomedullary junction, the CSF pathways may be obstructed and hydrocephalus may result.<sup>7</sup> In our patient, the tumor that was eventually diagnosed was indeed in this location, although the CT scan on presentation showed no evidence of a craniospinal lesion, and the child was well for six months after shunting. Interestingly, a literature review of 65 cases of spinal cord tumor and hydrocephalus showed the most common location for the tumor to be the thoracolumbar region.<sup>5</sup>

The case reported here illustrates the inherent danger in CSF shunting when the exact etiology of the hydrocephalus is unclear. The therapeutic implications are so important that every effort should be made to determine an underlying cause. We suggest that patients with hydrocephalus or increased intracranial pressure without a clearcut intracranial etiology should have an MRI scan that includes the whole spine, with special attention to the craniocervical region. This is particularly appropriate when the patient is an infant in the early stages of development. In this situation, even if the spinal MRI is initially negative, the child should be closely followed for signs of a developing spinal cord tumor.

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