CASE REPORT

James A. Filkins,[1] M.D., Ph.D.; Stephen Cohle,[2] M.D.; Beth K. Levy,[3] M.D.; and Michael Graham,[3] M.D.

Unexpected Deaths Due to Colloid Cysts of the Third Ventricle

REFERENCE: Filkins, J. A., Cohle, S., Levy, B. K., and Graham, M., **"Unexpected Deaths Due to Colloid Cysts of the Third Ventricle,"** *Journal of Forensic Sciences,* JFSCA, Vol. 41, No. 3, May 1996, pp. 521–523.

ABSTRACT: Colloid cysts of the third ventricle are rare central nervous system tumors that are a recognized cause of unexpected death in young, otherwise healthy adults and children. We report three adults and one child who died from colloid cysts of the third ventricle. Our report illustrates the difficulties of diagnosing these tumors premortem.

KEYWORDS: forensic science, forensic pathology, colloid cysts, brain tumors, forensic neuropathology, sudden death

Unexpected death due to primary neoplasms of the central nervous system is a recognized, if uncommon, occurence in medicolegal practice (1). Of the various central nervous system tumors that can produce unexpected death, colloid cysts of the third ventricle are unique. They are benign neoplasms that are amenable to uncomplicated surgical removal. These tumors may produce vague symptoms for years before abruptly causing catastrophic neurological deterioration and death. Since patients who die from colloid cysts do not always survive long enough to be taken to a hospital, or they die before the diagnosis is established, they may come to the attention of medical examiners as well as to hospital pathologists. The number of cases reported in the forensic literature is few (2) in comparison to those published in the neurosurgical or radiologic literature. In particular, reports of unexpected deaths in children due to colloid cysts are rare (3).

Case Reports

We report three adults and one child who died unexpectedly as a result of colloid cysts of the third ventricle. All of our patients were female and each presented with a severe headache as part of her terminal episode. Our findings are summarized in Tables 1 through 3. Also, see Fig. 1.

[1]Office of the Medical Examiner, County of Cook, Chicago, IL.
[2]Blodgett Memorial Medical Center, Grand Rapids, MI.
[3]Department of Pathology, St. Louis University School of Medicine, St. Louis, MO.

Received for publication 10 July 1995; revised manuscript received 11 Sept. 1995; accepted for publication 16 Oct. 1995.

Discussion

Colloid cysts of the third ventricle were first described by Wallman in 1858. The histological origin of these tumors remains uncertain as do the developmental conditions which produce them and the factors that stimulate their growth. The tumor consists of a fibrous capsule surrounding a mucin-filled cavity. Most often located in the anterior portion of the third ventricle the tumor may float loosely, attached only by a stalk, or completely occlude the ventricle. Microscopically, the cysts are lined by cuboidal or columnar epithelium with goblet cells and cilia.

Colloid cysts comprise less than one percent of intracranial tumors, occur at any age, and afflict males and females equally. In his monograph on colloid cysts, Walter E. Dandy described what has come to be known as the classical presentation of severe, paroxysmal headaches relieved by changes in head position (4). Subsequent investigators have seldom encountered Dandy's classical presentation. Instead, a headache without the positional component emphasized by Dandy seems to be the most common and most significant presenting symptom. Other common symptoms include nausea and vomiting, gait disturbances, mental status changes, seizures, and incontinence. Papilledema is the most common sign found during a neurological examination (5).

Since the advent of computerized axial tomography and magnetic resonance imaging colloid cysts have been diagnosed with greater frequency. Typically, the cyst will appear on a CT scan as a hyperdense or isodense mass at the level of the Foramen of Monro with enlargement of the septum pellucidum, collapse of the posterior portion of the third ventricle and dilation of the lateral ventricles (6). MRI imaging shows an increase in the T1 signal.

Dandy performed the first successful surgical excision of a colloid cyst of the third ventricle in 1921. Surgery remains the most common method of treatment, but in addition to traditional transcortical or transcallosal approaches, shunt placement and stereotactic aspiration are also employed. If the tumor is small enough, the patient asymptomatic, and there is no evidence of intracranial pathology other than the tumor on imaging studies, treatment may be deferred (7).

The attachment of some colloid cysts by a stalk to the anterior portion of the third ventricle suggested to Dandy and others that the tumor produced its symptoms by a ballvalve action, which intermittently obstructed the flow of cerebrospinal fluid. This was thought to explain why changes in head position produced relief. Eventually, the tumor would grow large enough to block permanently the Foramen of Monro. Then acute hydrocephalus, herniation and death would follow.

TABLE 1—*Headache history.*

	Age at death	Onset	Location	Pattern	Frequency	Duration	Associated signs and symptoms	Treatment	Releif with change in head position
Patient 1	13	3 mos. prior to death	fronto-occipital, then nonlocalizing	Intermittent, progressively more severe	Six week intervals	3 days	Lethargy, double vision	Aspirin, propoxyphene	No
Patient 2	30	2 days prior to death	nonlocalizing	Intermittent, progressively more severe	Unknown	2 days	Unknown	None	No
Patient 3	33	6 years prior to death	bilateral, progressing to frontal	Severe, "like a steel band"	3 year intervals	1 week	Confusion, photophobia, loss of consciousness	Acetaminophen, acetaminophen/isomethептene mucate/dichloralphenazone	Yes
Patient 4	26	Unknown*	nonlocalizing	Intermittent, severe	Unknown	2 hours	Unknown	Acetaminophen	No

*Although severe headaches were reported prior to the terminal episode.

TABLE 2—*Terminal episode.*

	Presentation	CT scan results	Surgical intervention
Patient 1	Severe, nonlocalizing headaches Nausea and vomiting	Massive hydrocephalus Lesion in 3rd ventricle	Bilateral ventriculostomies
Patient 2	Intermittent, progressively severe headaches Chills and vomiting	N/A	N/A
Patient 3	Severe headaches	Normal*	N/A
Patient 4	Severe headaches with neck pain Nausea and vomiting Lethargy, confusion deteriorating to coma	Severe hydrocephalus Hyperdense supersellar mass	Right frontal ventriculostomy

*This CT scan was done three years prior to death as part of a neurological evaluation for similar headaches. At an earlier neurological evaluation, six years prior to death, no CT scan was done.

More recent studies have noted that the tumors often completely fill the third ventricle below the level of the Foramen of Monro with reactive changes in the surrounding periventricular tissue indicating the chronicity of the tumor. The impaction obstructs cerebrospinal fluid flow which in turn elevates intracranial pressure and compromises the mechanisms by which variations in intracranial pressure are regulated. Acute variations in ventricular filling pressure known as plateau waves develop which cause the transitory symptoms of a colloid cyst and, if unrelieved, lead to permanent deterioration or death. Increased cerebral blood volume, decreased cerebral blood flow, impaired autoregulation, edema, and herniation result (8). In those instances where death occurs in the absence of herniation or significant hydrocephalus, such as in our Case 4, disturbance of neurally mediated cardiovascular regulation in the hypothalamus may be responsible (9).

Colloid cysts of the third ventricle have long been known to cause sudden and unexpected death in otherwise healthy children and adults. In large part, the reason for the high mortality of these tumors despite their ease of resection is the difficulty in establishing the diagnosis in a timely fashion. Only one of our patients presented with Dandy's classical presentation of a severe headache relieved by changes in head position. She was the only one of our decedents who reported mental status changes and loss of consciousness before the terminal episode. Her headaches occurred over the longest period of time in our series, six years. Two previous evaluations for headaches failed to demonstrate any anatomic abnormality. Her second evaluation included a CT scan of the brain that was interpreted as normal illustrating that present day imaging techniques are not always diagnostic, especially in the early stages of the tumor's development.

Our other patients reported headaches occurring over two days to three months. Their headaches varied in severity and response to analgesic measures. The only other associated symptoms were nausea and vomiting, and double vision. In these three patients mental status changes and neurological deficits occurred only during the terminal sequence of events. Autopsies confirmed a colloid cyst occluding the third ventricle in each case, with herniation and edema visible in three patients, and the microscopic findings of brain death in the fourth. Our report illustrates that even after seventy years of experience with these tumors and the availability of sophisticated imaging techniques the diagnosis of a colloid cyst can still remain difficult. Although rare, colloid cysts of the third ventricle are a continuing source of unexpected death with which medical examiners and hospital pathologists should be familiar.

Acknowledgments

The authors wish to thank Eldon Cassell, M.D. for the submission of Case 2.

TABLE 3—*Postmortem findings.*

	Colloid cyst of 3rd ventricle	Cerebral edema	Ventricular dilatation	Uncal grooving	Brain stem compression	Cerebellar tonsillar herniation	Cingulate gyrus herniation
Patient 1	Yes	Yes	Yes*	Yes	No	No	Yes
Patient 2	Yes	Yes	Yes	Yes	No	Yes	No
Patient 3	Yes	Yes	Yes	Yes	Yes	Yes	No
Patient 4	Yes†	No	No	No	No	No	No

*Dilatation of the left lateral ventricle; compression of the right lateral ventricle.
†There was no gross evidence of dilatation or herniation, but the microscopic findings of brain death were present.

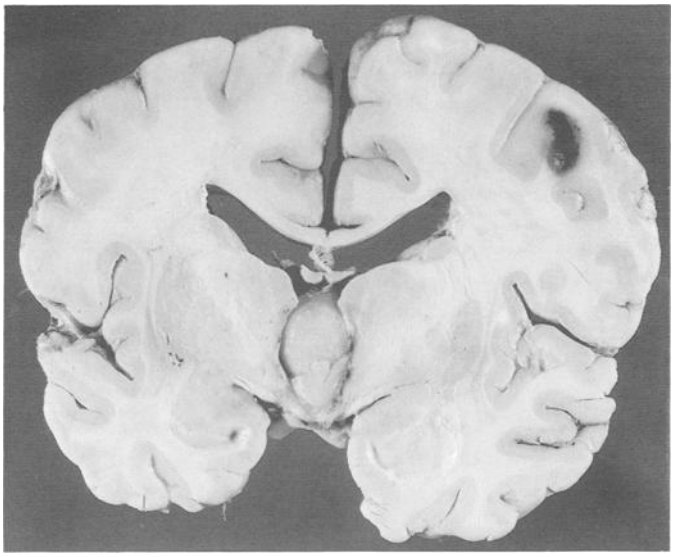

FIG. 1—*A colloid cyst occluding the third ventricle from Case 4. (The hemorrhage in the right frontal lobe is from the ventriculostomy.)*

References

(1) DiMaio T, DiMaio D. Sudden death due to colloid cysts of the third ventricle. NY State J Med 1974;1832.
(2) Leestma JE, Konakci Y. Sudden unexpected death caused by neuroepithelial (colloid) cyst of the third ventricle. J Forensic Sci 1981;26(3):486–91.
(3) Byard RW, Moore L. Sudden and unexpected death in childhood due to a colloid cyst of the third ventricle. J Forensic Sci 1993;38(1):210–13.
(4) Dandy WE. Benign tumors of the 3rd ventricle of the brain. London: Ballie, Tindall and Cox, 1933.
(5) Little JR, MacCarty CS. Colloid cysts of the third ventricle. J Neurosurgery 1974;39:230–35.
(6) Antunes JL, Louis KM, Ganti BR. Colloid cysts of the third ventricle. Neurosurgery 1980;7(5):450–55.
(7) Camacho A, Abernathy CD, Kelly PJ, Low ER. Colloid cysts: experience with the management of 84 cases since the introduction of computed tomagraphy. Neurosurgery 1989;24(5):693–99.
(8) Brun A, Egund N. The pathogenesis of cerebral symptoms in colloid cysts of the third ventricle: a clinical and anatomical study. Acta Neurol Scand 1973;49(4):532–34.
(9) Ryder JW, Kleinschmidt-DeMasters BK, Keller TS. Sudden deterioration and death in patients with benign tumors of the third ventricle area. J Neurosurgery 1986;64:216–23.

Address requests for reprints or additional information to
James A. Filkins, M.D., Ph.D.
Office of the Medical Examiner
County of Cook
2121 West Harrison St.
Chicago, IL 60612