CASE REPORT

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Sudden and Unexpected Death in Childhood Due to a Colloid Cyst of the Third Ventricle

REFERENCE: Byard, R. W. and Moore, L., **"Sudden and Unexpected Death in Childhood Due to a Colloid Cyst of the Third Ventricle,"** *Journal of Forensic Sciences,* JFSCA, Vol. 38, No. 1, January 1993, pp. 210–213.

ABSTRACT: A nine-year-old boy died suddenly and unexpectedly following a two day history of intermittent headaches. At autopsy a colloid cyst of the third ventricle was found that had obstructed the foramen of Monro and caused hydrocephalus with prominent cerebral edema. Colloid cysts are rare entities in childhood and are not usually included in the differential diagnosis of pediatric sudden death. This report describes the clinicopathological features of such a case.

KEYWORDS: pathology and biology, colloid cyst, sudden pediatric death

Intracranial causes of sudden and unexpected death in children are uncommon, and are often due to trauma, epilepsy, or to catastrophic hemorrhage associated with neoplasia or vascular malformations [*1–3*]. Intraventricular lesions that cause sudden and unexpected death are rare and are most often found in adults. The following case illustrates the clinicopathological features of a fatal, clinically unsuspected third ventricular colloid cyst in a nine-year-old boy. It is emphasized that this lesion must be considered as a cause of sudden pediatric death and that the antemortem presentation may be relatively nonspecific and of little help in formulating a diagnosis prior to performance of a full autopsy examination.

Case Report

A nine-year-old boy had complained of headaches on the two days prior to death and had been listless with one episode of vomiting on the day of death. He had slept in the afternoon and had later been found difficult to rouse. Death occurred soon after. His only significant past medical history was of mild asthma.

At autopsy, in addition to prominent pulmonary edema, there was marked cerebral edema with flattening of cerebral gyri and sulcal narrowing. The lateral ventricles were not compressed as expected, but were dilated and a 10 mm diameter unilocular cyst containing semitranslucent gelatinous material distending the third ventricle was identified (Fig. 1). Histologic examination of this lesion revealed features typical of a colloid cyst. The cyst had a thin wall composed of loose fibroconnective tissue and was lined on its

Received for publication 28 Jan. 1992; accepted for publication 20 May 1992.

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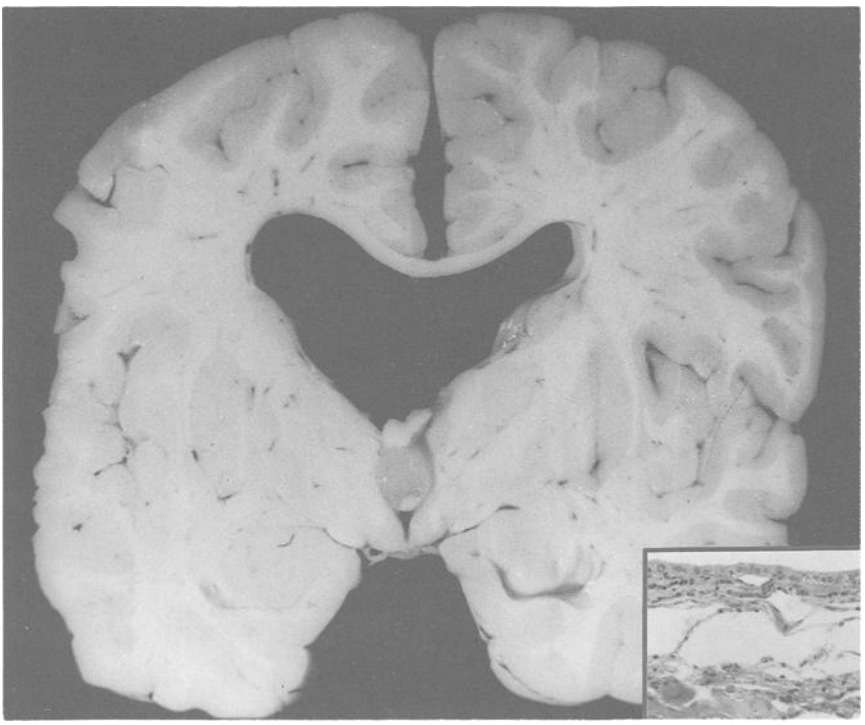

FIG. 1—*Coronal section of the brain demonstrating dilatation of the lateral ventricles with a 10 mm diameter colloid cyst within the third ventricle causing obstruction to the cerebrospinal fluid drainage pathway. Inset: Flattened cuboidal lining epithelium (Hematoxylin & Eosin × 600).*

inner aspect by a single layer of cuboidal cells surrounding periodic acid-Schiff diastase resistant material (Fig. 1 inset). The remainder of the autopsy was unremarkable and death was attributed to cerebral edema resulting from a third ventricular colloid cyst causing acute obstructive hydrocephalus with associated acute pulmonary edema.

Discussion

Central nervous system disorders that are associated with sudden and unexpected death in the pediatric age group are uncommon and, aside from trauma and epilepsy [*1,2*], often involve hemorrhage into a tumor or vascular malformation [*3–7*]. Very occasionally, sudden death can occur from a critically placed lesion, such as a colloid cyst of the third ventricle, which causes obstruction of cerebrospinal fluid drainage, with resultant acute hydrocephalus [*8*].

Colloid cysts are histologically benign cystic structures of uncertain derivation that arise within the third ventricle. Various theories have been suggested as to the tissue of origin of this entity with neuroepithelial derivation [*9,10*] from paraphyseal, ependymal or choroid plexus epithelium being favored by different authors [*11,12*]. Histologically, the cysts are unilocular structures containing eosinophilic debris lined by flattened cuboidal to columnar epithelium which may be ciliated [*13*].

The consistent localization of these cysts to the anterior portion of the third ventricle explains their clinical symptomatology [*14*]. Due to close proximity to the foramen of Monro, colloid cysts may cause obstruction to cerebrospinal fluid egress resulting in

severe headaches, collapse or rarely death [*15,16*]. Symptoms may be intermittent, as the pedunculated tumor may cause only temporary blockage of cerebrospinal fluid drainage with spontaneous relief of the acute hydrocephalus occurring with movement of the cyst [*16*].

The striking feature in the reported patient is the young age, as colloid cysts are most often found in adult life in the sixth decade, with Russell and Rubenstein describing the "paucity of examples in infancy and childhood" as "remarkable." In a review of 146 cases only two were found under ten years of age, and even those found incidentally have been in adults [*8*]. Only one other case of colloid cyst was found on review of the Department of Histopathology files at the Adelaide Children's Hospital over the 36 year period from 1962 to 1991. Although sudden death had also occurred in the second patient, a seven year old severely mentally retarded spastic quadraplegic boy, the history of epilepsy, and autopsy findings of significant gastric aspiration, made assessment of the role of the colloid cyst in the cause of death difficult to determine. As sudden death is a well recognized phenomenon in epileptic patients, due to a variety of mechanisms such as asphyxia, cardiac dysrhythmia or respiratory failure [*17*], epilepsy could not be excluded as the cause of death, and so the patient has not been described in detail in this report. Other reports of colloid cysts in children are found only infrequently in the literature [*18–21*].

In conclusion, the reported patient demonstrates the occurrence of sudden and unexpected death in a pediatric patient due to a large third ventricular colloid cyst. Although the cyst had caused obstructive hydrocephalus, the clinical presentation was brief and relatively nonspecific, emphasizing that these lesions may present unexpectedly at autopsy in quite young patients with minimal history.

Acknowledgments

We would like to acknowledge the secretarial help of Heather Cooke.

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