Fatal colloid cyst

Colloid cysts can result in sudden, unexpected and potentially lethal complications.

Mechanism

The mechanism(s) of death are still a controversial subject and several mechanisms have been postulated to explain the sudden onset of severe symptoms and of fatal rapid deterioration [1].

Postmortem findings of ventricular enlargement and brain herniation in fatal case of patients with colloid cyst of the third ventricle had led most authors to postulate that hydrocephalus may play a major role in fatal cases [2] [3] [4].

The accurate pathophysiologic lethal mechanism is not completely cleared. Moreover, it is interesting to note that neither the cyst size, nor the degree of ventricular dilatation appears to be a reliable predictor of outcome, as even small, asymptomatic cysts may result in sudden death [5] [6] [7].

Such findings suggest an alternative mechanism underlying the sudden death of patients with colloid cyst. As the hypothalamic structures which are involved in neuroendocrine and autonomic regulation playing a key role in cardiovascular control [8] [9] are located close to the walls of the third ventricle which is the most frequent anatomical site of colloid cyst, this may suggest that reflex cardiac effects due to the compression of the hypothalamic cardiovascular regulatory centers by the cyst explain the sudden death in patients harboring a colloid cysts when signs of hydrocephalus or brain herniation are lacking [10] [11].

Data suggests that the prodrome preceding sudden death appears shorter in adults with more adults presenting with symptoms for less than a day, and most colloid cysts measured one centimeter or larger [12].

Systematic Review

A systematic literature search of three popular databases was performed. Inclusion criteria were individuals with sudden-onset death and colloid cysts identified on imaging and/or autopsy. The cause of death must have been ascribed to the colloid cyst. Deaths precipitated by lumbar puncture were excluded. Clinical data were extracted and descriptive statistics were ascertained. Subgroup analyses were performed to compare adults to pediatric patients.

A total of 107 patients were included. The mean age was 28.13 years (SD 13.26, range 6-79 years). Males and females were equally affected. Headache was the most frequent symptom (n = 86, 80.37%). There were 6 patients (5 adults and 1 child) that reported positional headaches. The mean duration of signs and symptoms was shorter in adults versus pediatric patients (2.10 days vs 6.50 days, p = 0.02) and more adults presented with signs and symptoms for less than 24 hours than did pediatric patients (37.50% vs 5.56%, p = 0.01). Colloid cyst mean diameter was 2.00 cm (SD 1.07,

range 0.50-7.90 cm) and 95.71% measured 1 cm or larger.

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Cases

2015

A 45 year old male with hypertension with acute hydrocephalus due to a hemorrhagic colloid cyst in the third ventricle. An external ventricular drain on each side was introduced and he was admitted to the ICU, where brain death was certified. After removal of the cyst through transcortical frontal approach, the diagnosis of colloid cyst with remains of hemolyzed blood was confirmed.

Bleeding in colloid cysts is exceptional, and can occur in both symptomatic and asymptomatic patients, making it difficult to recognize this complication [14].

2012

One patient who was brought in an unconscious state, an external ventricular drain was inserted and she was ventilated. She died 4 hours after the admission [15].

In this case, macroscopic and histological findings addressed the diagnosis of colloid cyst of the third ventricle with diffuse myocardial injury (coagulative myocytolysis or contraction band necrosis, CBN) and led to conclude that acute cardiac arrest due to hypothalamus stimulation in the context of colloid cyst of the third ventricle was the cause of death. As the hypothalamic structures which are involved in neuroendocrine and autonomic regulation playing a key role in cardiovascular control are located close to the walls of the third ventricle which is the most frequent anatomical site of colloid cyst, this may suggest that reflex cardiac effects due to the compression of the hypothalamic cardiovascular regulatory centers by the cyst explain the sudden death in patients harboring a colloid cyst when signs of hydrocephalus or brain herniation are lacking [16].

Sudden death in a patient with a third ventricle colloid cyst [17].

2009

Three cases of sudden death resulting from colloid cysts of the third ventricle are presented. The first and second cases were treated for migraine headaches. In the first case, the patient was a 24-year-old woman who presented to the hospital with a severe headache and was sent back home after medical treatment. Six hours later, she was found dead in her bed. The second case was a 21-year-old woman who experienced a severe headache, dizziness and vomiting 1 day prior to her death. She was

transported to the hospital, where she was pronounced dead upon arrival. The third case was a 25-year-old man who experienced headaches and vomiting and was diagnosed with and medically treated for sinusitis. He lost consciousness and was taken to hospital, where he was pronounced dead on arrival. During the autopsy of all three cases, there was a grey transillumination area observed that occurred due to the stretching of tissue at the base of brain between the corpus mamillare and chiasma opticum. Dissection of the brain revealed a colloid cyst of the third ventricle. To avoid such fatal complications, prompt diagnosis using CT or MRI is essential in patients who have a long-standing history of intermittent headaches. During the autopsy of the sudden deaths of people with medical antecedents of headaches, if a grey color is observed between the chiasma opticum and the corpus mamillare in the base of the brain, a colloid cyst should be considered and this region should be dissected and examined carefully [18)].

A 32 years old omani woman G6P4A1 at 39 weeks of pregnancy presented with complaints of headache and picture of eclampsia. Emergency caesarean section was done for maternal and foetal distress. Post caesarean section patient could not be extubated from general anaesthesia. Emergency computed tomography scan of the brain was done which revealed colloid cyst of the third ventricle with severe acute obstructive hydrocephalus leading to irreversible brain damage and death. Colloid cyst of the third ventricle is a rare benign intracranial tumour, and pregnancy often masks the symptoms. High index of suspicion and early detection of tumour may be life saving [19)].

2005

An 18-year-old girl was found dead in her bed. The autopsy revealed a colloid cyst of the third ventricle. The cyst obstructed the flow of cerebrospinal fluid, leading to prominent internal hydrocephalus with consecutive brain edema and compression of the caudal medulla at the foramen magnum. The girl's only previous complaints were episodic headaches for the previous 2-3 years. Computed tomography and magnetic resonance imaging were not performed prior to her death. This case report highlights the importance of early diagnosis of colloid cyst of the third ventricle and the need to perform computed tomography and magnetic resonance imaging in patients with episodic headaches even when they show no neurologic deficit. Although it is a very rare disorder, it should be included in the differential diagnosis of headaches in children and young adults, and also in the differential diagnosis of conditions causing increased intracranial pressure, in view of the life-saving management required to prevent a fatal outcome [20)].

A 24 year old pregnant woman with familial colloid cyst, who presented with headaches and suffered a cardiorespiratory arrest [21)].

2003

A ten-year-old male child who presented with sudden neurological deterioration due to colloid cyst of the third ventricle resulting in death. The child had intermittent headache for three months, for which medical attention was not sought [22)].

2002

Four patients died suddenly and the cysts were discovered at autopsy. The overall mortality rate was 12%. Results of a multivariate logistic regression analysis demonstrated that no subgroup of patients presenting without acute deterioration could be identified on the basis of patient age, duration of symptoms, cyst size, or the presence of hydrocephalus. The national incidence of colloid cysts in The Netherlands is 1/10(6) person-years; the prevalence was estimated to be 1800 asymptomatic colloid cysts.

Acute deterioration was a frequent presentation among a national cohort of Dutch patients harboring symptomatic colloid cysts. The risk of acute deterioration in a symptomatic patient with a colloid cyst in The Netherlands is estimated to be 34%. The estimated risk for an asymptomatic patient with an incidental colloid cyst is significantly lower. These results strongly advocate the selection of surgical treatment for patients with symptomatic colloid cysts [23)]

2000

Two cases with fatal outcome. Autopsy revealed presence of colloid cyst in the third ventricle in both cases. It is proposed that to avoid fatal outcome in a patient reporting with frequent attacks of headache, the presence of colloid cyst in the third ventricle should be considered in differential diagnosis. In such cases, CT scan or MRI study of brain becomes an essential diagnostic tool [24)].

1999

A 25-year-old man presented with a 24-h history of headache and vomiting. He rapidly became unconscious and fulfilled the criteria for brain death on arrival at hospital. No surgical intervention was performed.

The patient's sister presented at the age of 41 with headaches and rapidly became unconscious. The sister had urgent bilateral ventriculostomies. followed by transcallosal removal of a colloid cyst.

These cases support the hypothesis that colloid cysts are congenital lesions and provide some evidence of a possible genetic predisposition to their formation. Sudden death remains a real risk for patients harbouring a colloid cyst [25)].

1997

Two cases of sudden death due to colloid cysts of the third ventricle are presented with a review of the literature. In the first case, a 40-year-old woman suffered an acute onset of severe frontal headache after an intercontinental air flight. The next day, she was found comatous and died 7 h after admission to a hospital. In the second case, a 33-year-old man with a medical history of recurrent headaches was found dead in his car. Autopsy in both cases revealed a colloid cyst of the third ventricle and hydrocephalus involving the lateral ventricles. These cases demonstrate that fatal cases still occur. Nevertheless, prompt diagnosis using computed tomography (CT) or magnetic resonance imaging (MRI) is essential, since colloid cysts are histologically benign tumors that can be removed safely by neurosurgical intervention [26)].

Thirty-seven consecutive patients with colloid cyst of the third ventricle seen at Karolinska Hospital between 1984 and 1995 were reviewed. Five patients were admitted in a comatose state, and two died despite emergency ventriculostomy. Three had recurrent cysts following previous aspiration procedure. During the study period, patients underwent a total of 10 ventriculostomies, 10 aspirations, 26 microsurgical operations, and two shunt operations. Twenty-four of 26 microsurgical operations were transcallosal and two were transcortical. Twenty-four operations (22 transcallosal and two transfrontal approaches) without permanent morbidity were performed by four surgeons. Transient memory deficit from forniceal traction was noted in 26%. The remaining two transcallosal operations, which led to permanent morbidity or mortality, were performed by two different surgeons. Aspiration of cysts performed by four different surgeons carried a 40% risk of transient memory deficit (10% permanent) and an 80% recurrence rate. One patient was found to be cured on radiological studies obtained at the 5-year follow-up review. Seven cysts were followed by means of radiological studies with no treatment for 6 to 37 months. Five of these cysts grew, indicating that younger patients with colloid cysts will probably need surgical treatment. The main causes of unfavorable results were: 1) failure to investigate symptoms that proved fatal; 2) subtotal resection; and 3) surgical complications. Transcallosal microsurgery produced excellent results when performed by experienced surgeons. A colloid cyst of the foramen of Monro is a disease that should be detected before permanent neurological damage has occurred. Permanent morbidity or mortality should not be accepted in modern series of third ventricle colloid cysts [27)].

A case report describes death in a young male, six months after an assault. The death was caused by a colloid cyst, a rare but important malformation in the brain. The possible relationship between the assault and the cyst is discussed [28)].

1995

In three of five cases of third ventricular colloid cyst the patients presented with or developed severe headache which was like no other they had experienced before. The headache was of long enough duration to enable detection and removal of the cyst before its fatal outcome. In one case involving a fight it was considered that the third vetricular colloid cyst played an important part in the fatal outcome. In the fifth case the patient was found dead in bed without any preceding history of illness [29)].

1993

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 [30)].

1992

Five cases of colloid cysts of the third ventricle discovered only at autopsy are presented. In 3 cases, colloid cysts were identified as the actual cause of death; clinical symptoms had been unspecific (mostly intermittent headaches) and of varying duration. The pathogenesis of “sudden death” as it may occur in some of these benign, dysembryogenic neoplasms, is still not fully understood. Neither the size of the cyst and the degree of ventricular dilatation, nor the duration of clinical symptoms, seem to provide reliable prognostic indicators of this potentially fatal complication. That an extended indication for surgery might be advisable even in cases of asymptomatic colloid cysts without hydrocephalus, as advocated by some authors, is considered in the discussion [31)].

1987

A 26 year-old man with a colloid cyst of the third ventricle (paraphyseal cyst; neuroepithelial cyst), which gave rise to a fatal increase in intracranial pressure. Despite repeated hospitalisation the diagnosis was not established prior to death. This report serves as a reminder that this rare disorder must be considered in the differential diagnosis of conditions causing increased intracranial pressure in view of the life-saving therapeutic management required to avert a fatal outcome [32)].

1981

Four cases of sudden unexpected death in young persons whose only major previous complaint was that the chronic relapsing headaches. Because of the sudden and unexpected nature of their deaths, these cases came to the attention of the medical examiner and the colloid cysts were discovered at autopsy. The history of the lesion, its histogenesis, its pathological physiology, and the experiences of others are reviewed [33)].

1957

Colloid cyst of the third ventricle in flyers; report of three fatal cases [34)].

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