

## Case Report

# Sudden death due to intracranial colloid cyst: About three cases

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The colloid cyst is a rare and benign congenital intracranial tumor, commonly located in the anterior part of the third ventricle. It is a rare cause of sudden death whose mechanisms of occurrence are still been discussed. We report three cases of sudden death due to colloid cyst(s) discovered at autopsy. Death occurred in a man and two women, aged 45, 43 and 33 years, respectively. Pre-mortem symptoms were mainly headache and/or syncope. The autopsy revealed the presence of colloid cyst(s) of the anterior part of the third ventricle, confirmed by a histological examination. A cerebral edema was found in two cases, and hydrocephalus in one case. We raise the clinical and pathological features of this tumor, and we discuss the mechanisms of death.

**Key words:** Colloid cyst, sudden death, autopsy.

## INTRODUCTION

Sudden death represents a dramatic situation for both the physician and the society. It accounts for 21% of the post mortem activity of the Department of Forensic Medicine of Sousse (Tunisia), causing an average of 73 deaths per year. Ten percent of these deaths are due to neurological affections; in particular bleedings, meningoencephalitis, brain infections, epilepsy and tumors including the colloid cyst. In the literature, sudden death caused by brain tumors represents 0.16 to 3.2% of all causes of sudden death (Büttner et al., 1999). Colloid cyst is a benign tumor developed from embryonic remnants, in the anterior part of the third ventricle (Turillazzi et al., 2012). Its involvement in sudden death is rare and related to controversial mechanisms (Bender et al., 2013). We stress the clinical and pathological features of this tumor through the report of three cases of sudden death due to colloid cyst.

## CASE REPORTS

### Case 1

A 45-year-old man, with no personal medical history, died suddenly after fainting during his work as a mason. At autopsy, we found two cystic formations of the third ventricle measuring 1.5 cm in diameter each (Figure 1), accompanied by a moderate cerebral edema and without dilatation of the ventricular system. Body cavities were intact, with no other pathological findings except for pulmonary edema. The anatomo-pathological examination of the cysts concluded in their colloid nature.

### Case 2

A 43-year-old woman complained about an isolated and

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**Figure 1.** Two colloid cysts.

intermittent headache, which lasted three months before her death. She died immediately after an intense crisis of headaches. At autopsy, we found a cyst of the anterior part of the third ventricle measuring 2.5 cm, accompanied by a cerebral edema and a slight dilation of the ventricular system. Pathological examinations concluded at the colloid nature of the cyst and confirmed the medium-sized brain edema with hydrocephalus.

### Case 3

A 33-year-old woman, with no medical history, died suddenly after a syncope which occurred in the morning. At autopsy, there was a cyst of the anterior part of the third ventricle of 4 × 2.5 cm, of bumpy appearance (Figure 2), associated with a nearby ventricular dilatation and a clear commitment of the cerebellar tonsils through the foramen magnum. When cut, the cyst had a gray-greenish aspect (Figure 3). During the pathological examination, the cyst wall was fibrotic. It was the seat of an inflammatory infiltrate and siderophages with abraded coating accompanied, in places, by bone metaplasia foci's. The content is composed of a colloid substance mixed with macrophage and crystals of cholesterol (Figures 4 and 5).

### DISCUSSION

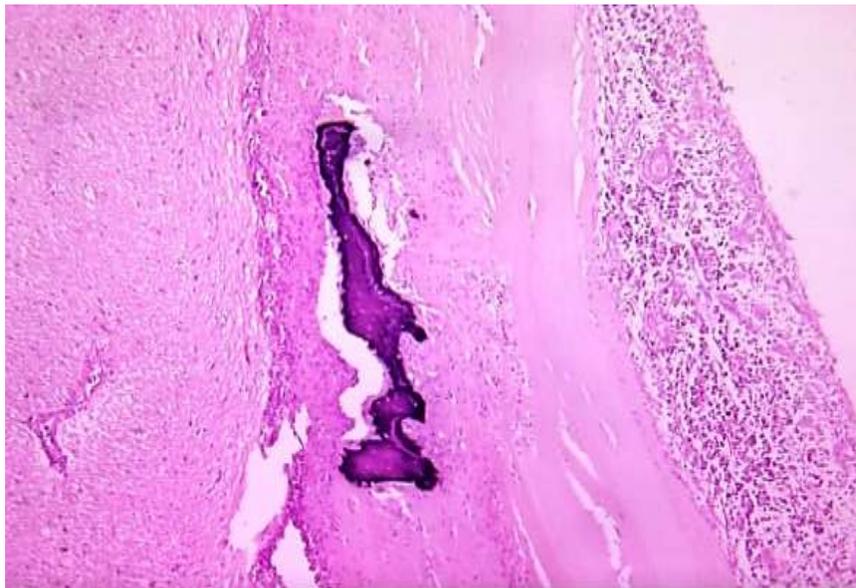
The colloid cyst is a rare cause of unexpected and sudden death (Hohenstein and Herdtle, 2010). Di Maio found, that for almost 11,000 autopsies, 19 deaths were due to brain tumors, and only one was due to a colloid cyst of the third ventricle (Dimaio et al., 1980). The colloid cyst of the third ventricle is a rare benign tumor, of dysembryoplastic origin. Related to its various proposed embryologic origins, other names for this tumor have been reported in the literature including paraphyseal and neuroepithelial cysts (Hwang et al., 1996). Family cases are rare but reported in the literature probably accompanied by autosomal dominant transmission (Joshi et al., 2005).

This congenital malformation was first described by Wallman in 1858, and Dandy, in 1922 (Bavil and Vahedi, 2007). In the literature, it represents 0.5 to 1.5% of brain tumors and its incidence is estimated at 3 individuals per million people per year (Roldan-Valdez et al., 2003).

The three cases reported in our series were two women and a man, aged from 33 to 45 years old, which confirms that these cysts are more common from the age of 30 to 50. However, they may occur at any age, even in children. In the literature, no gender predominance has been reported (El Gamal and Richards, 2006).



**Figure 3.** Greenish-gray appearance to the cut.

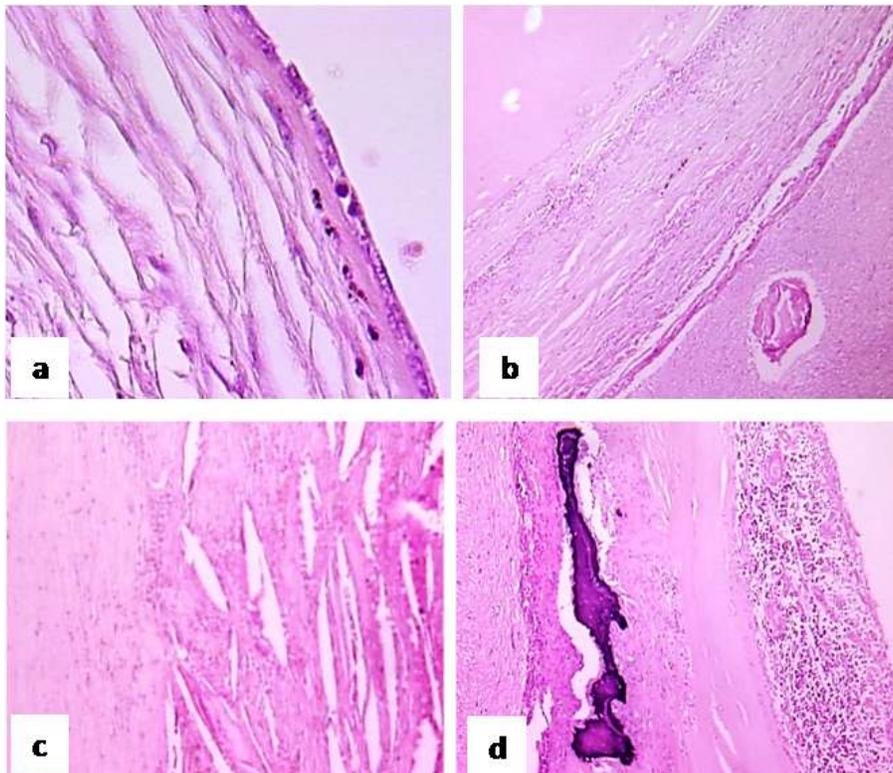


**Figure 4.** Osseous metaplasia of the cyst wall.

Macroscopically, this cyst is often smooth, spherical or oval in shape, of a diameter ranging from 3 to more than 40 mm. It is greenish, grayish or bluish in color. Its content is made of a homogeneous colloid substance of varying consistency (gel or liquid) (Andersen and Frost, 2015). During the autopsy, its small size and its possible rupture during the dissection can make it difficult to detect if a careful examination is not made. Therefore, the

diagnosis should be considered in the differential diagnosis of hydrocephalus and if a significant grey color is detected over the tuber cinereum (which is located between the chiasma opticum and the corpus mamillare) especially when the suddenly dead person has a medical history of headaches (Demirci et al., 2009).

Clinically, the cyst may be asymptomatic, incidentally discovered during a brain radiological investigation (brain



**Figure 5.** Microscopic feature of a colloid cyst: Cyst with Fibrotic wall adjacent to cerebral parenchyma, focally calcified, with macrophage resorption (b,d: HE  $\times$  100) , lined by a cuboid epithelium (a: HE  $\times$  400). The lumen is filled by a colloid fluid with cholesterol clefts (b, c: HE  $\times$  100, HE  $\times$  200)

scan, magnetic resonance imaging) indicated for another reason (Bender et al., 2013; Demirci et al., 2009). The typical appearance of the cyst on CT scan images (computed tomography) is due to its histological characteristics (viscous content surrounded by a fibrotic wall). Actually, with the advent of magnetic resonance imaging (MRI), detection of colloid cysts has become more frequent and accurate (Bender et al., 2013; Schmidek, 2006). The discovery of the cyst may occasionally happen at autopsy. In such condition, the cause of death may not be related to the cyst. However, it can be linked to this tumor although the subject doesn't present any clinical signs before death as it was the case in the first and the third observation.

When it is symptomatic, clinical signs of colloid cysts are not pathognomonic. The most common clinical symptom is an episodic headache, which may result from an intermittent obstruction of cerebrospinal fluid (CSF) flow through the foramina of Monro leading to a prompt increase in intracranial pressure (Spears, 2004). This symptom is usually accompanied by vomiting, visual disorders (diplopia, visual eclipses), flinch of the lower limbs, decreased tone for 5 to 15 min without consciousness troubles, vasomotor, thermal or fluctuating mental disorders (Roger et al., 2011). Symptoms are

often intermittent due to the cyst movements around its pedicle at the roof of the third ventricle, which may cause an episodic obstruction of Monro's foramen and an intermittent intracranial hypertension (Jaffree and Besser, 2001). Hydrocephalus is variable depending on the position of the head and the symptoms are usually relieved by the supine position, which is exceptional for the headaches that accompany brain tumors (Demirci et al., 2009).

Symptoms may be non-paroxysmal accompanied by continuous signs of intracranial hypertension, mental and memory troubles and endocrine disorders (amenorrhea, premature puberty). When the cyst is asymptomatic or small, especially without signs of hydrocephalus or increased intracranial pressure, the occurrence of sudden death raises the problem of physiopathological mechanism. In the literature, the two mechanisms most discussed are:

1. The obstruction of the foramen of Monro is the origin of acute obstructive hydrocephalus that leads to death, as it was the case in the second and the third observation. Nevertheless, the risk of sudden death is not proportional to the size of the cyst and the signs of hydrocephalus are not constant (Spears, 2004).

2. The irritation of the neurovegetative centers can result in neurogenic lung edema or cardiac disorders, such as a significant decrease in systolic ejection fraction which disappears after the excision of the cyst (Jarquin-Valdivia et al., 2005). These mechanisms are explained by the activation of the neuroendocrine system due to an increase of the intracranial pressure and may explain the occurrence of death in the first observation.

When it is diagnosed, the cyst must be treated because of the risk of sudden death's occurrence. Various surgical options are available. They include shunting of CSF, stereotactic cyst aspiration, transcortical-transventricular microsurgery, transcallosal-interforniceal microsurgery, and endoscopic surgery. The aim of surgery is to remove the blockage to CSF flow, restore normal intracranial pressure, and remove the entire lesion, because simple aspiration has been shown to lead to frequent recurrence of symptoms (Schmidek, 2006; David et al., 2013).

## Conclusion

These observations confirm that the colloid cyst of the third ventricle, although a rare benign tumor, is still responsible for sudden death. The diagnosis must be considered among the causes of intermittent headaches especially if they improve in the supine position and radiological investigations are required. Neither the size of the cyst and the degree of ventricular dilatation, nor the duration of clinical symptoms, appear to provide reliable indicators of the risk of sudden death. Surgical treatment of such tumor remains difficult because of its deep midline location. Its early detection with a total excision carries an excellent prognosis.

## Conflict of interest

Authors have none to declare.

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