Sudden Death due to Intracranial Colloid Cyst "A Case Report"

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Abstract

Intracranial colloid cysts are non-neoplastic epithelium-lined cysts of the central nervous system that almost always arise from the anterior third ventricle roof (immediately posterior to the foramen of Monro). These epithelium-lined, mucin-containing cysts can be found in asymptomatic patients; however, depending on their location, size, and degree of cerebrospinal fluid obstruction, patients may present with a variety of neurological symptoms. These symptoms can range from headaches to sudden death on rare occasions when there is acute hydrocephalus. This study describes a colloid cyst located between the two lateral ventricles at the level of foramen of Monro obstructing the cerebrospinal fluid (CSF) flow leading subsequently to sudden death and discovered incidentally during autopsy. The study was done in the Medico-legal institute in Baghdad from 1\textsuperscript{st} of November 2012 to 1\textsuperscript{st} of May 2013.

Key words  Sudden Death, Colloid Cyst, Autopsy, Intracranium.

Introduction

The dynamics of cerebrospinal fluid (CSF) production in the choroid plexus, its circulation throughout the ventricular system and subarachnoid space and then its reabsorption via the arachnoid granulations into the superior sagittal sinus is critical for homeostasis of the brain. Tumors can easily obstruct the circulation of the CSF and lead to abrupt enlargement and increase in the pressure of the ventricular system. This condition is known as acute hydrocephalus and it is potentially fatal. Example of this type of mechanism is the colloid cyst (which is not truly a tumor) \cite{1}.

Brain cysts are sacs filled with fluid and other organic debris. Almost all brain cysts form during fetal development; the colloid cysts form during embryonic development of the central nervous system \cite{2,3}. In 1910, Sjovall hypothesized that colloid cysts were remnants of the paraphysis, an embryonic midline structure with diencephalic roof immediately rostral to the telencephalic border (old name is paraphyseal cysts) \cite{4}. The origin of these cysts continues to be uncertain. Diencephalic ependymal, invagination of the neuroepithelium of the ventricle, or the respiratory epithelium of the endodermal origin are other etiologic possibilities so the colloid cysts are believed to derive from either primitive neuroepithelium of tela choroidea or from the endoderm \cite{5}. One leading theory is that colloid cysts form when ectopic elements migrate into the velum interpositum during ventral system embryonic development \cite{2,5}.

Colloid cysts usually arise in the anterior portion of the third ventricle between the fornices. The cysts are attached the roof of the third ventricle and frequently to the choroid plexus. Usually, the cysts are immediately dorsal the foramen of Monro. These cysts have also been reported to frequently arise in the septum pellucidum, the
fourth ventricle, and the sella turcica\(^6\). In 1858, Wallmann first reported on colloid cyst and in 1921, Dandy accomplished the first successful resection of a colloid cyst \(^7\).

Colloid cysts usually afflict adults (the youngest reported cases involve a 2-month-old infant) with uncertain gender differences; eight percent of the patients reported in the literatures are aged 30-60 years, approximately 0.1-1\% of all primary intracranial brain tumors and 15\% of all the intraventricular masses are colloid cysts and are the most common type of the neuroepithelial cysts, as well as the most common tumor in the third ventricle \(^2,4,5\). No known genetic relationship has been described, although familial occurrences of colloid cyst have been reported \(^5,7,8\). Typically, colloid cysts are clinical silent and are found incidentally when patients are imaged for other reasons and some types during autopsy, either computerized tomography scanning (CT) or magnetic resonance imaging (MRI) may help in diagnosing a colloid cyst, although MRI has fewer advantages. MRI typically demonstrates the location of the cyst, and the nature of intacystic contents \(^9,10\). These cysts can trigger headaches, fever and dizziness. The patient may also experience bouts of nausea and vomiting. Cysts rupture can be particularly dangerous; the contents spill into the brain can lead to meningitis. Cyst growth can cause hydrocephalus, a condition in which blockage prevents normal flow of fluid through the brain, causing the fluid to accumulate to dangerous level. In particularly severe cases, the afflicted person dies suddenly, without having experienced any prior symptoms \(^1-4\).

**Case report**

On 25\(^{th}\) of February, 2013; a natural sudden death case was referred by the police from Al-Mahmudiya to the medico-legal institute in Baghdad. The victim was 20 years old female with history of migrainous headaches since 2 years. She was treated with amitryptyline 25 mg once daily, 1 year before her death. Her medical history was negative apart from two previous cesarean sections. The night before her death; she complained of severe headache, which was relieved by a non-steroidal anti-inflammatory drug (oral Ibuprofen 200 mg) and then she went to bed after feeling well; during the night she had frequent projectile vomiting and the next morning she was found dead; face down in the bathroom. At autopsy the brain appeared swollen 1250 g in weight with flattening of the gyri throughout the convexities. The brain was symmetrical with no shifting of the midline structures. At the base; the unci of the temporal lobes were grooved and the hypothalamus appeared prominent but there was no definite herniation grossly.

On sagittal section the lateral ventricles were enlarged and containing a colloid cyst, 2.5 cm in diameter attached to the septum pellucidum at the level of foramen of Monro. The lateral ventricles were enlarged but the third and fourth ventricles were of normal size (Fig. 1.).

Fig. 1. Colloid cyst of the ventricles at the level of foramen of Monro attached to the septum pellucidum.

The cyst wall was elastic and yellowish in color, lined with mixed array of epithelial and goblet cells (Fig. 2) and full of proteinaceous mucinous fluid, slightly turbid-yellow and its consistency was thin gelatinous.
Fig. 2. Colloid cyst showing a cystic lesion lined by a thin fibrous capsule (X40 H. & E. stain).

It was concluded that her cause of death was due to acute hydrocephalus resulting from obstruction of CSF flow by the colloid cyst. No other pathological findings both grossly and microscopically were seen in her body and toxicological screening tests were negative.

Discussion

Undiagnosed brain tumors are rare cause of sudden natural deaths observed in medical examiners settings. Benign tumors are more likely than their malignant counter-parts to present with sudden death \(^{(11,12)}\). On rare occasions, a colloid cyst may obstruct the foramen of Monro completely and irreversibly, resulting in sudden loss of consciousness and, if patients are not treated, coma and subsequent death due to herniation. An alternative theory suggests that sudden death in patients with colloid cysts may be related to acute neurogenic cardiac dysfunction (secondary to acute hydrocephalus) and subsequent cardiac arrest rather than herniation \(^{(2)}\). The risk of sudden death remains difficult to predict, the risk of sudden death does not seem to correlate with tumor size, degree of ventricular dilatation, or duration of symptoms \(^{(13)}\). A study found that 8% of asymptomatic patients with a colloid cyst of the third ventricle eventually became asymptomatic \(^{(4)}\), whereas a different study found 34% patients presented to a hospital with acute deterioration and in some cases sudden death \(^{(1)}\). Cyst size and extent of ventricular dilatation do not seem to predict for acute deterioration \(^{(6)}\). Pollock et al. found that ventriculomegaly is the most important variable associated with cyst-related symptoms, other variables include age and cyst size \(^{(2)}\). Eberhardt et al. described their 20 years of experience of previously undiagnosed brain tumors resulting in sudden death. In Eberhardt series of 11 neoplasms associated with sudden death, seven (63%) were gliomas, two (18%) were intraventricular colloid cysts, one (9.09%) was pituitary adenoma and one (9.09%) was schwannoma \(^{(12)}\). Following age-related regional frequency of brain tumors, lesions that involve the fourth ventricle are more common in children, whereas those of the third and lateral ventricles predominate in adults and this agrees with this study where the tumor was found in 20 years old female \(^{(1,4,7,14)}\). A study by Shemie et al. from Hospital for Sick Children in Toronto indicated that from 1990 through 1997, seven children died unexpectedly with acute obstructive hydrocephalus. All children had an intracranial tumor located at a critical site for CSF circulation, three with colloid cyst, two with astrocytoma, one with ependymomma and one with suspected lymphoma. This study was limited to children only where colloid cysts are more prevalent \(^{(14)}\).

References


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