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CASE REPORT Open Access

Sudden unexpected death caused by a colloid cyst in the third ventricle: case report

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Abstract

Background Colloid cysts arise from congenital abnormalities of the endodermis and are characteristically comprised of an external fibrous layer, with a lining of cuboidal and columnar epithelial cells and endodermal ciliated cells. They contain a gelatinous matrix, produced by mucus-producing epithelial cells.

Case presentation An 8-year-old Saudi girl died in Dammam Central Hospital, to which she was admitted by her parents following her complaints of intense headaches. When the body was examined externally, it was noted that she had bilateral eye congestion, evidence of needle marks on all four limbs and in the region of the femoral triangle and bruising in the central chest and left shoulder areas. No other evidence of injury was noted. A cyst in the third ventricle of the brain was identified at post mortem. Investigations for alcohol, toxins and illicit substances were negative.

Conclusion It was surmised that the child had died as a result of a cerebral colloid cyst and its clinical sequelae.

Keywords Child, Cerebral, Colloid cyst, Unexpected death, Case report

Background

Colloid cysts have an annual incidence of approximately 2.3 instances per million population and form 1% of intracranial malignancies. Post mortem studies have suggested that the majority of colloid cysts, i.e. 1 in 8500, cause no symptoms as they are slow growing. The incidence of this pathology is equivalent in males and females. The majority of colloid cysts are diagnosed in adults within the age range, 20–50 years; between 1 and 2% affect paediatric patients aged less than 10 years (Turillazzi et al. 2012). Irrespective of age, the presentation of a colloid cyst, which is a benign tumour, is typically related to raised intracranial pressure (Farooq et al. 2016). Typically, colloid cysts arise in the anterior portion of the third ventricle, bordered by the forniceal columns. This location results in blockage of the foramina of Monro and acute onset hydrocephalus, which is the principal aetiology underlying their mortality rate (Lannon et al. 2016).

The natural course of colloid cysts is not clearly delineated owing to their relative infrequency and low case numbers in the majority of series published in the literature. Thus, prognostic markers and well-described indicators for operative intervention, particularly in individuals in whom such a mass has been detected incidentally, are lacking. In excess of 50% of patients with a colloid cyst within the third ventricle report symptoms in keeping with hydrocephalus, i.e. headache, nausea, vomiting, blurred vision, an ataxic gait and impaired cognition, and therefore undergo operation. In the current case study, the child was complaining of headaches on

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admission to hospital, and post mortem findings demonstrated obstructive hydrocephalus. Following presentation with acute obstructive hydrocephalus, a colloid cyst may be fatal in a low percentage of individuals who fail to respond to diversion of the cerebrospinal fluid and supportive therapy (Beaumont et al. 2016).

Case presentation

Since the 8-year-old child's cause of death was unknown, her body was admitted to the morgue for a post mortem study.

External examination

Clothes

No clothes were present on the body.

External appearance

The body was that of a female Saudi child with a 140 cm height and a typical body build and weight for her age. Rigor mortis was present although affected by refrigeration. A red hue of hypostasis was noted on her back but did not involve the pressure areas. There were no signs of decomposition.

Head and neck

No injuries were seen in the head and neck area (Fig. 1). When the face was looked at, there were no broken bones; both eyes were congested and free of injuries; there were no injuries or foreign objects in the nose or nostrils, and there was no rhinorrhea. The lips and mucous membrane were free from injuries.

Chest, abdomen, back and pelvis

Bruising was evident in the chest centre (Fig. 2). Needle marks were obvious on either side of the femoral triangle (Fig. 3). These findings were consistent with clinical procedures. No injuries were observed on the abdomen or back.

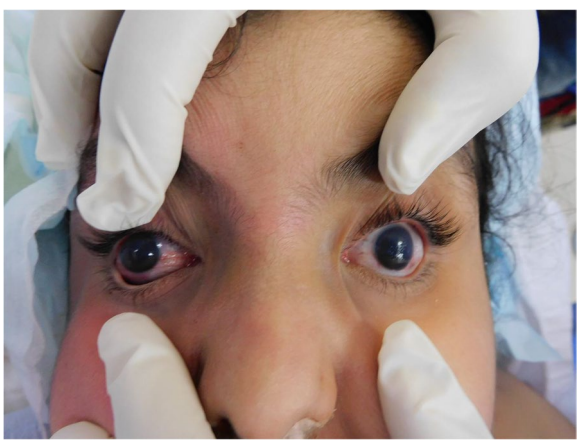


Fig. 1 Both eyes were congested and no facial injuries

Fig. 2 Contusion over the mid of the chest, indicating medical intervention

Upper limbs

A red, circular bruise, 3 cm in diameter, was present on the left shoulder, relating to clinical procedures (Fig. 4). Numerous puncture wounds relating to clinical procedures were seen on the back of both hands (Fig. 5), wrists (Fig. 6) and forearms, the latter 3 cm inferior to the cubital fossae (Fig. 7).

Lower limbs

A needle mark, associated with a clinical procedure, was identified on the lateral side of the left ankle (Fig. 8). The lower limbs were otherwise of a normal appearance.

Autopsy

Head

No bruising was observed in the reflected layers of the scalp. No injuries were noted on the calvarium or skull, and no intracranial bleeding was identified. The brain appeared symmetrical but was increased in size, weighing 1700 g, and there was evident oedema and congestion. The definition of the gyrae was lost, and the sulcus was narrowed (Fig. 9). On dissection, the third ventricle was found to contain a cyst, which was filled with a mucoid substance, yellow-green in colour (Fig. 10).

Face and neck

No injuries were present on the face, and no haemorrhages were identified within the muscles in the cervical region.

Chest and abdomen

An incision was made in the midline which extended between mental and pubic symphyses. Exposure of the thoracic wall and rib cage showed the structures to be intact without any bruising or fractures. The lungs were

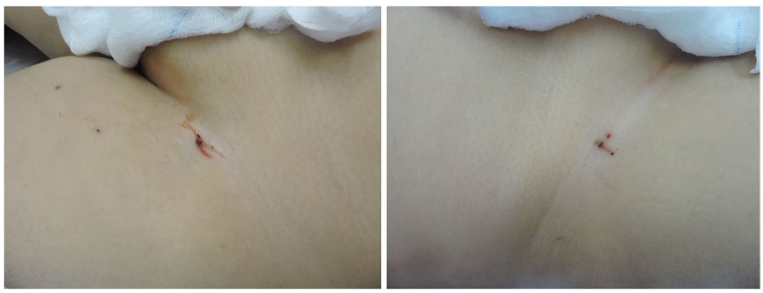


Fig. 3 Needle marks in both femoral triangles

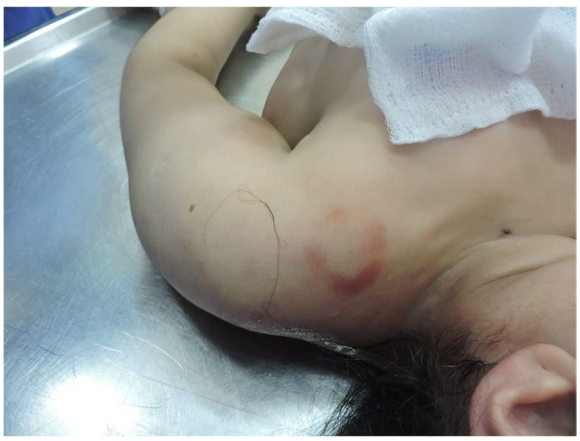


Fig. 4 Contusion over the left shoulder, indicating a medical intervention

of normal dimensions and appeared outwardly normal. The cardiac structures were also normal; no congenital anomalies were seen, and the coronary arteries were free from obstruction. No bruising was evident within the abdominal wall; the organs within the abdominal cavity had a normal external appearance.

Histopathological samples

A complete set of pathological specimens was obtained, which included samples from the cerebral cyst and third ventricle. The findings are detailed below.

Brain

The presence of a cyst, filled with a mucoid substance and with a simple epithelial lining, was confirmed. A colloid cyst was queried (Fig. 11 a and b). Cerebral oedema was reported.

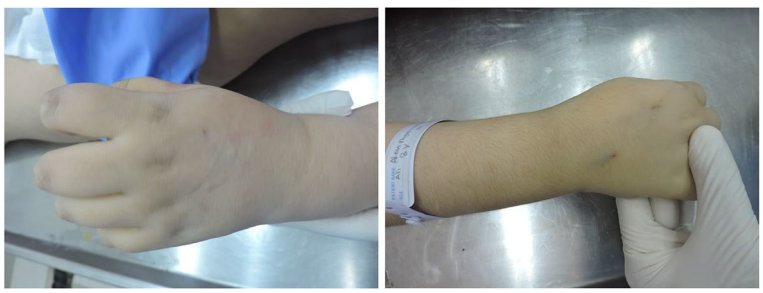


Fig. 5 Needle marks over dorsum of both hands

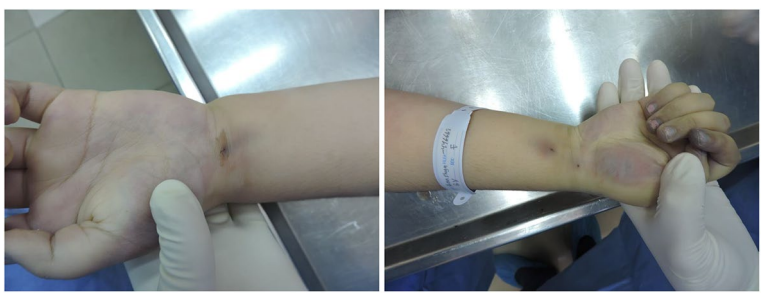


Fig. 6 Needle marks over both wrists

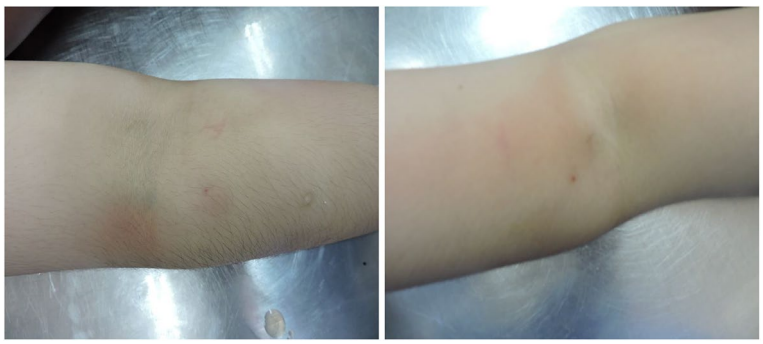


Fig. 7 Needle marks below both cupital fossa

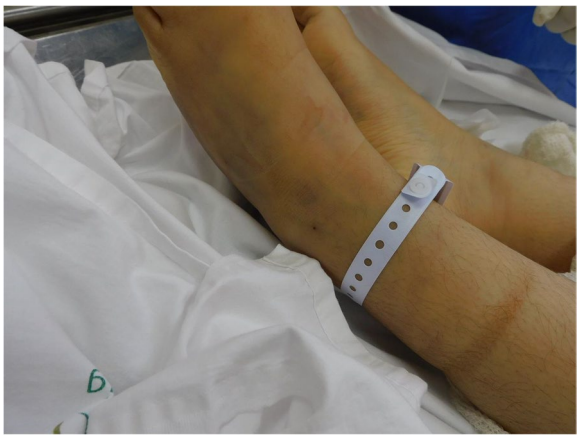


Fig. 8 Needle mark over the lateral aspect of the left ankle

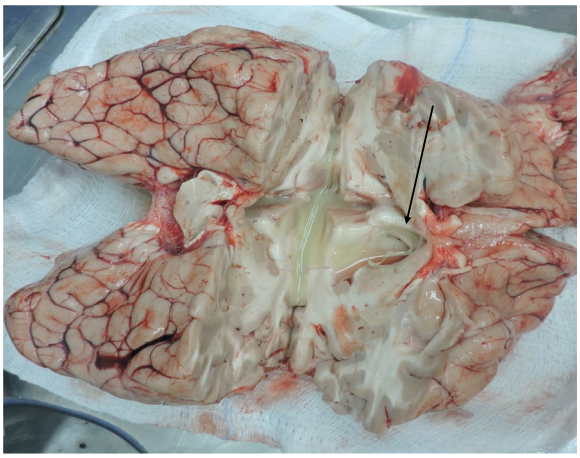


Fig. 10 Mucoid materials and cyst in the brain's third ventricle

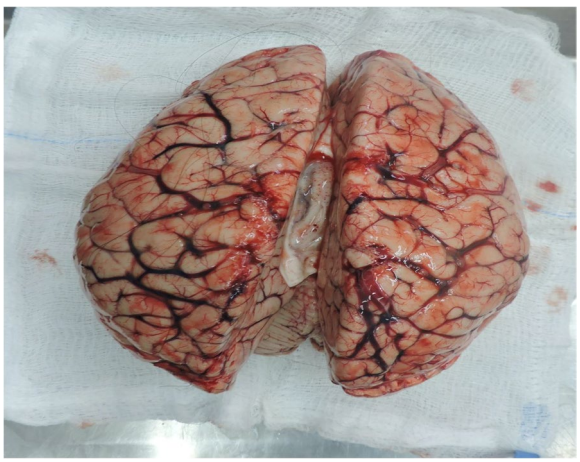


Fig. 9 Significant brain swelling with gyral flattening, sulcal narrowing and congestion

Heart

The heart was normal in shape and size with no congenital anomalies. All heart valves were free of congenital malformations or pathological diseases. Two coronary ostia were found, and both were anatomically normal. The heart ventricles wall thickness was normal, with the left one measuring about 1.8 cm.

Lungs, liver, kidneys, spleen and suprarenal glands

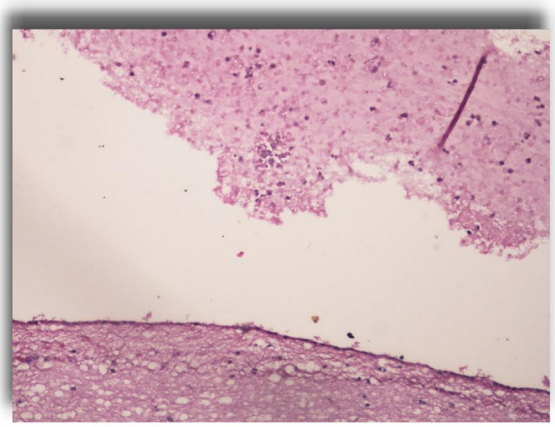
Congestion was noted in these viscera.

Toxicology samples

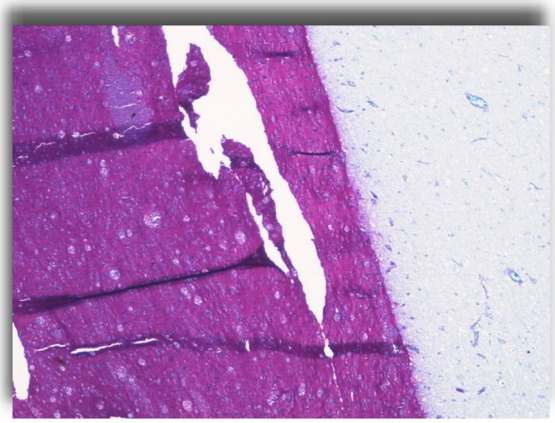
Specimens of blood and serum were taken for toxicological analysis, which included alcohol, narcotics, toxins and illicit substances; the results were negative in all cases.

Discussion

Colloid cysts arise from congenital abnormalities of the endodermis and are characteristically comprised of an external fibrous layer, with a lining of cuboidal and columnar epithelial cells and endodermal ciliated cells. They contain a gelatinous matrix, produced by mucus-producing epithelial cells (Coce et al. 2011). Their usual



a



b

Fig. 11 **a** This microscopic picture stained with H&E stain showing mucinous debris in the top of the picture and brain tissue down lined by simple epithelium. **b** PAS stain: mucin (left) and brain tissue (right)

locus is the rostral area of the roof of the third cerebral ventricle; they form a space-occupying lesion within the anterosuperior portion of the ventricle in proximity to the foramina of Monro (Turillazzi et al. 2012). Cerebral masses which should be considered in the list of differential diagnoses include abnormalities of the choroid plexus and ependymal, arachnoid and enterogenous cysts, respectively. In particular, the latter may have similar clinical, radiological and macroscopic characteristics to a colloid cyst (Lavrador et al. 2016).

Headaches and nausea are commonly experienced by patients with a colloid cyst, and drop attacks, which are indicative of a greater risk of death, occur in approximately 1 in 3 individuals. Over twenty familial studies, four of which were conducted in monozygotic twins, have demonstrated that there is a significant genetic contribution to the aetiology of colloid cysts, with heightened concordance in twins regardless of whether they were mono- or dizygotic. This increased incidence in the twin of an affected individual may also relate to the common environment in utero. However, some studies have suggested that epigenetic differences may be relevant as there are some reports of colloid cysts presenting in just a single twin (Lannon et al. 2016). Given the increased incidence of this pathology in identical twins, however, the authors suggest that genetic investigations or screening, in the context of one twin being diagnosed with a colloid cyst, could be indicated in order to recognise any associated clinical syndromes (Lannon et al. 2016). There are also reports of familial colloid cysts in the absence of twins (Weisbrod et al. 2016).

In individuals in whom colloid cysts have proven fatal, it is common to find dilated ventricles and cerebral herniation at autopsy. Thus, it is commonly believed that death is likely to be related to the presence of hydrocephalus, although the exact underlying mechanisms are still to be elucidated. Sudden death may arise even when a colloid cyst is only of modest dimensions; the size of the lesion and the extent of associated ventricular enlargement do not appear to be indicators of the likely prognosis. Thus, it is probable that other processes, in addition to the physical presence of the cyst, contribute to unexpected fatality. The hypothalamus gland is situated in proximity to the borders of the third ventricle, where colloid cysts are most likely to form. This gland controls the neuroendocrine and autonomic systems. The latter has an important regulatory effect on the heart, and there is therefore the potential for aberrations in the hypothalamic cardiovascular pathways to arise as a consequence of the mass effect of the tumour, which then cause reflex cardiac abnormalities and possible sudden fatality. Acute cerebral hydrocephalus is another mode of death linked with colloid cysts; this is manifest by raised intracranial pressure, cerebral herniation, compression of the hindbrain and medullary impairment which, in turn, leads to impaired cardiorespiratory function and fatality (Turillazzi et al. 2012).

In this case, the individual concerned was a child. Colloid cysts arise most frequently in children and teenagers but rarely cause symptoms until the individual concerned reaches their third or fourth decade. Often, the lesion is found incidentally during a post mortem examination (Skerbinjek Kavalar et al. 2016).

Conclusions

In view of the clinical context, external examination, post mortem findings and the histopathology and toxicology analyses, it was surmised that the child had died as a result of a cerebral colloid cyst and its clinical sequelae.

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N/A.

Authors' contributions
AA, autopsy and preparation of medicolegal report, critical review and proof reading. AA, autopsy and preparation of medicolegal report, critical review and proofreading. GA, autopsy and preparation of medicolegal report, critical review and proofreading. AA, review of literature and preparation of manuscript. MH, review of literature and preparation of manuscript. BA, critical review and proofreading. MA, critical review and proofreading. The authors read and approved the final manuscript.

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Availability of data and materials
Available from the corresponding author upon request

Declarations

Ethics approval and consent to participate
This case was approved by ethics committee of Forensic Medicine Center, general directorate of health affairs, eastern province, Ministry of Health, KSA.

Consent for publication
Informed consent for publishing and presentation of the case report was obtained from the father of the deceased girl.

Competing interests
The authors declare that they have no competing interests.

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