Spontaneous Decrease in Size and Change in MRI Signal Characteristics of a Colloid Cyst in a Pediatric Patient

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Abstract
Colloid cysts are rare benign tumors that can cause acute obstructive hydrocephalus and sudden death. The literature indicates that many of these tumors are likely to be asymptomatic and that some may spontaneously regress in adults. However, the majority of colloid cysts in young patients are more clinically and radiologically overt. Conservative management is recommended in asymptomatic adults. While early diagnosis followed by surgical resection is recommended for the pediatric age group. Given that there have been few reported cases of spontaneous regression of colloid cysts in pediatric patients not requiring surgical intervention, management guidelines for pediatric patients are unclear. We present a case of a pediatric patient with a colloid cyst that underwent spontaneous decrease in cyst size along with changes in signal characteristics prior to surgery.

Keywords: Colloid cyst; Pediatrics; Treatment; Surgery; Imaging

Introduction
Colloid cysts are histologically benign tumors that have the potential to cause fatal outcomes. They arise from ectopic endodermal elements at the anterior margin of the third ventricle where they can remain clinically silent or enlarge and cause sudden death by abrupt obstructive hydrocephalus [1]. Colloid cysts usually affect adults in their third to fifth decades. Their incidence is unknown in the pediatric population [2].

The natural progression of colloid cysts is poorly understood [3]. Some studies suggest that in adults, colloid cysts are more likely to be asymptomatic and can even regress. While in young adults and children, colloid cysts tend to be more clinically and radiologically aggressive [2,4-6].

Studies in the adult population support the current standard of practice to surgically manage symptomatic cysts and conservatively manage asymptomatic ones. The few studies conducted within the pediatric age group advocate early diagnosis and surgery, even in incidental cases [2,6]. However, three reported cases of spontaneous regression of colloid cysts in pediatric patients question this rationale [7,8].

We present a case of a 16-year-old female who experienced progressive headache and acute vision changes and was found to have a colloid cyst with resultant hydrocephalus. Prior to surgery, the patient experienced improvement of her symptoms. MRI demonstrated decrease in cyst size and signal changes. This case serves as another example that spontaneous regression in cyst size can occur in the pediatric population and that conservative management may be considered.

Case Presentation
A 16-year-old female with six months history of headaches associated with photophobia, dizziness and vertigo, presented to the emergency room with worsening headache and new onset of blurry vision. She had no other significant past medical history and was neurologically intact on physical exam.

CT demonstrated a 3.7 x 3.6 x 3.8 cm well circumscribed, isodense mass at the foramen of Monro/roof of the third ventricle with associated hydrocephalus (1) MRI demonstrated a non-enhancing lesion that is hypointense on T1WI; (2a) and hyperintense on T2WI; (2b) and FLAIR sequence; (2c) No hemorrhage was noted on the SWI; (2d) The imaging features were consistent with a colloid cyst. Surgery was scheduled in a few weeks, when most convenient for the patient, as she was hemodynamically and neurologically stable.
A follow up preoperative MRI showed interval decrease in cyst size (3.0 x 3.3 x 3.3 cm) and mildly improved hydrocephalus (3) Interestingly, increased, homogenous T1 signal; (3a) and low T2; (3b) and low FLAIR; (3c) signal changes were noted. There was also interval development of mild susceptibility artifact within the cyst; (3d)

Despite the decrease in size, the neurosurgical team and patient’s parents elected to undergo endoscopic resection. Resection of the cyst was reported to be difficult given the thick nature of its content. Post-operative course was uncomplicated and patient was discharged in excellent condition.

Discussion

Colloid cysts comprise about 1% of all intracranial tumors and 15-20% of all intraventricular tumors [9]. Their potential for fatal outcomes, likely stems from the mucin secreting goblet cells and simple/ pseudostratified columnar epithelial cells that line the cyst wall [9]. Due to these mucin secreting cells and their location near the foramen of Monroe, colloid cysts can enlarge and cause abrupt obstruction of the ventricles leading to increased intracranial pressure and brain herniation [1]. However, many tend to remain clinically silent. The management of colloid cysts remains challenging as there is limited information regarding their growth rate and natural evolution [1,3,6,9].

Symptomatic cysts are managed surgically due to evidence that clinical deterioration is typically preceded by symptoms of increased intracranial pressure [3,10]. Pollock et al’s retrospective studies in 155 symptomatic and asymptomatic patients helped to define management in the adult population [4]. Pollock et al investigated 87 symptomatic patients from an initial patient cohort of 155 patients in an attempt to understand the natural history of colloid cysts [4]. The study discovered that the risk factors associated with symptomatic cysts were patient age, cyst size, ventricular dilatation and increased signal on T2WI. This information was used to establish distinct patient classes, which showed that younger patients (<50 years old) with large cyst size (>10mm) and hydrocephalus were likely to become symptomatic while older patients were likely to remain asymptomatic depending on cyst size, ventricular dilatation and presence or absence of high T2 signal. A second study analyzed the remaining 68 asymptomatic patients and demonstrated that observation with serial imaging resulted in only 8% of the patients becoming symptomatic during a 10-year period [5]. This latter study concluded, that conservative management was possible in asymptomatic patients unless there was occurrence of symptom development, cyst enlargement or hydrocephalus development [5].

Subsequent case studies in the adult population have further validated conservative management by showing that incidentally discovered asymptomatic to mildly symptomatic cysts can also regress [11-14]. The mechanism of this regression is unclear. One theory for regression is cyst rupture. A study from Motoyama et al reported a case of spontaneous cyst rupture with follow-up imaging demonstrating a shrunken cyst wall attached to the anterior roof of the third ventricle [15]. Peeters et al reported a case of cyst regression.
which showed residual high T1 signal at the time of cyst disappearance, perhaps representing discharge from cyst rupture [1]. However, worsening symptoms and imaging findings, such as hydrocephalus, were associated with these examples. There are other examples of cyst regression in patients who have demonstrated improved or resolved symptoms without evidence of cyst rupture. The etiologies in these cases are unclear, but may be related to cyst hydration status' effect on cyst size and symptoms. Kachara et al reported case of pediatric patient who demonstrated decreased colloid cyst size with increased density on CT that may have been due to water extraction in the setting of CSF diversion [7]. Pollock et al's study demonstrated association between high T2 signal and symptomatic cysts [4]. It was concluded that high T2 signal indicated higher water content and ongoing expansion of the cyst while low T2 signal indicated desiccated, more solid material and less growth capability of the cyst.

Studies in children suggest that colloid cysts in the pediatric population tend to be clinically and radiographically aggressive [2,6,16]. For example, Buttner et al studied cases of colloid cysts that were associated with sudden death and 7 out of the 21 cases were pediatric patients [17]. Alnaghmoosh et al describes a retrospective study of 43 patients with colloid cysts in which 7 patients ranging from 6-18 years old had aggressive clinical presentation of increased intracranial pressure with imaging findings of hydrocephalus, large cyst size and high T2 signal [2]. This study concluded that the pediatric population requires early surgical management.

However, a recent case report by Hamidi et al of a pediatric patient with a colloid cyst found on imaging who experienced diminished symptoms after ten months, without surgical intervention. This paper concluded that in light of the clinical course of this patient, a more conservative approach could be considered for young patients, especially given the risks typically involved with surgery.

Although our patient ultimately underwent surgical intervention, our case also raises the possibility of adopting a more conservative approach to treating colloid cysts in pediatric patients, even when they are initially asymptomatic. Pre-operative imaging demonstrated decreased cyst size and decreased hydrocephalus with concomitant decrease in patient's symptoms (3). The lesion changes signal characteristics from iso-intense T1/high T2 signal (2) to high T1/low T2 signal with development of susceptibility artifact (3). Ultimately, the mechanism of cyst regression is unknown as there was no evidence of cyst rupture. The susceptibility artifact within the cyst is of unclear significance as hemorrhagic colloid cysts are usually associated with acute deterioration [18]. The interval change in the signal characteristics of the patient's colloid cyst is likely due to a change in hydration status raising the possibility that the change in cyst size may have occurred due to cyst desiccation. The altered signal characteristics, may also explain the difficulty experienced during removal of the cyst as low T2 signal has been reported to suggest high viscosity of the tumor and difficulty with aspiration during endoscopic procedures [19].

Beaumont et al recently recommended conservative management of symptomatic patients with colloid cysts in adults [3]. This study found that only a subset of symptomatic patients presented with acute obstructive hydrocephalus and sudden death [3]. Similar to Pollock et al, this study evaluated the predictive factors for symptomatic colloid cysts [4,5]. Notably, it further stratified risk factors for obstructive hydrocephalus by developing a Colloid Cyst Risk Score. Yet, since this study was a retrospective analysis, confirmation of these findings will be needed through a prospective study prior to establishing management guidelines for determining the need for surgical intervention in pediatric patients with colloid cysts.

Conclusion

This case suggests that a conservative approach to colloid cyst management may be possible in pediatric patients. Furthermore, symptomatic patients can also show cyst regression. Since surgery is associated with inherent risks, it is important to investigate which factors determine risk for obstructive hydrocephalus and poor outcomes in pediatric patients.

References