CASE REPORT

Neurology



Colloid cyst of the third ventricle

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Abstract

A colloid cyst is a malformation composed of an amorphous, gelatinous material surrounded by epithelial and connective tissue. It is typically located in the third ventricle, leading to signs of increased intracranial pressure and hydrocephalus. In this case report, we discuss a classic presentation of a patient presenting with a colloid cyst of the third ventricle. This includes the patient's symptoms before arriving at the emergency department and presentation upon arrival and during their hospital stay. We also discuss the diagnostic approach to colloid cysts, providing radiographic imaging to support the diagnosis. Finally, we discuss the approach to treatment of a colloid cyst, including temporizing measures to relieve symptoms and definitive measures for removal of the colloid cyst. The overall approach from diagnosis to management to definitive treatment of a colloid cyst requires an interdisciplinary approach but prompt recognition of the signs and symptoms in the ED can decrease both morbidity and mortality associated with this potentially fatal diagnosis.

1 | INTRODUCTION

Increased intracranial pressure (ICP) is a rise in the pressure inside the skull that can be caused by a number of pathologies including, but not limited to, traumatic head injuries, infections, and space-occupying lesions like brain tumors. There are many uncommon or rare causes of increased ICP, one of which includes colloid cysts of the third ventricle, which represent 2% of intracranial tumors. The incidence of colloid cysts of the third ventricle are about 0.9 per 1 million. 10 The prevalence is estimated to be around 1 in several thousand. 9 Colloid cysts are benign intracranial tumors usually located in the third ventricle of the brain. They are composed of connective tissue and are mucin producing. ⁶ These benign, slow-growing tumors have the potential to cause obstruction to the flow of cerebral spinal fluid (CSF), which lead to signs of increased ICP, including headaches, gait ataxia, nausea, vomiting, and vision difficulties. Of these findings, headache is by far the most common.⁷ Prompt recognition and diagnosis via

computed tomography (CT) or magnetic resonance imaging (MRI) and treatment with neurosurgical intervention is necessary, especially for symptomatic patients, as these tumors can lead to an acute decompensation and rarely sudden death.9

1.1 | Case presentation

The patient is a 54-year-old female who presented to the emergency department via private vehicle because of a frontal headache for 2 days. She stated that the headache was exacerbated by positional changes. Specifically, the headache was worse when sitting upright and improved when lying down or flat. The patient also complained of increased fatigue, overall soreness, and a feeling of heaviness. She denied any history of migraines. However, she previously had a headache that led to a diagnosis of a colloid cyst, which was removed in 1995 by a neurosurgeon in Cincinnati. The patient's physical

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FIGURE 1 Computed tomography brain scan of 54-year-old female with lateral ventriculomegaly

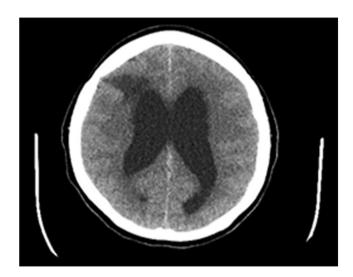


FIGURE 2 Computed tomography brain scan of same 54-year-old female with lateral ventriculomegaly

examination was grossly negative, with the exception of an abnormal gait, which exhibited unsteadiness with ambulation. Cranial nerve testing, sensory deficit testing, and strength testing were normal.

1.2 | Primary findings

The patient underwent a CT brain scan with and without contrast that showed findings suggestive of obstructive hydrocephalus. The lateral ventricles were dilated and the third and fourth ventricles were normal in size. Although we did see hydrocephalus and lateral ventriculomegaly, the CT did not readily identify the obstructing mass (Figures 1–3). An MRI with and without contrast was then performed, which also showed lateral ventricular dilation, confirming the previous

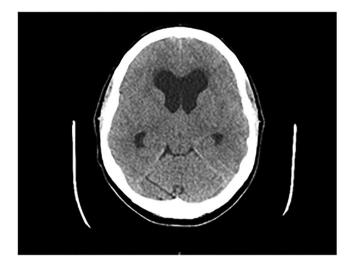


FIGURE 3 Computed tomography brain scan of same 54-year-old female with lateral ventriculomegaly, not showing cystic mass

findings seen on the CT brain scan. Evidence of a cystic mass was seen in the region of the foramen of Monro, which appeared to obstruct CSF flow and cause the ventricular dilatation. Collaboratively, the patient's case was discussed with a neurosurgeon and she was scheduled for surgical removal of the obstruction. Before the surgery was performed, the patient began to show further signs of neurologic decompensation, most noticeable being ataxic gait. She quickly underwent ventriculoperitoneal (VP) drainage to relieve the increased intracranial pressure and decrease hydrocephalus. Soon after, she ultimately was transferred to a different facility where she underwent surgical intervention to remove the colloid cyst and the VP drain. Post surgery, the size of the lateral ventricles decreased closer to normal. The patient improved neurologically and showed no signs of residual deficits.

2 | DISCUSSION

Colloid cysts of the third ventricle are benign intracranial tumors and represent approximately 2% of intracranial neoplasms. 1 Although colloid cysts are rare causes of intracranial tumors, it is thought that these tumors account for 1 in 5 primary intraventricular tumors.⁸ The prevalence of colloid cysts of the third ventricle, based on some studies, is estimated to be lower than 1 in several thousand. The incidence rate is thought to be even lower at \approx 0.9 per million. ¹⁰ The composition of these cysts usually includes an outer layer made of connective tissue and an inner epithelial layer that is ciliated and mucin producing.⁶ The cysts consist of an amorphous, gelatinous material that stains periodic acid-Schiff positive. As colloid cysts are a developmental malformation, they may be present early in childhood but can remain asymptomatic, which is partially due to the slow-growing nature of this intracranial tumor. However, if this cyst does become symptomatic, it will typically do so in the third to sixth decade of life. Studies have demonstrated that up to 34% of patients who become symptomatic have rapid and acute deterioration, similar to our case.9

Some cases have shown cyst sizes of 1-2 cm in diameter, though larger lesions have been found.² Cysts enlarge over time because of the accumulation of desquamated and secretory products. Though not inherently dangerous, the endodermal congenital malformations can cause complications as seen in this case. Symptoms are usually nonspecific and include signs of increased intracranial pressure, such as headaches, nausea, vomiting, papilledema, intracranial hypertension, hemorrhagic changes, and rarely sudden death.^{3,4} These symptoms manifest when there is obstruction of flow to the CSF. Patients may also present with neurological symptoms like unsteady gait and idiopathic drop attacks. Headaches are by far the most common presenting complaint, being present in 68% to 100% of diagnosed cases. They can be mild to severe and are usually intermittent and paroxysmal in nature. The intermittent nature of the headaches could potentially be caused by the colloid cyst acting as a "ball-and-valve" blocking the flow of CSF out of the lateral ventricles.⁸ These headaches are positional; worse when sitting upright and improved when lying supine. These headaches are often very brief, lasting only seconds to minutes but have been shown to last for days.⁷ Acute obstruction of CSF, most commonly located at the foramen of Monro, can cause intense, severe headaches (thunderclap headaches) and can be associated with nausea and vomiting. It is thought that the combination of intermittent headaches and drop attacks are pathognomonic for obstruction via colloid cyst, but this constellation of symptoms is present in only about a third of diagnosed patients.⁶ A combination of clinical presentation and imaging are used to diagnose colloid cysts of the third ventricle. CT brain scan with and without contrast is usually the first imaging modality used and typically reveals a hyperdense, isodense, or hypodense rounded or ovoid lesion within the third ventricle. MRI brain scan will show a hyperintense lesion on T1-weighted imaging and a hypointense lesion on T2-weighted imaging. Both CT and MRI will show ventriculomegaly of the lateral ventricles with normal sized third and fourth ventricles. This pathology is because of the colloid cyst obstructing the CSF at the foramen of Monro, which is the most common location of blockage (Figures 4-6). As in our case, the CT brain scan may not show the cystic mass or obstruction. Based on the clinical presentation and history, this should prompt the physician to consider more definitive imaging like MRI brain scan to visualize the obstruction.

Early identification of third ventricular colloid cysts is necessary because of the risk of sudden death. Acute hydrocephalus due to obstruction is treated with surgical intervention. Based on the patient presentation, a ventriculostomy may be needed as an initial treatment to effectively decrease intracranial pressure and relieve symptoms, but it is temporary. Performing this procedure can assist in decreasing the risk of persistent neurological deficits. In the long term, neurosurgical intervention for removal of the obstruction is imperative. There are multiple possible approaches to the removal of these cysts that have been successful, such as the transcortical or transcallosal approaches as well as stereotactic or endoscopic aspiration. Though uncommon, patients with a prior history of neurosurgical intervention can have reoccurrence of the cyst, as seen in our patient. It is important that this diagnosis be recognized early. On initial presentation to the ED, patient history and physical examination can assist immensely in increasing the

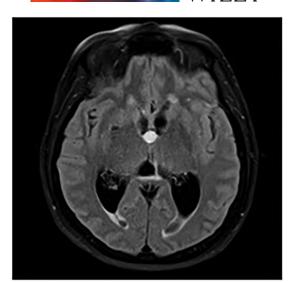


FIGURE 4 Magnetic resonance imaging brain scan of 54-year-old female showing an obstructive mass at the foramen of Monro

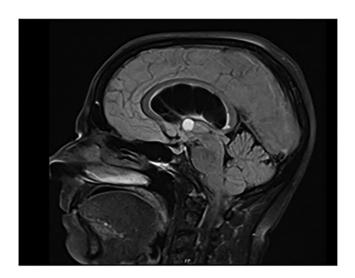


FIGURE 5 Sagittal view of magnetic resonance imaging brain scan showing an obstructive mass and lateral ventriculomegaly

physician's suspicion for an intracranial lesion such as a colloid cyst of the third ventricle. One such finding in the patient's history may be waxing and waning symptoms. Because of the intermittent obstruction of the flow of CSF, patients may have decreased mentation, vision difficulties, headaches, nausea, and gait ataxia, which improve over a period of time, corresponding to the actual obstruction by the colloid cyst.⁸ In the case of a known past diagnosis, this should prompt a consideration of MRI use and a team-based approach with neurology and neurosurgery consults early in the ED stay with a goal of rapid identification of the cyst to ultimately reduce the morbidity and mortality of the patient.

CONFLICT OF INTEREST

None.

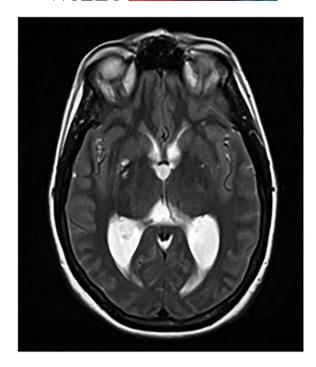


FIGURE 6 Magnetic resonance imaging brain scan of 54-year-old female showing an obstructive mass

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