Neurosurgical management of a giant colloid cyst with atypical clinical and radiological presentation

Case report

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The finding of a colloid cyst on neuroimaging is often incidental. These lesions are usually located at the foramen of Monro, are hyperdense on CT scans, and generally demonstrate signal intensity of water on MR images, although this depends on their content. When symptomatic, they frequently present with headaches and nausea due to an obstructive hydrocephalus. The authors describe a case of a giant colloid cyst in a patient presenting with complete left-sided vision loss and progressive memory loss, two very atypical findings in colloid cyst presentation. Imaging findings were also atypical, and this case proved to be a diagnostic dilemma because of its clinical and radiological presentation. Histopathological investigation was of utmost importance in the final diagnosis of a colloid cyst. To the authors’ knowledge this colloid cyst is larger than any other described in the literature.

Key Words • intracranial colloid cyst • intraventricular neoplasm • neuroradiology • neurosurgery • neuropathology • oncology

Colloid cysts are benign intracranial lesions usually located in the anterosuperior part of the third ventricle. Rarely, they arise from the lateral ventricles, fourth ventricle, or outside the ventricular system. Between 40% and 50% are asymptomatic and account for approximately 0.5%–1% of intracranial tumors. A large colloid cyst is usually defined as > 3 cm and is extremely rare. Few cases have been reported of so-called huge colloid cysts that are > 5 cm. We present a case of a giant 8-cm third ventricle colloid cyst; larger than any other reported in the literature, it was a diagnostic challenge due to its atypical clinical presentation and unique imaging appearance.

Case Report

History and Examination. A 42-year-old woman presented to the emergency department with acute onset of complete vision loss in her left eye that later resolved. She also reported a 6-month history of progressive memory loss, left hemiparesis, and an unsteady gait. On physical examination she had an ataxic, unsteady gait, and funduscopic examination showed bilateral papilledema.

Neuroimaging. Contrast-enhanced head CT scans demonstrated a large cystic mass, probably within the ventricular system. It measured approximately 8 cm in maximum diameter, lacked calcification, contained a small area of enhancement, and was causing obstructive hydrocephalus (Fig. 1A and B). A contrast-enhanced MRI study of the brain showed a largely cystic lesion arising from the third ventricle that was predominantly T2 hypointense (Fig. 1C) and T1 hyperintense, with an enhancing nodule (Fig. 1D and E). It demonstrated a focus of restriction on diffusion-weighted imaging and foci of blooming, suspicious for hemorrhage on susceptibility-weighted imaging (Fig. 1F and G). The differential diagnosis at that time included craniopharyngioma, ependymoma, neuroenteric cyst, teratoma, cystic intraventricular cavernous malformation, and a large colloid cyst.

Operation. A left frontal craniotomy with resection of the cystic lesion was performed. Intraoperatively the cyst wall was very tough, and a thick “anchovy paste”-type substance was drained. Under intraoperative mi-
croscopy, the cyst was further decompressed and its wall was excised. There was a small nodular segment that was quite adherent to the deep venous system, and this was carefully dissected in a subtotal fashion to preserve venous flow.

Pathological Findings. Histological evaluation demonstrated a cystic structure, lined by a single layer of epithelium resting on a basement membrane lying on a thin layer of connective tissue (Fig. 2A). The contents of the main cyst were altered by acute and chronic hemorrhage; however, in small microcysts in the cyst wall, colloid content, brightly stained by PAS-D, was demonstrated (Fig. 2B and C). Focally the epithelium demonstrated its ciliated, columnar nature (Fig. 2D). The epithelium expressed cytoplasmic low- and high-molecular-weight cytokeratins (Fig. 2E), epithelial membrane antigen (Fig. 2F), and focally nuclear S100 protein (Fig. 2G). There was no expression of glial fibrillary acidic protein or carcinoembryonic antigen. Lymphocytes, epithelioid histiocytes, and occasional plasma cells were demonstrated in the underlying connective tissue, indicating chronic inflammatory changes. There was no evidence of malignancy.

Postoperative Course. Postoperatively the patient developed generalized tonic-clonic seizures from which she recovered. She made a satisfactory recovery and on follow-up was neurologically intact. Postoperative imaging 1 year after surgery demonstrated a tiny, partially enhancing intraventricular lesion involving the septum pellucidum, without hydrocephalus.

Discussion

Colloid cysts are benign, slow-growing tumors that account for 0.5%–1% of primary brain neoplasms and 15%–20% of all intraventricular masses. Colloid cysts occur more commonly in men and are usually diagnosed between the 3rd and 5th decades, although there have been some reported cases in children. Histopathologically, the cyst is usually lined by simple to pseudostratified columnar or cuboidal epithelium. Colloid cysts are usually filled with various gelatinous materials such as blood products, mucin, or cholesterol crystals. Bertalanffy et al. reported a large colloid cyst of similar size; however, the histopathological examination could not identify any epithelial lining, and this makes our case the largest reported colloid cyst with typical epithelial lining.

Colloid cysts may be incidentally discovered on imaging. Clinical presentations and the majority of symptoms, however, can be explained by the raised intracranial pressure caused by the obstruction of the foramen of Monro. The most commonly reported symptom is headache, which is usually described as intermittent, se-
vere, and intense—it is relieved by lying down, is of short duration, and is usually frontal in location. Our patient presented with very atypical symptoms of an acute onset of complete vision loss in her left eye against a background of a 6-month history of progressive memory loss, left hemiparesis, and an unsteady gait. This provided a diagnostic dilemma with respect to the etiology of her symptoms and prompted further imaging investigation.

On imaging, colloid cysts typically arise from the foramen of Monro. On CT scans they are usually nonenhancing, well-defined hyperdense masses because of their heterogeneous and mucinous contents. Their MRI appearance can vary depending on their intracystic content, but they are typically nonenhancing lesions that are hyperintense on T1-weighted imaging and isointense to brain on T2-weighted imaging. Colloid cysts rarely hemorrhage, and such bleeding has been associated with rapid clinical deterioration. Colloid cysts do not typically demonstrate enhancing components, with rim enhancement being the only common feature. Colloid cysts range in size from a few millimeters to 3 cm, with a mean size of 1.5 cm. The radiological appearance of the colloid cyst in our patient was extremely unusual—measuring 8 cm, it is the largest colloid cyst ever described in the literature, and it was a lobulated, extremely heterogeneous mass with hemorrhage and an enhancing nodule (all atypical imaging findings).

Management of colloid cysts should be performed by a multidisciplinary team including professionals from neurosurgery, neurology, radiology, nursing, physical therapy, occupational therapy, and rehabilitation services. Although observation can be considered in some patients with colloid cysts who are asymptomatic, resection remains the optimal treatment.

**Conclusions**

This case proved to be a diagnostic challenge from both a clinical and imaging perspective. Clinically, the typical description of headache was not the predominant finding; our patient presented with acute onset of complete vision loss in her left eye, with a 6-month history of progressive memory loss, left hemiparesis, and an unsteady gait. The radiological appearance of an 8-cm lobulated, heterogeneous lesion probably located within the ventricular system, with an enhancing and hemorrhagic component, was so atypical for a colloid cyst that it was initially not considered high in the differential diagnosis. Pathological examination following resection, however, revealed this lesion to be the largest and one of the more unusual colloid cysts when compared with those described in the literature.

**Disclosure**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.
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