

Pure Endoscopic Management of Fourth Ventricle Arachnoid Cyst: Case Report and Literature Review

Abstract

Arachnoid cysts (ACs) within the fourth ventricle are rare, and only a few cases have been reported in the literature. These are benign lesions within the arachnoid membrane, and they have been reported to occur in almost all locations where arachnoid is present. Different procedures have been performed to restore a normal cerebrospinal fluid dynamic and/or pressure, including shunting and partial or complete excision of the cyst by open microsurgery or endoscopic fenestration. We report the case of a fourth ventricle AC successfully treated using only endoscopic anterior trans-frontal cyst fenestration/marsupialization and standard third ventriculostomy. Clinical and technical features are discussed, along with the pertinent literature.

Keywords: Arachnoid cyst, endoscopy, fenestration, fourth ventricle, marsupialization

Introduction

Arachnoid cysts (ACs) are common anomalies of the central nervous system constituting about 1% of all nontraumatic intracranial space-occupying lesions. They are benign, congenital, nonneoplastic, extra-axial, arachnoidal lesions filled with fluid similar to cerebrospinal fluid (CSF). However, ACs in the fourth ventricle are particularly uncommon; the first report was published in 1979 on a pediatric patient.^[1] Since then, to the best of our knowledge, only 12 cases have been reported to date [Table 1]. The incidence of AC in the general population is estimated to be around 0.1% from autopsy series.^[4] The incidence is similar in computed tomography (CT) imaging series (0.2%)^[8] and greater in magnetic resonance imaging (MRI) series (0.8–1.7%).^[11] The pathologic hallmarks of ACs have generally been established, but their pathogenesis is still controversial and debated. The occurrence of cysts in neonates and siblings, the anatomic relationship of cysts to the cisterns, and the association of ACs with other developmental anomalies all lend credence to the supposition that these cysts are usually developmental in origin and not acquired from other pathologic conditions. ACs are most commonly present during childhood. About 75% of intracranial ACs

present before 3 years of age. Typical manifestations of AC of the fourth ventricle are chronic hydrocephalus, vertigo, ataxia, and less frequently, symptoms related to brain stem compression. In the neurosurgical literature, considerable debate continues regarding whether it is best to place a shunt, excise, fenestrate, or undertake a nonsurgical observational approach. We report our experience with a patient harboring a large fourth ventricle AC causing hydrocephalus and thus progressive increased intracranial pressure (ICP).

Case Report

We report the case of a 55-year-old woman, otherwise healthy, presenting to our outpatient clinic with a 6-month history of progressive deterioration of short-term memory associated with loss of balance, walking problems, and repeated episodes of falling. An urgent brain CT scan followed by MRI showed a dilated fourth ventricle due to an AC causing tri-ventricular hydrocephalus with trans-ependymal CSF resorption [Figure 1]. The patient was immediately admitted, whereupon neurological examination revealed a Mini-Mental State Examination (MMSE) score of 22 with light disorientation in time and space and unsteady waking with paraparesis globally estimated at 3/5 (MRC). After patient information

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Table 1: Literature review

| Author (reference) | Age | Symptoms | Procedure | Outcome |
|--|-----------|--|--|---|
| Di Rocco <i>et al.</i> ^[1] | 7 years | Chronic hydrocephalus, delayed mental and physical development, worsening of preexisting hemiparesis | VP shunt Complete excision | Temporary improvement Good outcome with persistent slight hemiparesis |
| Korosue <i>et al.</i> ^[2] | 67 years | Chronic hydrocephalus | VP shunt Complete excision | Improvement of urinary function persistent of intellectual impairment Good outcome |
| Turgut <i>et al.</i> ^[3] | 29 years | Chronic hydrocephalus | Complete excision | Progressive improvement of symptoms |
| Nadkarni <i>et al.</i> ^[4] | 18 months | Delayed mental and physical development, ataxia, signs of raised ICP, vomiting | VP shunt Partial excision | Symptoms of raised ICP improved, ataxia worsened Slow improvement after rehabilitation |
| Azzimondi <i>et al.</i> ^[5] | 68 years | Chronic hydrocephalus | Subtotal excision | Progressive ataxia, persistent urinary incontinence and memory impairment |
| Acar <i>et al.</i> ^[6] | 73 years | Normal pressure hydrocephalus | Complete excision | Died from ischemic stroke; size of the fourth ventricle had normalized postoperatively |
| Szucs <i>et al.</i> ^[7] | 32 years | Depression, signs of acute occlusive hydrocephalus | Subtotal excision | Good outcome |
| Bonde <i>et al.</i> ^[8] | 27 years | Headache, truncal ataxia | Complete excision | Good outcome |
| | 3 years | Headache, vomiting, drowsiness, ataxia | VP shunt Partial excision | Symptoms of raised ICP improved, ataxia worsened Good outcome |
| | 52 years | Headache, vomiting, truncal ataxia | VP shunt Partial excision | Good outcome Symptoms of raised ICP improved, ataxia worsened Good outcome |
| Westermaier <i>et al.</i> ^[9] | 34 years | Progressive vertigo, slight ataxia | Complete excision | Good outcome |
| Martinoni <i>et al.</i> ^[10] | 32 years | Normal pressure hydrocephalus | Endoscopic cyst marsupialization and 3 rd ventriculostomy | Good outcome |
| Present case | 55 years | Progressive loss of memory, unsteady gait, paraparesis | Endoscopic cyst marsupialization and 3 rd ventriculostomy | Good outcome |

VP – Ventriculoperitoneal; ICP – Intracranial pressure

and consent, surgery was performed by pure anterior trans-frontal endoscopic cyst fenestration/marsupialization and standard third ventriculostomy. The wake-up was uneventful; the patient improved rapidly and can be discharged home on the seventh postoperative day. Four weeks later, clinical follow up showed a patient in a good condition able to walk independently, and her MMSE score had improved from 22 to 28/30. Brain MRI checks at 2, 6, and 12 months showed a progressive reduction in the size of the cyst and aqueduct as well as of the ventricular cavities [Figure 2]. Patient follow-up has continued for 22 months, with all checks showing neurological stability; neither delayed hydrocephalus and oculomotor problems nor hypothalamic dysfunction has, so far, been recorded, and the patient reports a generalized sense of well-being.

Surgical Technique

The procedure is usually performed with the patient under general anesthesia and in a supine position. The head is

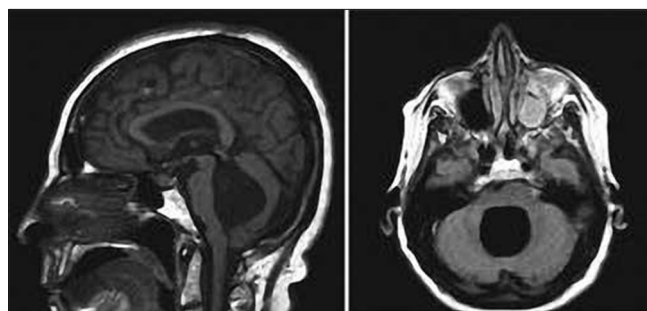


Figure 1: Sagittal and axial magnetic resonance imaging T1-Weighted showed a dilated fourth ventricle due to an arachnoid cyst causing tri-ventricular hydrocephalus with trans-ependymal cerebrospinal fluid resorption

lightly flexed. The operation is planned and completed with the aid of a rigid endoscope of 0° and 30° using a free-hand technique. We used the Hopkins II optical system (Endoscopy-America, Charlton, MA), with an operative channel by Karl Storz (Tuttlingen, Germany), 30 cm long and 2.9 mm in diameter. An Image-Guided

Surgery (IGS) system was used (BrainLab VectorVision). A single frontal right-sided burr hole is performed, and its position was chosen and verified by the IGS to obtain a trajectory to reach easily the aqueduct, the fourth ventricle, and the floor of the third ventricle simultaneously. The operative approach consists of a linear (1.5 cm) right frontal incision placed rather anteriorly, possibly along the hairline [Figure 3], to easily direct the endoscope toward the posterior portion of the third ventricle and aqueduct.^[12] Following placement of the burr hole, dural coagulation, and pial piercing, the endoscope is advanced toward the lateral ventricle; after entering the foramen of Monro, the endoscope direction is readjusted and following the ventricular landmarks and the indications of the neuronavigation system, aimed toward the aqueduct and then the fourth ventricle. The cyst is identified and after visual inspection, a careful coagulation of its surface is performed aiming for as large a fenestration as possible, completed by forceps [Figure 4]. Following cyst marsupialization, a standard endoscopic third ventriculostomy was done via the same hole/approach to treat the hydrocephalus.

Discussion

ACs within the fourth ventricle have rarely been reported in the literature. It is reasonable to think that ACs are congenital anomalies resulting from a duplication of the arachnoid membrane during embryological arachnoid cistern formation.^[13,14] Frequently, ACs are found incidentally; when clinical symptoms arise, they are directly in relation to the cyst location. MRI is currently the diagnostic gold standard for such a pathology as it allows detailed study and analysis of the various cysts features. MRI flowmetry is most important to delineate the flow of CSF, which can differentiate an AC from a trapped fourth ventricle. Ventriculoperitoneal (VP) shunting can provide transient or incomplete improvement of symptoms,^[1,2,8] and a third ventriculostomy may only release ICP in occlusive hydrocephalus.^[2] However, unchanged or even worsened signs of cerebellar or brainstem dysfunction following VP shunts have been reported,^[1,2,4,8] suggesting that the CSF diversion procedure does not constitute a durable solution. Currently, endoscopic fenestration is a worldwide accepted procedure for managing ACs in many locations except within the fourth ventricle. A fourth ventricle AC may be approached either via the third or the fourth ventricle.^[15] Endoscopic fenestration of a fourth ventricle AC cyst has already been described with good postoperative results, although, until date, there have been only a few attempts to treat such a pathology in a purely endoscopic fashion. The first case of a fourth ventricle AC successfully managed by an endoscopic procedure alone via the third ventricle was recently reported by Martinoni *et al.*;^[10] the former technique has the advantage of allowing both a cyst fenestration and a third ventriculocisternostomy at the

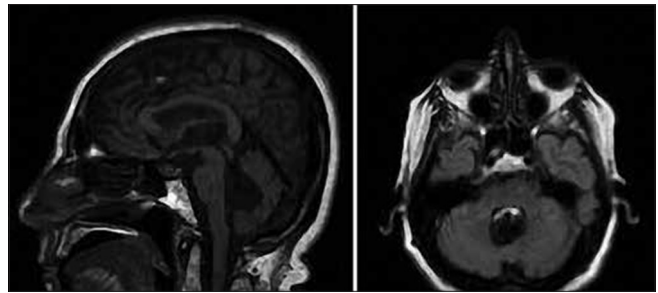


Figure 2: Sagittal and axial magnetic resonance imaging T1-Weighted showed a progressive reduction in size of the cyst and aqueduct as well as of the ventricular cavities

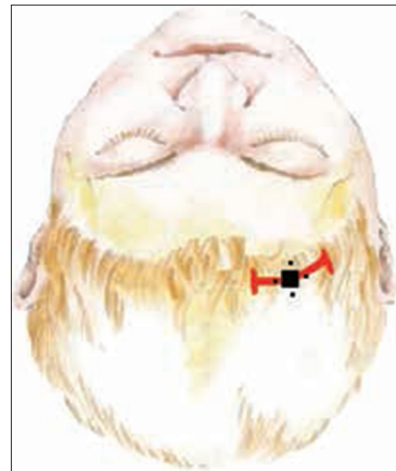


Figure 3: Linear (1.5 cm) right frontal incision placed anteriorly on a frontal wrinkle or along the hairline

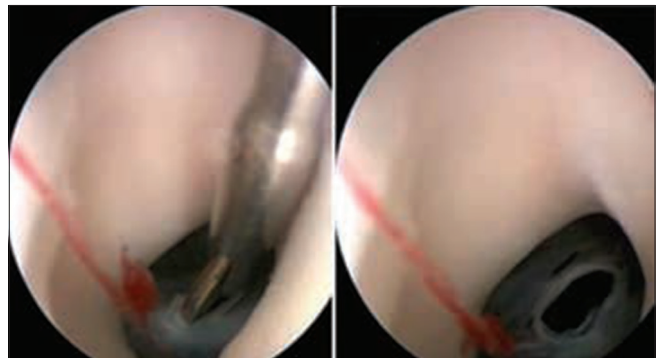


Figure 4: Endoscopic photos showing cyst fenestration/marsupialization and third ventriculostomy

same stage. This procedure might prevent the occurrence of hydrocephalus in case of impeded free pericystic CSF circulation or cyst recurrence, representing a remarkable advantage compared to simple CSF diversion. In fact, a case of sudden death in a fourth ventricle AC has been reported due to shunt failure.^[16] From our experience, we believe that, when possible, pure endoscopic cyst fenestration/marsupialization and third ventriculostomy could represent a valid and less invasive option to manage such a pathology as an alternative to cyst open complete or partial microsurgical resection. However, the latter

should still be considered in case of tonsillar herniation; it is also important to consider that the former technique presents a few limitations as follows: (1) The absence of hydrocephalus (ventriculomegaly constitutes a crucial anatomical condition), (2) the presence of a narrow (not dilated) aqueduct, and (3) the presence of intracystic septation, each of which make endoscopic AC fenestration/marsupialization not possible. Finally, it is worth of noting that open microsurgical procedures have a globally higher postoperative morbidity related to vermis manipulation/splitting.

Conclusion

Fourth ventricle ACs are rare entities and sometimes either mistaken or misdiagnosed as communicating hydrocephalus. MRI is the most important diagnostic tool, which can help to identify and analyze cysts in detail. Different procedures have been performed to restore normal CSF dynamic and pressure, including shunting and partial or complete excision of the cyst by open microsurgery. Endoscopic fenestration is now considered the procedure of choice for the treatment of symptomatic ACs in other locations, and the procedure could constitute an effective, less invasive, less morbid, and elegant way to manage the fourth ventricle AC as well.

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Conflicts of interest

There are no conflicts of interest.

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