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Original Article

Surgery Section

Endoscopic Management of Intracranial Arachnoid Cysts: A Retrospective Study

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ABSTRACT

Introduction: Endoscopic method is an alternative to shunt placement and open surgery; it creates a communication between cyst, ventricular system and subarachnoid system.

Aim: To describe the demographic, clinical profile, operative technique, complications and outcome following endoscopic management of intracranial arachnoid cysts at a tertiary Care Hospital in Southern India.

Materials and Methods: A retrospective observational study was conducted in tertiary care hospital in southern India in which case records of all 42 symptomatic patients diagnosed with intracranial arachnoid cyst and who underwent surgical management by endoscopic techniques from 1995 to 2010 were included. Demographics, clinical presentations, type of surgery, complications and clinical outcome were recorded and results were expressed in terms of frequency and percentages. The data was entered in Microsoft excel was imported to IBM SPSS version 20.

Results: Study included 30 males and 12 females with a mean (SD) age of 14.8 (15.5) years. The most common presenting feature was raised intracranial pressure (38.1%). Cysts were commonly located in posterior fossa (31.0%) followed by intraventricular region (26.2%). All patients (100%) underwent endoscopic fenestration with either cystoventriculostomy or cystocisternostomy. Endoscopic Third Ventriculostomy (ETV) was done in 21.4% of patients associated with hydrocephalus. Postoperative complications (11.9%) included Cerebrospinal Fluid (CSF) leak, subdural hygroma and metabolic complications. Mortality rate was 2.4%. Follow-up period ranged between 1-99 months (median 24 months). Success rate of the procedure was 95%.

Conclusion: Prevalence of intracranial arachnoid cyst is common in males and is often located in the posterior fossa. The endoscopic management of intracranial arachnoid cyst is safe and is associated with superior clinical outcome.

Keywords: Cystocisternostomy, Cystoventriculostomy, Endoscopic third ventriculostomy, Fenestration

INTRODUCTION

Arachnoid cysts are extra parenchymal, intra-arachnoidal collection of fluid within the CSF cisterns and major cerebral fissures bordered by arachnoid membrane. Arachnoid cyst was first described by Richard Bright in 1831, it accounts for 1% of all intracranial space occupying lesions [1,2]. Prevalence of intracranial arachnoid cyst among the paediatric and adult population is estimated to be 2.6% and 1.4%, respectively [3]. Most cysts are congenital, appear to originate from alteration in CSF flow in the early phase of subarachnoid space formation; while, the acquired cysts are secondary to neurological insults including, head injuries, meningitis, haemorrhage or tumours [4].

Although, most of these are asymptomatic, the raised intracranial pressure or local mass effect on adjacent structures result in symptoms such as headache, nausea, vomiting, visual impairment seizures and abducens nerve palsy, which necessitates surgical decompression [5]. Management of intracranial arachnoid cyst is variable, ranging from conservative approach with serial imaging, medical management with acetazolamide or surgical decompression through craniotomy, shunt placement, or fenestration of cyst (endoscopic or open) [6]. The use of endoscopic method to create communication between cyst and ventricular system and subarachnoid system offers attractive alternative to traditional shunt placement and open surgery. Previous studies have suggested that neuro-endoscopic techniques and instrumentation is safe, effective and is associated with better clinical outcome [7,8]. Moreover, it's cheaper, associated with fewer complications and shorter hospital stay [9].

As there are limited reports on endoscopic management of intracranial arachnoid cyst in southern India, the present study was carried out with an objective to describe the demographic, clinical features, operative technique, complications and outcome (success or failure of procedure) following endoscopic management of intracranial arachnoid cysts at a tertiary Cancer Hospital in Southern India.

MATERIALS AND METHODS

A retrospective observational study was conducted in National Institute of Mental Health and Neuro Sciences, Bengaluru, India, from December 1995 to December 2010. All patients or guardians (in case of paediatrics patients) had signed an informed consent form before the procedure. The duration of the data analysis was from December 1st 2020 to May 31st 2021. Considering the retrospective nature of the study, ethics committee clearance was deemed not necessary at the time of the study.

Inclusion and exclusion criteria: The study included all patients who underwent endoscopic management of the intracranial arachnoid cyst by different surgeons. All the patients below 60 years who presented with raised Intracranial Pressure (ICP) or focal neurological deficits without significant co-morbidities were included in the study. Those patients who were not fit to undergo procedure under general anaesthesia and asymptomatic smaller arachnoid cysts which did not cause mass effect were excluded from the study.

Study Procedure

The main objective of the procedure was to marsupialise the arachnoid cyst in to normal basal cisterns and ventricles, depending on the anatomical features. In patients with a combination of arachnoid cyst and hydrocephalus, ETV or Cystoperitonial Shunt (CPS) was carried out in addition to the endoscopic technique, whenever required. A rigid endoscope (Carl Zeiss 4 mm) was used for the procedure. We attempted the fenestration using bipolar electrode and dilated with Fogarty catheter. Based on the

anatomical locations, the entry point was selected according to the best trajectory determined after proper assessment of Computed Tomography (CT) or Magnetic Resonance (MRI) images. For suprasellar cysts, a right frontal burr hole was made just anterior to coronal suture and 3 cm off the midline. In children with an open fontanelle, the entry point was at the lateral margin of open anterior fontanelle. For posterior fossa cysts, retro mastoid sub occipital burr hole was made. We used precoronal frontal burr hole for Intraventricular arachnoid cyst, temporal burr hole for sylvian fissure cyst and interhemispheric arachnoid cyst was approached through frontal burr hole.

Upon entering the arachnoid cyst, all patients with cysts in the suprasellar region underwent cystoventriculostomy followed by cystocisternostomy; while patients with arachnoid cyst in the posterior fossa, sylvian fissure and interhemispheric cysts underwent cystocisternostomy. Cystoventriculostomy was a preferred choice for intraventricular arachnoid cysts. At the end of the procedure wide opening with pulsating CSF between arachnoid cyst and prepontine cistern was confirmed. Postoperative CT head was performed in all patients before the time of discharge. Patients were followed up for a median of 24 months (1-99 months). Follow-up data was available in 70% patients.

Details on demographic characteristics including age, sex; details on clinical presentations including site, symptoms at presentation, duration of symptoms and diagnosis based on the radiographic findings of CT and MRI images of the lesions; surgery related factors including previous history of surgery, preoperative clinical status, type of surgery and operative procedure; intraoperative and postoperative complications and its outcome, clinical outcomes were recorded as improved, unchanged or deteriorated based on subjective improvement in symptomatology, neurological examination and radiological findings.

STATISTICAL ANALYSIS

The data was entered in Microsoft excel was imported to IBM Statistical Package for the Social Sciences (SPSS) version 20 for statistical analysis. Continuous variables were expressed as mean and standard deviations; while, categorical variables were expressed as frequency and percentages.

RESULTS

A total of 42 patients (males: n=30, 71.4%; females: n=12, 28.6%) were included for the final analysis. Patients belonged to the age range of two months to 55 years with a mean age of 14.8±15.5 years. Most common symptom at presentation was raised ICP 16 (38.1%) followed by increasing head circumference 14 (33.3%). Cysts were commonly located in posterior fossa 13 (31.0%) followed by intraventricular region 11 (26.2%). Two patients (aged 4 and 9 years) had presented with bobble head doll syndrome [Table/Fig-1].

Demographic and clinical characteristics	Frequency
Age in years	
Mean (SD)	14.8 (15.5)
Median (min-max)	9.5 (0.2-55)
Age; n (%)	
0 to <1 year	3 (7.1)
1 to 3 years	11 (26.2)
4 to12 years	10 (23.8)
13-17 years	2 (4.8)
≥18 years	16 (38.1)
Gender; n (%)	
Male	30 (71.4)
Female	12 (28.6)
Symptoms at presentation; n (%)	
Increasing HC	14 (33.3%)
Raised ICP	16 (38.1)
Delayed milestones	6 (14.2)
Seizures	8 (19.0)
Focal neuro deficit	8 (19.0)
Others	6 (14.2)
Duration of symptoms in months	
Mean (SD)	8.3 (9.5)
Median (min-max)	5 (1-36)
Diagnosis; n (%)	
Tumour with/without hydrocephalus	1 (2.4)
Intraventricular cysts with hydrocephalus	5 (11.9)
Arachnoid cyst	33 (78.6)
Congenital hydrocephalus with arachnoid cyst	2 (4.8)
Arachnoid cyst with shunt malfunction	1 (2.4)
Anatomic locations of the cyst; n (%)	
Intraventricular	11 (26.2)
Posterior fossa	13 (31.0)
Sylvian	5 (11.9)
Suprasellar	9 (21.4)
Intraventricular+Posterior fossa+Interhemispheric	1 (2.4)
Intraventricular+Posterior fossa	1 (2.4)
Others	2 (4.8)

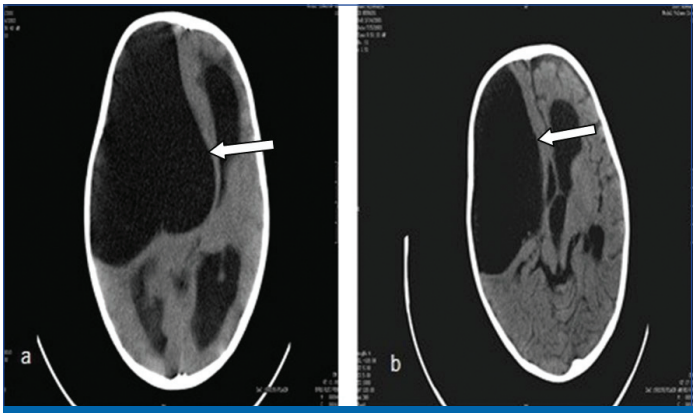
[Table/Fig-1]: Demographic features and clinical characteristics of the study participants.
SD: Standard deviation; Min: Minimum; Max: Maximum; n: Number; HC: Head circumference; ICP: Intracranial pressure

Most of the patients 35 (83.3%) had no previous history of surgery. Almost all patients 39 (92.9%) were conscious and alert before the surgery. All 42 patients underwent CT scan and 38 patients underwent MRI scan before surgery. All patients underwent endoscopic fenestration with either cystocisternostomy or cystoventriculostomy or both. Additionally, ETV to divert the CSF (particularly in posterior fossa and intraventricular arachnoid cysts) was performed in 9 (21.4%) patients and endoscopic placement of CPS was done in 1 (2.4%) patient [Table/Fig-2].

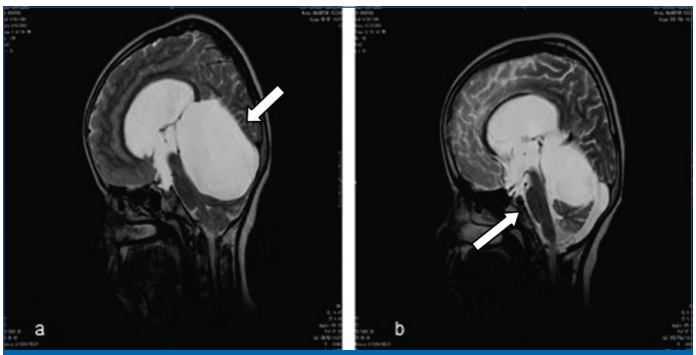
Surgery related factors	Frequency n (%)
Previous history of surgery	
Yes-endoscopy	2 (4.8)
Shunt placement	5 (11.9)
No	35 (83.3)
Preoperative clinical status of the patient	
Conscious, alert	39 (92.9)
Drowsy, arousable	1 (2.4)
Altered sensorium	2 (4.8)
Type of surgery	
Endo assisted microsurgery	32 (76.2)
ETV for CSF diversion	9 (21.4)
Shunt placement	1 (2.4)
Type of procedure	
Biopsy and or decompression	1 (2.4)
Cyst excision/cysternostomy/fenestration of loculi	37 (88.1)
Cyst excision/cysternostomy with aqueduloplasty	2 (4.8)
Cyst excision/cysternostomy with endo shunt	1 (2.4)
Cyst excision/cysternostomy with septostomy	1 (2.4)

[Table/Fig-2]: Distribution of patients based on previous history of surgery, type of surgery and procedure.
CSF: Cerebrospinal fluid; ETV: Endoscopic third ventriculostomy

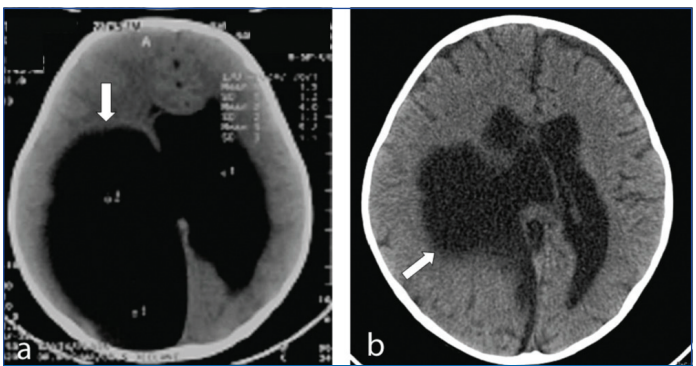
Intraoperative haemorrhage was noted in 3 (7.1%) patient which was controlled without requiring a craniotomy. Follow-up period ranged from 1-99 months (median of 24 months). All patients were investigated with CT head during follow-up and details regarding reversal of the symptoms, any fresh symptoms were noted. We defined success in the present study depending on the symptom reversal, lack of need for shunt and decrease in the size of the cyst based on imaging [Table/Fig-3-6]. Failure rate was 2 (4.8%) cases. One patient who was operated for posterior third ventricular arachnoid cyst had persistence of ventriculomegaly on follow-up at six months. This patient underwent Ventriculoperitoneal (VP) shunt and recovered well. Another patient was operated for posterior fossa arachnoid cyst. On follow-up at 13 months after surgery patient had again presented with raised ICP. Patient underwent re-exploration and endoscopic cystocisternostomy was done [Table/Fig-7].



[Table/Fig-3]: CT head shows: (a) Galassi type 3 sylvian fissure arachnoid cyst causing significant mass effect on ventricles and midline shift; (b) postoperative scan showing reduction in the size of the cyst following fenestration.



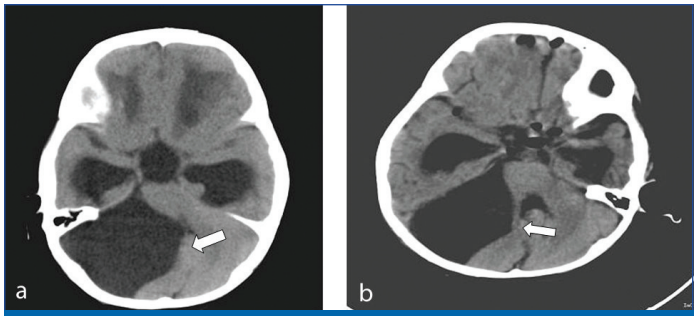
[Table/Fig-4]: MRI shows: (a) large quadrigeminal cisternal arachnoid cyst causing significant compression on the brainstem and cerebellum; (b) shows post op scan eight months following surgery. There is reduction in size of the cyst and mass effect on brainstem.



[Table/Fig-5]: (a) Intraventricular arachnoid cyst; (b) at four months follow-up after cystoventriculostomy.

DISCUSSION

Arachnoid cysts account for around 1% of intracranial lesions [1]. They are most probably developmental in nature and arise because of incomplete separation of perimedullary mesh during embryogenesis [10]. Alteration in the CSF flow during early phase of



[Table/Fig-6]: (a) Arachnoid cyst at cerebellopontine angle with squashing of 4th ventricle; (b) postoperative image following cystocisternostomy and opened up 4th ventricle.

Complications and outcome	Frequency n (%)
Intraoperative complications	
Bradycardia	2 (4.8)
Haemorrhage- significant but not requiring craniotomy	3 (7.1)
None	37 (88.1)
Postoperative complications	
CSF leak	2 (4.8)
Subdural collection	1 (2.4)
Metabolic complications	1 (2.4)
CSF leak+subdural collection	1 (2.4)
None	37 (88.1)
Outcome of complications	
Complete recovery	41 (97.6)
Death	1 (2.4)
Outcome of procedure	
Success	40 (95.2)
Failed endoscopy	2 (4.8)

[Table/Fig-7]: Intraoperative and postoperative complications and outcome of the procedure in the study population.
CSF: Cerebrospinal fluid

subarachnoid space formation leads on to formation of arachnoid cysts. According to the reports by Choi JU and Kim DS, arachnoid cysts develop following minor closed head injuries during infancy in some cases [11]. Arachnoid cysts especially interhemispheric cysts are associated with neurodevelopmental anomalies like partial or complete agenesis of corpus callosum, gyral abnormality [12,13]. Most of the arachnoid cysts are static fluid compartments, some increase in their size and exact pathogenesis is yet to be elucidated. Possible proposed theories include, ball valve mechanism suggesting a communication between the cyst and arachnoid space; osmotic gradient theory suggesting fluid transport between subarachnoid space and cyst; developmental malformation theory suggests trapped fluid content in cerebral agenesis to be arachnoid cyst; and hypersecretion of the neuroepithelium theory [14].

Many arachnoid cysts are recognised during the first two decades of life. In the present series, 61.9% patients were below 18 years of age which is lower than 75% reported by Deopujari CE et al., [10]. The male to female sex ratio in the present series was 2.5:1, which was comparable to previous reports [12,13]. Although, the most common site of arachnoid cysts is middle cranial fossa ranging from 25-80% [13,14], in present series posterior fossa was the most common location which could have been because of referral bias. Location of cyst also determines the success rate of the procedure. Linares Torres J et al., reported a higher success rate in intraventricular cysts followed by quadrigeminal cysts, suprasellar cysts and sylvian cysts [15]. In the present study, among three failed endoscopy procedures, two were seen in superstellar region and one in posterior fossa. Presence of concomitant hydrocephalus is common with arachnoid cysts [10]. In present study, the presence

of hydrocephalus was much lower than the prevalence reported by Linares Torres J et al., (17.7% vs 70%) [15].

Due to variable clinical presentation, indication for surgical treatment is debatable. According to Ali M et al., age at presentation (younger age) and size of the cyst (≥68 cm^3) are independent predictors of surgery [16]. Although, for seizures or headaches, medical management is preferred, surgery is definitive indication in patients with concomitant hydrocephalus, raised ICP or those with focal neurological symptoms [5]. The indication for surgery is uncertain in patients with seizures, headache or delay in developmental milestones, because a causal relationship between symptomatology and cyst is difficult to establish. Similar to Copley P et al., we performed surgery in symptomatic patients with increasing head circumference, raised ICP, delayed milestones, seizures and focal neural deficit [5]. In the present series, two patients of suprasellar arachnoid cysts had presented with bobble head-doll syndrome, which is a rare movement disorder characterised by episodic forward and backward movement of the head at 2-3 Hz frequency. These movements are absent during sleep and attenuate during volitional activities, occurs due to compression over dorsomedial nucleus of thalamus and dentatorubrothalamic pathway compression. Neuroimaging is essential for early identification of this disorder [17]. In the present study, both patients showed complete resolution of symptoms following fenestration procedure.

Previous studies have reported management with open craniotomy, cystoperitoneal shunting, endoscopic fenestration, microsurgery, each with its own risk and benefits [18,19]. Amelot A et al., observed no significant difference between the postoperative reduction of size and cyst volume between surgical procedures including microsurgery, endoscopic cyst fenestration, cytoperitoneal shunting or subdural shunting [13]. Event free survival was shorter with endoscopy group (67.7%), compared to CPS (71.5%) and microsurgery (90.5%). They suggested that microsurgery is effective treatment with longer event free survival and few complications. In the authors experience endoscopic management provides an effective modality for the treatment of arachnoid cyst in both children and adult population.

The primary goal of surgery in arachnoid cyst management is to establish a direct communication between the cyst and neighboring CSF spaces. We performed endoscopic assisted microsurgery in all patients. Although, complete excision of the cyst membrane is essential to avoid recurrence; however, close relationship between the membrane and underlying neural tissues and presence of large ruptured cysts limits the procedure. Hence, selective opening with limited resection of outer cyst wall is advised which limits the CSF access to subdural space and subsequent postoperative symptoms [1]. Similar to Duz B et al., they used burr hole as a working channel [20]. Based on the anatomic locations, patients with cysts in the suprasellar region underwent cystoventriculostomy followed by cystocisternostomy; while, patients with arachnoid cyst in the posterior fossa, sylvian fissure and interhemispheric cysts underwent cystocisternostomy. Cystoventriculostomy was a preferred choice for intraventricular arachnoid cysts. Furthermore, ETV for CSF diversion and shunt placement was performed in 21.4% and 2.4% patients, respectively. The frequency of cystoperitoneal shunt is much lower than 23.8% reported by Wang Y et al., [21]. ETV should be considered for those presenting with hydrocephalus secondary to intraventricular and posterior fossa arachnoid cysts.

Frequency of complications reported by Schulz M et al., was 3% and Karabatsou K et al., was 8% [9,22]. In the present study, complications were seen in 11.9% of patients which were effectively managed. However, one patient (2.4%) of suprasellar arachnoid cyst succumbed because of inadvertent hypothalamic damage during cystoventriculostomy due to intractable diabetes insipidus.

Schulz M et al., reported better reduction in acute symptoms than chronic symptoms [9]. In a study by Bir SC et al., clinical outcome was better in adults than children and those having communicative hydrocephalus than obstructive hydrocephalus [23]. We observed successful treatment in 40 (95.2%) patients, while treatment failed in 2 (4.8%) patients. One patient underwent VP shunt, while other underwent re-exploration and endoscopic cystocisternostomy. Failure rate and re-exploration is much lower than reported by Boutarbouch M et al., (29%), Schulz M et al., (16%), Deopujari CE et al., (7%) and Karabatsou K et al., (8%) [1,9,10,22]. Similarly, present success rate is much higher than the previous reports (60-92%) [18,23]. Present success rate is higher probably due to double fenestration technique. Additionally, selection of patients and a thorough preoperative evaluation also determines the clinical outcome.

Based on the results of the present study, we believe that endoscopic surgery has to be individualised to each patient depending on its anatomical location and features. Therefore, proper anatomical knowledge with utmost accuracy in operative technique is most important in achieving higher success rate. Endoscopic management in arachnoid cyst is safe but margin for error is narrow as more anatomical structures can be visualised due to free movement of endoscope and instruments within the large cystic cavities. Moreover, the ventricular CSF aids in better image transmission. Successful endoscopic surgery can prevent the need for shunt placement. Nonetheless, further prospective research with randomisation of surgical techniques and comparing the outcome in different anatomical location is warranted to understand the management of intracranial arachnoid cysts.

Limitation(s)

There are study related limitations including the inherent limitations of the retrospective review. Although, the sample size was adequate for the descriptive analysis, we could not perform interferential analysis due to lack of predefined grouping and non homogenised distribution. Moreover, preoperative volume of cyst and postsurgical reduction in size was not assessed. Long term (12-24 months) follow-up imaging was necessary to note the decrease in cyst volume which is in most cases is very slow. In present study, long term follow-up details were available in only 70% of patients.

CONCLUSION(S)

Minimally invasive endoscopic fenestration and marsupialisation is safer, and associated with superior clinical outcome. Proper knowledge of anatomical structures and utmost accuracy of operative technique prevented the complications and also reduces the need for additional procedures including CPS and VP shunt. Therefore, endoscopic management can be safely advised as a first line of treatment for symptomatic arachnoid cyst patients.

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