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Long Term Follow-up Results of Neuroendoscopic Management of Suprasellar Arachnoid
Suprasellar Araknoid Kistlerde Nöroendoskopik Tedavinin Uzun Dönem Sonuçları

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ABSTRACT

INTRODUCTION: Suprasellar arachnoid cysts (SACs) represent a unique subset of intracranial arachnoid cyst with their clinical presentation and diverse surgical treatment options. This study aims to delineate the long-term results of endoscopic ventriculocystocisternostomy (VCC) in suprasellar arachnoid cysts.
METHODS: A total of 22 suprasellar arachnoid cyst patients operated on with ventriculocystocisternostomy between 2013 and 2022 were enrolled in this study. Their clinical and radiological findings were recorded both pre and post-operatively. Clinical presentation, clinical improvement, cyst collapse ratio, and Evans index values (to evaluate hyrdocephalus) were recorded. Cyst collapse ratio and Evans index values were compared both between pre and post-operatively at last follow-up, and between different subtypes.
RESULTS: Ventriculocystocisternostomy showed excellent clinical result regarding increased intracranial pressure in 18 patients (82%). Three out of 4 patients that did not benefit from ventriculocisternostomy had previous surgical interventions. Success ratio in primary patients were 95%. While all 4 abducens nerve palsies recovered post-operatively, patients did not show progress in endocrine dysfunction. While Evans index values show significant decrease post-operatively ($p<0.05$), cyst collapse ratio and Evan index values did not differ among the morphologically described suprasellar arachnoid cyst subtypes ($p>0.05$).
DISCUSSION AND CONCLUSION: Ventriculocystocisternostomy may suggest best treatment modality for suprasellar arachniod cysts. Morphological subtypes of suprasellar arachnoid cysts did not show significant difference following surgery and their prognostic value is lacking.

Keywords: suprasellar arachnoid cyst, neuroendoscopy, ventriculocystocisternostomy, ventriculocystostomy

ÖZ

GİRİŞ ve AMAÇ: Suprasellar araknoid kistler klinik prezentasyonları ve cerrahi tedavilerindeki çeşitlilik nedeni ile diğer araknoid kistlerden ayrılırlar. Bu çalışmanın amacı ventrikülokistosisternominin suprasellar araknoid kist tedavisinde uzun dönem sonuçlarını değerlendirmektir.
YÖNTEM ve GEREÇLER: Kliniğimizde 2013-2022 yılları arasında ventrikülokistosisternomi yöntemi ile tedavi edilmiş 22 hasta çalışmaya dahil edildi. Hastaların klinik ve radyolojik bulguları ameliyat öncesi ve sonrası değerlendirildi. Klinik prezentasyonları, tedavi yanıtları, kistlerin küçülme oranları ve bir hidrosefali belirteci olarak Evans indeksi kullanıldı. Kist küçülme oranları ve Evans indekslerinin ameliyat öncesi ve ameliyat sonrası son kontroldeki değerleri karşılaştırıldı. Ayrıca bu değerler farklı suprasellar araknoid kist alt tipleri arasında karşılaştırıldı.
BULGULAR: Ventrikülokistosisternostomi tekniği 18 hastada artmış intrakraniyal basıncı kontrol etmede başarılı olmuştur (%82). Tedaviye yanıt vermeyen 4 hastanın üçünün öyküsünde daha önce geçirilmiş cerrahi tedavi vardı. Primer olgular için başarı oranı %95 idi. Abdusens siniri paralizisi olan 4 hastada bu bulgu ameliyat sonrası düzelirken, endokrin fonksiyon bozuluğu olan 4 hasta cerrahi tedaviden fayda görmedi. Evans indeksi değerleri ameliyat sonrası anlamlı düşüş gösterdi ($p<0.05$). Ancak Evans indeksi vekist küçülme oranları alt tipler arasında karşılaştırıldığında, grupral arasında anlamlı farklılık yoktu ($p>0.05$)
TARTIŞMA ve SONUÇ: Ventrikülokistosisternostomi tekniği mevcut cerrahi yaklaşımlar arasında suprasellar araknoid kist tedavisi için en uygun seçenek olabilir. Suprasellar araknoid kistler morfolojik olarak sınıflandırılmaya çalışılsa da, cerrahi tedaviye yanıtları açısından bu alt tipler arasında anlamlı farklılık bulunmamıştır ve prognostik değerleri net değildir.

Anahtar Kelimeler: suprasellar araknoid kist, nöroendoskopi, ventrikülokistosisternostomi, ventrikülokistostomy

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INTRODUCTION

Suprasellar arachnoid cysts (SACs) are non-tumoruos congenital cerebrospinal fluid (CSF) collections. They are believed to be developed from an abnormality of the Liliequist membrane of the interpedincular cistern, and they mainly produce symptoms either by obstructing CSF flow and causing obstructive hydrocephalus, or by compressing adjacent structures. They represent 9-21% of all arachnoid cysts, and 8-15% of arachnoid cysts that require treatment in childhood (1,2). Several explanations were given in terms of their pathogenesis including ball valve action, osmotic gradient, slit-valve mechanism, and transudation from choroid plexus remnants or cyst wall. Their special anatomical location makes them different from other arachnoid cysts in several aspects, especially in clinical features that can lead to obstructive hydrocephalus, endocrine dysfunction, or visual problems (3). There are also efforts on the classification of SACS, which will lead to appropriate management options, although no consensus has been reached to date (1,4). Recently, SACs are classified into 3 subtypes based on radiological features, although their role in prognosis is unclear (1).

The aim of this study is to delineate long-term clinical and radiological results of endoscopic ventriculocystocisternostomy (VCC) of SACs. A secondary aim is to try to clarify the role of SAC subtypes in clinical results and prognosis.

MATERIALS and METHODS

This retrospective study is by the Local Ethics Committee by decision number 09.2022.820, and performed in accordance with the 'Declaration of Helsinki'.

Patient selection

This study included patients who were diagnosed with SAC and treated with VCC in our clinic between 2013 and 2022. Exclusion criteria were: patients have been found to have other types of cystic lesions at surgery (epidermoid cyst, craniopharyngioma, etc), patients having other cystic intracranial lesions in conjunction with SAC, patients having metabolic or genetic disorders that may affect brain development, and history of intracranial hemorrhage.

Surgical technique

All endoscopic fenestrations were performed with the patient in the supine position, with the head flexed approximately 15-20°. The typical burr hole location was 3 cm lateral to the midline and just anterior to the coronal suture on the right side, although a pre-operative evaluation was carried out in each patient using magnetic resonance imaging (MRI) to carefully plan the entry site and optimal route to both fenestrate the apical and basal cyst membranes without excessive manipulation of the endoscope. In patients with previously had a shunt on the right side,

the contralateral side was used. A curvilinear incision was used in case a further shunt insertion would be necessary for the future. After entering the lateral ventricle, the apical cyst membrane was fenestrated and enlarged enough to secure future patency (ventriculocystostomy, VC). Then the cyst was entered and the basal part of the cyst was evaluated. The basal membrane or floor of the cyst was exposed and perforated until the abducens nerves and the prepontine cistern is clearly visible using a neuroballoon and scissors, which will complete dual opening and VCC.

Clinical and radiological follow up

All patients were evaluated by our neurosurgery team, ophthalmology, and endocrinology departments both pre and post-operatively. Past medical records, clinical charts, surgical reports, and picture archiving and communications systems were used in this study. Criteria for successful surgical treatment are composed of improvement of the patient's symptoms, absence of developmental delay and stabilization of excessive head circumference growth following surgical treatment in pediatric patients, and a good adaptation of the patient to life following surgery.

Cyst size was evaluated using maximum diameters in three MRI planes using CISS sequences (constructive interference in steady state) and reduction in cyst size or cyst collapse ratio was classified as less than 10%, between 10-50% and over 50% on long term follow up images. Evans Index (EI) is used to evaluate ventricle size and hydrocephalus.

According to the new classification system, patients were classified into 3 subtypes: SAC-1, SAC-2, and SAC-3 as described in the literature (1). Basically, SAC-1 represents the blockage of both Foramen of Monro and the diencephalic leaf of the Liliequist membrane was displaced caudally. SAC-2 represents prepontine expansion and the Liliequist membrane stayed rostrally. SAC-3 is the asymmetrical form with cysts expanding laterally into the temporopolar region or the Sylvian fissure. The collapse ratio of the cysts, pre and post-operative EI, and change in EI were analyzed between SAC subtypes and the whole study group to identify the prognostic value of the new classification and factors affecting good long-term prognosis. Since the classification system is described in a pediatric patient population and there is very little number of adult SAC cases reported in the literature, 4 adult patients were excluded from this analysis.

Statistical analysis

Patient data were first recorded in a Microsoft Excel spreadsheet (Microsoft Corporation, Redmond, Washington, USA). The data was then imported to SPSS (version 22, IBM Corporation, New York, USA) where statistical analysis was performed; $p<.05$ was considered statistically significant. To determine if there are statistically significant

differences between the different SAC subtypes, one-way ANOVA test was used for parametric variables while Kruskal-Wallis test was used for nonparametric variables. A paired-sample t-test was used to compare the mean pre-operative EI values versus the post-operative EI values.

RESULTS

Demographics

A total of 22 patients were included in this study. There were 19 males and 3 females. There were 18 pediatric and 4 adult patients. The median age at surgery was 48.5 months for the 18 pediatric patients, ranging between 9-173 months. Four adult patients were between 32 and 78 years old. The mean clinical follow-up duration was 43.8 months and the mean radiological follow-up duration was 36.0 months for the whole cohort.

Clinical presentation and outcomes

The most common presenting symptoms were related to increased intracranial pressure including headache, vomiting, and excessive head circumference growth (68%), followed by motor and mental retardation, a dysfunctioning shunt, abducens nerve palsy, epilepsy, endocrine dysfunction, and incidental. All but 4 of the patients showed either complete relief or marked improvement of their symptoms related to increased intracranial pressure (82%). Three of the four patients that did not benefit from endoscopic VCC include 3 patients which had previous ventriculoperitoneal shunts inserted in other institutions and their presenting symptoms were related to shunt dysfunctioning. Although VCC procedure was accomplished successfully, they showed shunt dependency and neither of them could be discharged without a functioning shunt. The remaining patient that did not benefit from endoscopic VCC had a fever post-operatively and was diagnosed with bacterial meningitis. He had external ventricular drainage immediately after surgery and multiple courses of antibiotherapy. He was not able to hold without external ventricular drainage at the end of his multiple courses of antibiotherapy and a ventriculoperitoneal shunt was inserted as a definitive treatment for hydrocephalus.

There were 4 patients having endocrine dysfunction in conjunction with increased intracranial pressure symptoms. Although they benefited from VCC procedure and had relief regarding increased intracranial pressure, their endocrinological status did not change. All continue their follow-up in the pediatric outpatient clinic and were still on the course of hormone replacements. Four patients had abducens nerve palsy pre-operatively and all of them showed marked improvement or full recovery immediately after VCC. One patient that presented with epilepsy was seizure free at his final follow-up.

Radiological and prognostic analysis

Regarding the cyst collapse ratio, there was no significant difference between SAC subtypes and the

whole study group (p=0.162, Kruskal-Wallis) (Table 1). Both pre and post-operative EI values were also not significantly different across all SAC subtypes and the whole study group (p=0.650 and p=0.812, respectively, One-way ANOVA) (Table 1). On the other hand, EI showed marked decrease post-operatively and mean post-operative EI was significantly less than the pre-operative value either for the whole study group or each individual SAC subtype (p<0.05 for each, paired sample t-test) (Table 1). Although EI evaluation showed a marked decrease, there was also no significant difference between the amount of reduction in EI (Δ EI) across all SAC subtypes and the whole study group (p=0.986, Kruskal-Wallis) (Table 1).

Spearman's rank correlation showed two significant correlations. Age at surgery was positively correlated with Δ EI at 0.05 level (2-tailed) and post-operative EI was positively correlated with pre-operative EI at 0.01 level (2-tailed) (Table 2).

Table 1. Realtions of Cyst Collapse Ratio, EI and Δ EI between SAC Subtypes and the Whole Study Group

	All Cysts	**SAC-1**	**SAC-2**	**SAC-3**	**p-value**
Cyst collapse ratio (no.)					**p=.162**
<10%	1	0	0	1	
10%<X<50%	5	2	3	0	
>50%	12	9	1	2	
EI Pre-op (mean±SD)	49.2±9.2	49.6±7.5	51.3±15.0	44.7±7.6	**p=.650**
EI Post-op (mean±SD)	44.9±10.1	44.8±10.4	47.3±11.0	42.0±11.1	**p=.812**
p-value	**P=.009**	**P=.014**	**P=.037**	**P=.022**	
Δ EI, median (range)	-2.0 (-23 to 1)	-2.0 (-23 to 1)	-1.5 (-12 to -1)	-3.0 (-6 to 1)	**p=.986**

EI: Evans Index, Δ EI: Evans Index post operative-Evans Index pre operative, SAC: suprasellar arachnoid cyst

Table 2. Correlation Matrix (Spearman's Rank Correlation)

	Age at surgery	Clinical follow-up	Radiological follow-up	EI Pre-op	EI Post-op	Δ EI
Clinical follow-up	-.127					
Radiological follow-up	-.173	**.957****				
EI Pre-op	-.289	-.197	-.112			
EI Post-op	-.115	-.259	-.229	**.824****		
Δ EI	**.484***	-.131	-.196	-.044	.384	
Cyst collapse ratio	.316	.448	.441	.048	-.119	-.020

*: correlation is significant at the 0.05 level (2-tailed), **: correlation is significant at the 0.01 level (2-tailed), EI: Evans Index, Δ EI: Evans Index post operative-Evans Index pre operative, SAC: suprasellar arachnoid cyst

DISCUSSION

Arachnoid cysts represent 1% of all intracranial masses, and their incidence is 2.6% among the pediatric population (5). SACs are also predominantly seen in the pediatric population and adult patients are rare (6) and mostly reported in small numbers in pediatric series (7,8). Our series includes 4 adult patient and 18 pediatric patient that is in concordance with the literature. There is a tendency for male predominance in SACs and our series shows a higher male-to-female ratio (19 male, 3 female) than reported in the literature (1,5,9).

Various surgical treatment options were adopted for SACs. Before the widespread use of neuroendoscope, microsurgical excision and cystoperitoneal shunting were the principle surgical treatments (8,10). There has been a paradigm shift toward neuroendoscopic management of SACs for the last two decades and today neuroendoscopy became the treatment of choice. Regarding the neuroendoscopic management of SACs, there are two options. Ventriculocystostomy (VC) consists of apical membrane fenestration alone and ventriculocystocisternostomy (VCC) consists of both apical and basal membrane fenestrations. There has been a debate about whether VC is sufficient or basal membrane fenestration is required (5,11,12). Although there is a lack of high-level scientific evidence comparing these two techniques, there is a tendency among neurosurgeons to choose VCC (5,9,12,13). It has been reported in a literature review that 16% of patients having VC require further surgery while 8% of patients having VCC require further intervention (13). Since VCC includes both apical and basal membrane fenestrations and connects the

ventricular system directly to the subarachnoid space in basal cisterns, VCC was used in all patients in our series. Of 19 primary patients treated with VCC in this series, 18 of them benefited regarding their symptoms of increased intracranial pressure and did not require further intervention throughout the follow-up period. The only primary patient that required ventriculoperitoneal shunting, suffered from meningitis following VCC procedure which may cause reduced CSF absorption, and transform the clinical picture to communicating hydrocephalus rather than obstructive hydrocephalus. The other 3 patients were already admitted with symptoms related to shunt dysfunctioning which were inserted previously in other clinics and they continue their shunt dependency although VCC procedures were completed uneventfully. In one of the largest series reported on SAC in the literature consisting of 247 patients, authors found that no history of previous treatment was an independent favorable factor for good prognosis(9). Although the current series include a limited number of patients, 3 out of 4 patients that did not benefit from VCC procedure had previous surgical treatments which may also indicate the possibility of a bad prognosis in patients with previous surgical treatment. These results show a 95% success rate for primary SAC patients and 82% overall for the current series, which are comparable with the current literature and may suggest that VCC can be the best modality of surgical treatment for SACs.

There are few efforts to classify SACs to date. They are mostly based on anatomical and morphological features that are present in imaging studies (1,4,5). Although they are useful in distinguishing imaging characteristics pre-operatively, their value in determining treatment options and prognosis is low. Based on our findings both SAC subtypes that were described by Andre et al. had a significant reduction in cyst size and also EI post-operatively (1). However, there was no significant difference between SAC subtypes or the whole study group. Although this classification system is based on the morphological aspects of the SACs and they were described to be related to the clinical presentation, its prognostic value is lacking based on our findings.

Reduction in cyst size is achieved almost always after VCC procedures (1,3,5,9,13-15). On the other hand, little has been reported regarding the ventricular size following VC and VCC. Our findings indicate a mild decrease in EI from 49.2±9.2 pre-operatively to 44.9±10.1 post-operatively which was significant. The only quantitative analysis regarding ventricular size found a mean 9.13% EI decrease in 1-year follow up and they showed that this decrease mainly occurred within the first 3 months following surgery (9). They found paraventricular edema as a good

prognostic factor for the decrease in ventricle size. They stressed the nature of brain compliance associated with long-term stress irritation and suggest that paraventricular edema implies acute hydrocephalus which is why this finding favors more decrease in ventricle size. Our findings on the other hand showed a positive correlation between the age at surgery and the decrease in EI which we also believe that this finding is concordant with paraventricular edema. While smaller children have open sutures and more tolerance to chronic hydrocephalus, older children and toddlers have sutures closed and less tolerance to chronic hydrocephalus. This may cause the positive correlation between age at surgery and decrease in EI.

While visual symptoms show a good response to surgical treatment in terms of ocular movements and visual field defects, visual acuity has a poor prognosis (5). Four of our patients in this series had abducens nerve palsy and all had a good response to VCC. Endocrinological problems encountered in SAC patients seem to have worse benefits from surgical treatments. The persistence of signs and symptoms related to the hypothalamo-hypophyseal axis was highlighted in several publications in the literature (5,15,16). Our results also indicate that surgical treatment does not resolve endocrinological dysfunction, at least during the follow-up period. It may seem logical to not primarily aim to resolve hormonal problems seen in SAC patients by neuroendoscopic or surgical interventions.

Conclusion

SACs constitute a unique subset of intracranial arachnoid cysts regarding their clinical picture and treatment options. VCC may suggest the best surgical option owing to its low morbidity and excellent results, however previous surgical treatment may show a poor prognosis. Although there are morphologically distinguishable subtypes of SACs pre-operatively, data regarding their clinical importance and response to surgical interventions is lacking.

Ethics Committee Approval: Marmara Univercty Clinical Research Ethics Committee (09.2022.820, protocol code , ethics committee number)

Authors Contributions:

Concept: M.S., A.D., Design: M.S., A.D., Supervision: A.D., Materials: A.D., M.S., Data Collection:M.S., Analysis: M.S., A.D., Literature search:M.S., Writing:M.S., Review:A.D.,

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