Research Article

Surgical management of Posterior Fossa Arachnoid Cysts

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Abstract
Posterior fossa arachnoid cysts are rare lesions that of congenital origin. 12 patients who underwent surgical treatment included. The common presentation were ataxia and headache. The diagnosis was based on computed tomography or magnetic resonance imaging. Cyst wall excision with fenestration was done in 9 cases and shunting procedure in three cases. In 11 cases, the postsurgical investigations showed decreasing in size of the ventricle, the cerebellum re-expanded, with decreased hydrocephalus, one case died. The period of follow up ranged between 6 months to 2 years. All patients are free of symptoms and imaging showed no recurrence. A review of literature was done including classification, pathophysiology, differential diagnosis and surgical management.
Key Words: Posterior fossa, arachnoid cysts**,** surgical treatment

Introduction
Arachnoid cysts are rare central nervous system lesions account for 1% of all intracranial masses. 25-35 of all arachnoid cysts are in the posterior fossa[1-3], it produces a range of clinical presentations. Its symptoms result from hydrocephalus or compression on the nearby structures. Management, include cyst wall excision with fenestration or cysto-peritoneal (CP) shunt. There is a controversy about the classification, pathophysiology, differential diagnosis (D.D) and its management. This study is retrospective study carried on 12 patients with symptomatic posterior fossa arachnoid cysts who underwent surgical treatment in Minia University Hospitals, Egypt, we discuss the classification, pathophysiology, D.D and surgical management of these lesion.

Methods
Between January 2012 and December 2015, from total 36 intracranial arachnoid cysts, 12 patients were presented with posterior fossa cyst. There were 6 males and 6 females, the age ranged from 12 months to 59 years. All patients had at least one C.T. or MRI at 6-12 months postoperatively and were followed up clinically for a period between 6 months to 2 years.

Results
Clinical presentation (Table 1)
The main complaint was gait ataxia and headaches. Enlarged head was the common finding in infancy. The duration of symptoms before diagnosis was short. One patient was in coma at the time of admission.

Table (1): symptoms, signs, operation and outcome for all patients in this study.

No	Age (yrs)/ Sex	Presentation	Neurologic Signs	Cyst location + hydrocephalus	Operation	Outcome	Follow-up
1	24/F	Headache	Papilledema	Retrocerebellar mild HCP	Cyst excision	No complaint	2 y
2	30/F	ataxia + vertigo	Trunkal ataxia + nystagmus	Retrocerebellar	Cyst excision	No complaint	2 y
3	35/F	1 headache	Papilledema	Superacerebellar mild HCP	Cyst excision	Small residual cyst, No. complaint	2 y
4	59 M	Headache, vertigo	Limb ataxia, left V, VII nerve palsy	Left CPA	Cyst excision	No complaint	2 y
5	29/M	Headache, ataxia	Trunkal + limb ataxia, nystagmus	Retrocerebellar mild HCP	Cyst excision	No complaint	1 y
6	50/M	Ataxia, nausea + vomity	Left VIII nerve palsy, limb ataxia	Intracerebellar mild HCP	Cyst excision	No complaint	2 y
7	31/F	Tinnitus in right ear	Right VIII nerve palsy	Right CPA	Cyst excision	No cyst, No complaint	2 y
8	38/M	Headache ataxia	Papilledema, trunkal ataxia	Retrocerebellar mild HCP	Cyst excision	No complaint	2 y
9	16/M (months)	Macrocephaly	Bulging fontanel	Retrocerebellar severe HCP	Y connected VP + CP shunt	Cyst Dec. no signs	2 m
10	17/M (months)	Macrocephaly	Bulging fontanel	Supracerebellar severe hydrophus	VP+CP shunt	Vent. size Dec. no signs	1.5 y
11	12/F (months)	Macrocephaly, vomiting	Sunset eye, bulging fontanel	Retrocerebellar severe HCP	Ext. Vent. drainage + cyst excision	Death	-
12	12/F (months)	Macrocephaly	Bulging fontanel	Retrocerebellar + severe HCP	VP+CP shunt	No signs	2 y

Radiological findings:

The diagnosis was made by CT in 2 cases, by MRI in 7 cases, and by CT and MRI in three cases. In 7 cases the location was retro-cerebellar (figure 1), in 2 cases, supra-cerebellar (figure 2), in 2 cases (figure 3) and one case, intracerebellar (figure 4). On CT it appeared as uncalcified, low density mass with regular wall that did not enhanced. The fourth ventricle and vermis were visualized by axial CT or MRI in all cases except in one case, where the CT was misinterpreted as a communicating hydro-cephalus. A CT 3 months after of insertion of a ventriculoperitoneal shunt showed an isolated cyst at the incisura. MRI showed a

huge cyst in the posterior fossa. A supracerebellar arachnoid cyst compressing the fourth ventricle was diagnosed. In one case, metriza-mide CT ventriculography was performed in order to identify any communication between the cyst and fourth ventricle. The cysts contents on MRI had the same signal characteristics as cerebrospinal fluid (CSF) on both Tl and T2 images. No surrounding edema was seen.

Management and outcome

Cyst wall excision with fenestration into the 4^{th} ventricle subarachnoid cisterns was done in 9 cases and shunting procedures in 3 cases.

In 9 cases, histologically the wall was similar to the arachnoid structure. A VP shunt was inserted in one case and 3 months later a cystoperitoneal (SP) shunt was done. Y connected VP shunt and CP shunt was inserted at the same time in one case. Neurological deterioration occurred in case 11 before surgery. An external ventricular drainage (EVD) was inserted urgently, then the cyst was excised. However, the patient died on the fourth day. A V-P shunt was inserted at 12 months of age in case 12 as the first treatment. The child showed normal milestone till the 3^{rd} years. MRI of this case then revealed severe compression of the cerebellum and brain stem by the cyst.

The follow up periods ranged from 6 months to 2 years. All cases are free of symptoms and there is no recurrence (table 1).

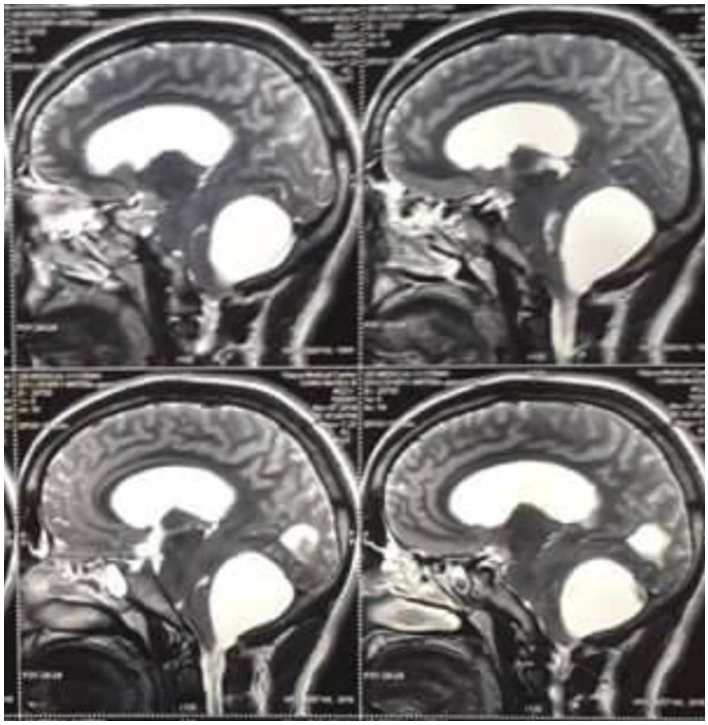

Figure (1): T_2 – weight image MRI showing retro cerebellar arachnoid cyst

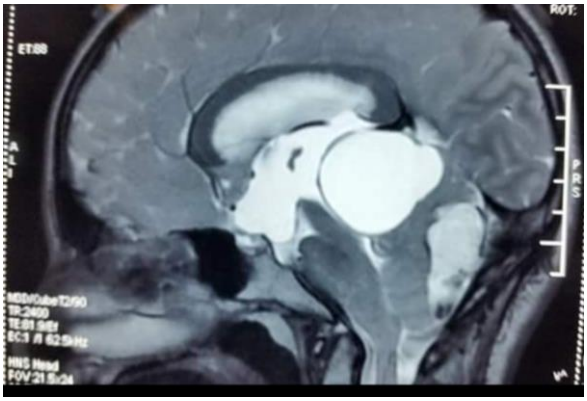

Figure (2): T_2 – weight image MRI showing Supra cerebellar arachnoid cyst

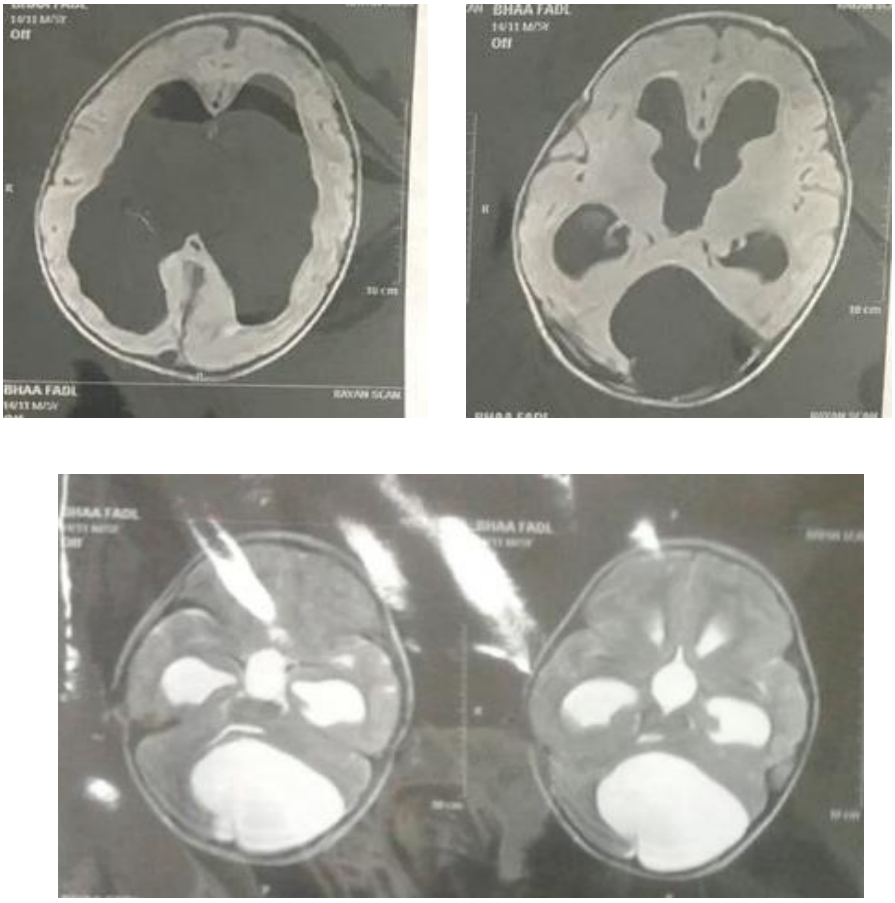

Figure (3): MRI Show Hydrocephalus with retro cerebellar cyst

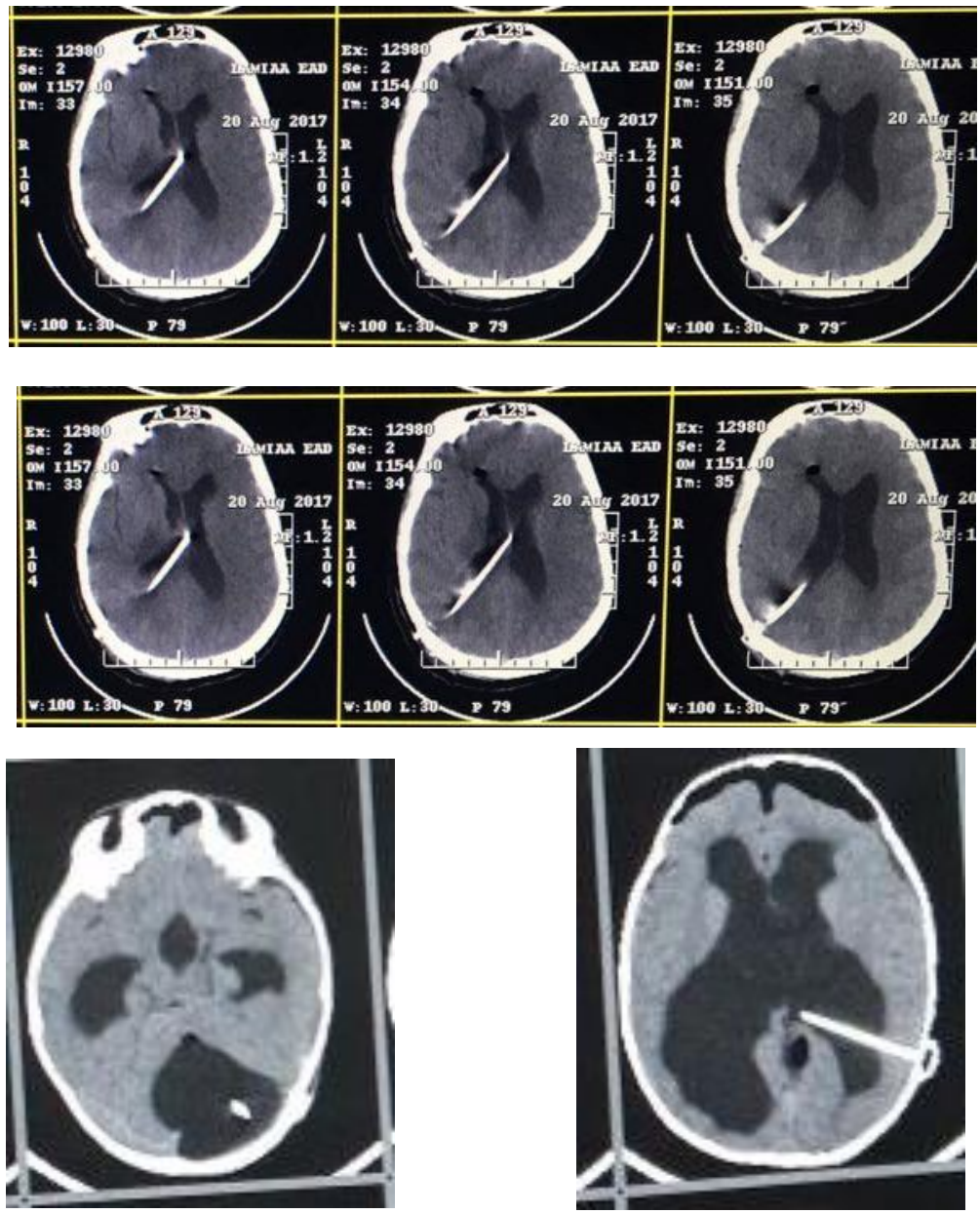

Figure (5): MRI show: Y connected VP + CP Shunt

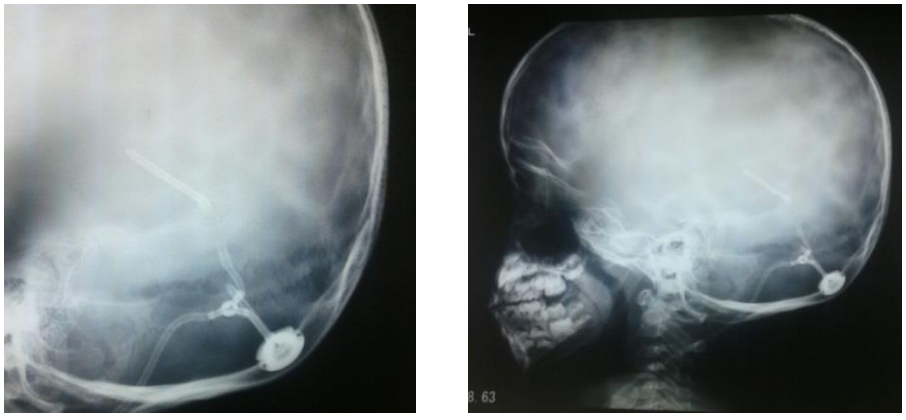

Figure (6): Plain X-Ray show: Y connected VP + CP Shunt

The 2nd most common location of arachnoid cysts is the posterior fossa [2-4]. The incidence of posterior fossa cysts among intracranial arachnoid cysts is 20 and 30% in pediatric series[3,5]

Clinical presentation

Symptoms are usually caused by compression of the surrounding nervous structures which give rise to symptoms and/or obstructive hydrocephalus[6,7]. The follow up data of case 12, we found that the retrocerebellar arachnoid cyst gradually compressed the cerebellum and the brain stem within 2 years of follow up which reveal the gradual increase in the size of the cyst in spite of a functioning V-P shunt. Three theories for explaining the filling of the cysts by CSF: The ball valve mechanism [8], active secretion of CSF by cyst wall [9] and filling by osmotic gradient [10]. The anatomical communication may also be intermittent [2].

Galassi et al., found that a minor head injury can provoke the onset of symptoms.. [12] Suggested that the clinical picture is of long duration with an episodic course in adults. Headache or vertigo can have a long history with a chronic increased in the intracranial pressure, gait ataxia or cranial nerve palsy was usually presented in short duration in our cases. If the cyst is associated with other CNS developmental abnormalities or with trauma or an inflammatory process[2,11}, inflammatory or traumatic aetiology in the history of our cases can't be detected, nor any association of any other severe CNS developmental abnormalities.

The hydrocephalus is frequently associated with infratentorial arachnoid cysts, some authors have described the pathogenesis either due to defective CSF absorption [14] or due to the developmental block of the cisterns rostral to the cyst[15,16] In our study in pediatric cases, macrocephaly was the alerting signs but in the adult cases the presenting symptoms were related to the posterior fossa. In the 5 adult cases there was mild hydrocephalus, a decrease in ventricular size after simple partial cyst excision. It seems that in adult cases mechanical obstruction of the CSF circulation pathways by the cyst may be the main cause of hydrocephalus. All pediatric cases presented with a rapid progression of hydrocephalus were treated with VP and CP shunting except the one who died following cyst excision

Classification

In recent reports, importance is attributed to the formation of the floor of the fourth ventricle and vermis[7,11]. Barko-vich et al., [7] define posterior fossa arachnoid cyst as all discrete posterior fossa. They categorize Dandy-Walker malformation and mega-cisterna magna as a separate entity of developmental anomalies of the posterior fossa[7]. A true arachnoid cyst is believed to has congenital origin, but is not associated with other developmental anomalies of the cerebrum and cerebellum.[11].

Differential diagnosis

The differential diagnosis includes cerebellar cystic astrocytoma, cystic hae-mangio-blastoma, hydatid cysts, and abscesses, epidermoid or dermoid cyst. A few literature in which an epithelial, glioependymal or choroidal cyst was diagnosed histolo-gically[11,19,20]. The characteristic appearance of an uncalcified, low density, extra-axial mass with regular borders that do not enhance with administration of contrast medium make the diagnosis from simple in most cases. In contrast to epidermoid tumors which are hyperintense on MRI images, encase and engulf arteries and cranial nerves, the arachnoid cysts displace adjacent structures[21].

The difference between arachnoid cysts and Dandy-Walker malformation, vermian cerebellar hypoplasia and mega cisterna magna is a problem[22]. In this study, the main point for the diffentiation was visualization of the 4th ventricle and vermis on CT or MRI. In all cases the posterior fossa was normal-sized, the torcula was normally positioned and the 4th ventricle and vermis were seen on CT or MRI except in one case which was misdiagnosed as a communicating hydrocephalus, although the preoperative CT were suggestive of a posterior fossa cyst. When MRI was available the non- communicating cyst and normally positioned torcula were easily

diagnosed. The 4th ventricle and brain stem were compressed by the cyst, but the vermis was intact.

The same morphology with posterior fossa arachnoid cysts[2,18,23]. In none of the six cases of posterior fossa arachnoid cyst reported by Bar-kovich et al.,[7], was there an enlarged posterior fossa or an elevated torcula. A normal-sized posterior fossa with a normal position of torcula are the key point of this D.D even in infants. Our four pediatric cases accorded with this observation.

The distinction between a communicating and non-communicating cyst is not necessary. In cases of mega-cisterna magna, if the posterior fossa size is normal, with no elevation of the tentorium and the torcula, filling of the cyst by contrast makes a clear differentiation between an arachnoid cyst and a mega cisterna magna[11,26]. There were no cases in this study that needed to be differentiated from a mega cisterna magna. In one case in which we performed CT ventriculography, we decided to insert a Y connected ventriclocysto-peritoneal shunt because of the non-communicating nature of the cyst.

Surgical treatment

There is controversy on the surgical treatment of arachnoid cysts either by shunting or by cystectomy[3]. In most of cases we performed cyst excision in a single operation with no recurrence. In 3 cases that shunting was done. Our results were satisfactory in all cases

Conclusion

If there are no developmental anomalies and if the clinical examination were only of a space occupying lesion. the surgery, will lead to cyst decreases in size, the cerebellum, re-expands, and the symptoms were relieved. But if there is a congenital origin, or developmental the management is debated. Elevation of the torcula and its sinuses, an enlarged posterior fossa are features that suggest poor surgical outcome than for a true arachnoid cyst.

The main cause of hydrocephalus in adult patients with posterior fossa arachnoid cysts is obstruction at the level of the 4th ventricle or aqueduct of Sylvius in this cases the treatment with the restoration of CSF circulation pathways by cyst excision without the need for shunting.

Successful results can be obtained by shunting alone in pediatric patients with hydrocephalus.

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