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# Symptomatic Surgically Treated Non-neoplastic Cysts of the Central Nervous System: A Clinicopathological Study From Pakistan

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## ABSTRACT

**Objective:** To report clinicopathologic features of symptomatic surgically removed non-neoplastic cysts of the central nervous system (CNS).

**Study Design:** Case series.

**Place and Duration of Study:** The Aga Khan University Hospital, Karachi, from 2003 to 2012.

**Methodology:** All non-neoplastic CNS cysts reported during the study period were retrieved and reviewed. Age, gender, location, histologic type and clinical features were noted.

**Results:** A total of 124 cysts were diagnosed in the study period. These included 44 epidermoid cysts (mean age  $30.5 \pm 13.8$  years), 35 colloid cysts (mean age  $31 \pm 13.2$  years), 32 arachnoid cysts (mean age  $24.8 \pm 20.2$  years), 6 dermoid cysts, 3 enterogenous cysts and Rathke's cleft cysts each and 1 ependymal cyst. All cyst types mainly presented in young adults in both genders with signs and symptoms of a mass lesion.

**Conclusion:** Non-neoplastic cyst mainly presented like a CNS mass lesion in young adults. Epidermoids were the most common type of these cysts in the present series followed by the colloid and the arachnoid cysts.

**Key Words:** Non-neoplastic cysts. Central nervous system. Epidermoid cyst. Dermoid cyst. Arachnoid cyst. Colloid cyst. Rathke's cleft cyst. Enterogenous cyst. Ependymal cyst.

## INTRODUCTION

Non-neoplastic cysts of the central nervous system (CNS) are mostly benign and congenital in origin. Often, these cysts are discovered incidentally during routine radiological examination or at autopsy. However, in many cases these cysts can present clinically as space occupying lesions of the CNS.

CNS cysts can arise from defects entirely within the nervous system and may be static (cysts arising in infarcts etc.) or progressive (arachnoid cysts, ependymal cysts etc.). They can also arise as a result of non-nervous tissue intruding into the neuraxis (epidermoid, dermoid, Rathke's cleft and colloid cysts etc). This latter group is congenital and the cysts expand and ultimately become symptomatic although some may become symptomatic in adult life.<sup>1</sup>

The primary non-neoplastic CNS cysts can arise from the cranial and spinal meninges, the cerebral hemispheres and cerebellum, the ventricles of the brain, sellar region, pineal gland, spinal canal etc. The main non-neoplastic CNS cysts include epidermoid cyst,

dermoid cyst, colloid cyst of the third ventricle, enterogenous cyst, choroid plexus cyst, perineural cyst and ependymal cyst.<sup>2</sup>

Epidermoid cysts slowly but invariably progress and ultimately require surgical treatment.<sup>3</sup> They probably arise from the inclusion of embryonic ectodermal elements relatively late in embryogenesis during closure of primitive neural groove. Colloid cysts are usually considered to be neuroectodermal in origin<sup>1</sup>, but immunohistochemical and ultrastructural studies support an endodermal origin for these cysts.<sup>4,5</sup> Congenital arachnoid cysts probably result from focal splitting of the developing arachnoid membrane during embryogenesis. Most arachnoid cysts are found incidentally on intracranial imaging in all age groups and can be managed conservatively. However, a small number of cases which are symptomatic should be treated surgically regardless of age and location.<sup>6-9</sup> Spinal arachnoid cysts can be traumatic or inflammatory in origin but if accompanying histologic changes are not present, they should be considered as congenital malformations.

To date, no local data or figures regarding non-neoplastic CNS cysts is available. Autopsies are not routinely performed in this country due to religious and cultural reasons. However, as the largest referral center for Histopathology in Pakistan, the authors aimed to report here the non-neoplastic CNS cysts which presented as symptomatic space-occupying lesions

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(SOL) and were surgically removed and sent for histopathological examination

## METHODOLOGY

The authors retrieved hematoxylin and eosin (H and E) stained slides of 124 cases of CNS cysts reported between 2003 and 2012 in the Section of Histopathology, the Aga Khan University Hospital. The cases were archived by electronic search of each type of lesion. Cystic CNS neoplasms and inflammatory lesions were excluded. The variables noted included age, gender, exact location, histologic type of cyst, and clinical signs and symptoms. All collected data was analyzed on SPSS software version 20. Mean and standard deviation was calculated for continuous variables. Frequency and percentage was calculated for categorical variables. Since this was a retrospective observational study, ethical approval was not sought.

## RESULTS

A total of 124 cysts were diagnosed in the study period (Table I). Epidermoid cysts were the commonest, closely followed by colloid cysts of the third ventricle, and arachnoid cysts. Males were affected much more commonly than females (Table II). For epidermoid cysts, overall age range in both males and females was 8 to 69 years with mean age of  $31.1 \pm 13.8$  years. In males, age

range was 8 to 69 years with mean age of  $32.1 \pm 14.5$  years. In females, age range was 12 to 56 years with mean age of  $29.7 \pm 13.1$  years. For colloid cysts, overall age range in both males and females was 11 to 63 years with mean age of  $30 \pm 13.2$  years. In males, age range was 11 to 63 years with mean age  $31.5 \pm 13.8$  years. In females, age range was 15 to 62 years with mean age of  $30.4 \pm 13.8$  years. For arachnoid cysts, overall age range in both males and females was 1 to 57 years with mean age of  $25.3 \pm 20.2$  years. In males, age range was 1 to 55 years with mean age of  $25 \pm 19$  years. In females, age range was 2 to 57 years with mean age of  $24.8 \pm 20$  years. For dermoid cysts, overall age range was 12 to 34 years with mean age of 24 years. The three patients with enterogenous cysts were 12, 25 and 30 years of age. The three patients with Rathke's cleft cyst were 27, 30 and 69 years of age, while the only patient with ependymal cyst was 18 years of age.

Cerebellopontine angle (CPA) was the most common site for epidermoid cysts (Table III). Of the 10 cases in the cerebral hemispheres, 5 were in the temporal lobe, 4 in the frontal lobe and 1 in the parietal lobe. All the 6 cases in the spine were in the thoracolumbar region. Of the 6 dermoid cysts, 3 were in the posterior fossa, 2 in the spine, and 1 in the anterior cranial fossa. All colloid cysts were located in the third ventricle. Similarly, all Rathke's cleft cysts were located in the pituitary fossa. All three enterogenous cysts were located in the cervical (1) and upper thoracic (2) regions of the spinal canal. Out of the 32 arachnoid cysts, 8 (25%) were located in the posterior fossa, located either between the cerebellar hemispheres or in the cerebellopontine angle, while another 8 (25%) were located supratentorially

**Table I:** Breakup of histologic types of Non-neoplastic CNS cysts (n=124).

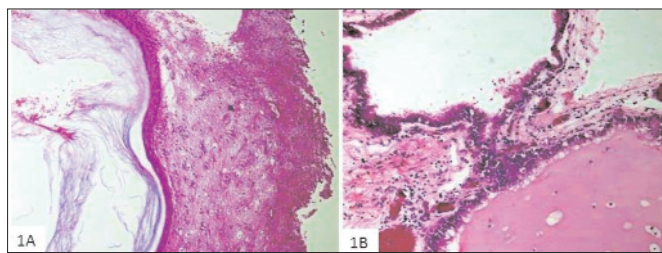
Histologic Type	Number	Percentage (%)
Epidermoid cyst	44	35.5%
Colloid cyst	35	28.2%
Arachnoid cyst	32	25.8%
Dermoid cyst	6	4.8%
Enterogenous cyst	3	2.4%
Rathke's cleft cyst	3	2.4%
Ependymal cyst	1	0.8%

**Table II:** Distribution of cases according to gender (n=124).

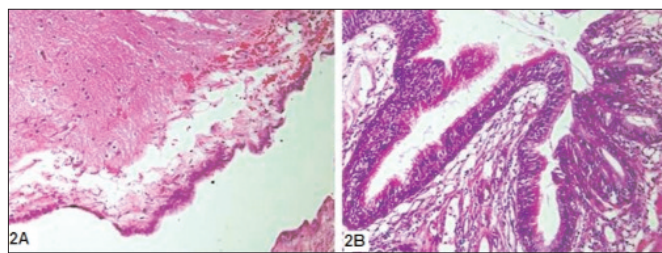
Histologic Type	Number	Males	Females
Epidermoid cyst	44	26 (59.1%)	18 (40.9%)
Colloid cyst	35	21 (60%)	14 (40%)
Arachnoid cyst	32	24 (75%)	8 (25%)
Dermoid cyst	6	3 (50%)	3 (50%)
Enterogenous cyst	3	1 (33.3%)	2 (66.6%)
Rathke's cleft cyst	3	2 (66.6%)	1 (33.3%)
Ependymal cyst	1	1 (100%)	-

**Table III:** Distribution of epidermoid cysts according to site (n=44).

Site	Number	Percentage (%)
Cerebellopontine angle	15	34.1%
Cerebral hemisphere	10	22.7%
Spine	6	13.6%
Sellar region	6	13.6%
Lateral ventricle	3	6.8%
Skull bone	2	4.5%
Site not specified	2	4.5%



**Figure 1:** (A) Epidermoid cyst of CPA. The cyst has keratinized stratified squamous lining. Glial tissue is present beneath lining. (B) Colloid cyst of third ventricle. The cyst has ciliated columnar lining with lumen containing pink colloid like material [H&E, 400x].



**Figure 2:** (A) Arachnoid cyst of CPA of a 13-year old boy. The cyst lining is meningotheial resting on thin collagenous membrane overlying brain tissue. (B) Enterogenous cyst of spinal cord. The cyst lining is mucinous pseudostratified ciliated columnar epithelium [H&E, 200x].

in the frontotemporoparietal region. However, the commonest site was the spine. Out of 32, 11 (34.4%) cases were located in the spine. Site was not stated in 4 (12.5%) cases while 1 (3.1%) case was located in the suprasellar region. Of the 11 spinal arachnoid cysts, 7 (63.6%) were in the thoracic, 3 (27.3%) in the lumbar, and 1 (9.1%) in the cervical region.

Histologically, both epidermoid and dermoid cysts in our series were lined by stratified squamous epithelium with shed anucleate keratin squames in the lumen (Figure 1A). In dermoid cysts, adnexal appendages such as sweat glands and sebaceous glands were seen. Foreign body giant cell reaction secondary to cyst rupture was seen in a number of cases of both epidermoid and dermoid cysts. Colloid cysts were lined by a single layer of ciliated epithelium often with mucin production (highlighted on PAS stain). Beneath the epithelium, thin fibrous connective tissue was seen (Figure 1B). Arachnoid cysts had thin, delicate fibrovascular membranes lined by nests of meningotheial cells (Figure 2A). Enterogenous cysts were lined by pseudostratified, ciliated and mucin producing cells (Figure 2B). Rathke's cleft cysts histologically showed a lining composed of a single layer of columnar, mucin producing cells (similar to enterogenous cells). The single endymal cyst in our series had simple cuboidal to columnar endymal lining with a zone of fibrillar glial tissue intervening between the epithelium and the brain parenchyma.

## DISCUSSION

Epidermoid cysts (35.5%) were the commonest type in this series. They were most common in young adults especially males (Table II) and were located in the cerebellopontine angle (34.1%) followed by the cerebral hemispheres. Epidermoid cysts were also the commonest CNS cysts in two separate studies from India.<sup>10,11</sup> Cerebellopontine angle is the most common site for epidermoid cysts.<sup>12,13</sup> In a study by Lopes *et al.*<sup>3</sup> which evaluated at 44 epidermoid cysts, the number of males and females was equal (22 each) and mean age was 39.9 years. Epidermoid cysts of cerebellopontine angle are commonly symptomatic.<sup>14</sup> Epidermoid cysts in this series were also seen in the spine, sellar region, lateral ventricles etc (Table III). In the spine, all were located in the thoracolumbar region, which is the commonest spinal site for these cysts.<sup>1</sup> There are a number of studies in literature documenting the occurrence of epidermoid cysts in many of these locations, as well as in the fourth ventricle, brain stem etc.<sup>15-18</sup> Colloid cysts of the third ventricle were the second commonest type in our series comprising 28.2% (Table I). Like arachnoid cysts, they were also most common in young adults and males were affected more commonly than females (Table II). Colloid cysts were also the second commonest cysts in the series by Sundaram *et al.*<sup>11</sup> Other studies have also shown that

these cysts are more common in males, and most commonly occur in the fourth or fifth decade.<sup>19</sup> In this study, mean age was around  $31 \pm 13.2$  years. Colloid cysts can also be infra or suprasellar.<sup>20</sup>

Arachnoid cysts were the third commonest histologic type in our series comprising 25.8% (Table I). They were also most commonly seen in young adults albeit at a younger age (mean age around  $25 \pm 20.2$  years) compared to epidermoid and colloid cysts, and were much more common in males (Table II). In two separate studies by Al-Holon *et al.*, arachnoid cysts were much more common in males in both adults and children.<sup>6,7</sup> A study by Duz *et al.*<sup>8</sup> reported an age range of 2 months to 45 years and a mean age of 20 years. Arachnoid cysts were the second and third commonest cysts in the two studies from India.<sup>10,11</sup> In this study, 25% each were located in the posterior fossa (infratentorial) and middle fossa (supratentorial, fronto-temporo-parietal region). However, the commonest location was spine (34.4%). Of the spinal arachnoid cysts in this study, over 63% was in the thoracic spine. Various studies have shown that the commonest locations for intracranial arachnoid cysts are the middle fossa followed by posterior fossa (retrocerebellar).<sup>6,7</sup> In the spine, the low or mid-thoracic region is the commonest location.

The 6 dermoid cysts in our study were equally distributed among males and females (Table II) and mean age was 24 years. Posterior fossa was the commonest location followed by the spine. In the two studies from India, dermoid cysts were more common than in our study being the second and third most common type.<sup>10,11</sup> Cranial dermoid cysts are most commonly located in the posterior fossa, and in the spine almost always occur in the lumbosacral region.<sup>1</sup> Dermoid cysts usually present at a younger age than epidermoid cysts.<sup>15</sup> In this study, mean age was younger compared to that for epidermoid cysts (24 years compared to 31 years). Dermoid cysts like epidermoid cysts are believed to arise from ectodermal elements which become entrapped within the developing neural tube before it closes.<sup>13</sup>

All three enterogenous cysts in this study were located in the spinal cord and occurred in young adults, two in females and one in male. Enterogenous (or neurenteric) cysts occur mainly in the spinal canal, are mostly diagnosed in young adults, and are more common in males.<sup>21</sup> Enterogenous cysts may not produce any characteristic clinical features, but can present with slowly progressive myelo-radiculopathy.<sup>21</sup> They probably result from an abnormal embryological connection between the primitive foregut and the developing neural tube.<sup>21</sup>

Only a single case of brain enteric cyst has been published from Pakistan.<sup>22</sup>

There were 3 Rathke's cleft cysts in this study accounting for 2.9% of all cysts. Rathke's cleft cysts

were similarly uncommon in other studies.<sup>10,11</sup> Symptomatic Rathke's cleft cysts are very rare, can present from the second to the eighth decade, are more common in females, and represent remnants of Rathke's cleft.<sup>23</sup>

The epidermoid and dermoid cysts in this series presented with symptoms and signs of a slowly enlarging mass. These included headache, drowsiness, blurring of vision, slurring of speech, gradual impairment of memory, progressive cranial nerve palsy, left sided weakness, difficulty in walking, trigeminal neuralgia etc. These clinical features are well documented for these cysts.<sup>24</sup> Colloid cysts in this series presented with headaches, hydrocephalus and sudden episodes of loss of consciousness. Chronic headaches are a common presenting symptom of colloid cysts and sudden 'drop attacks' due to acute impaction in the foramen of Monro sometimes occur and may even lead to coma or death.<sup>25</sup> Cranial arachnoid cysts in this series presented with clinical features of a mass lesion producing symptoms and signs such as headache, seizures, vomiting, vertigo, hydrocephalus etc. Spinal arachnoid cysts presented with backache, girdle paresthesias, paraparesis, lumbar swelling etc. Arachnoid cysts of posterior fossa also presented with eighth nerve dysfunction, a feature well documented for arachnoid cysts in this location.<sup>26</sup> Enterogenous cysts in our series presented with paraparesis. The three Rathke's cleft cysts in this series presented with headache and visual disturbances. These presenting features are well documented in literature.<sup>23</sup>

All the cysts included in this series were symptomatic, treated surgically, and sent for histopathological examination. The results show that the clinical features, epidemiological data (age, gender, site etc), and incidence of CNS cysts are similar to those reported in both regional and western studies.

## CONCLUSION

Non-neoplastic-cyst mainly presented like a CNS mass lesion in young adults. Epidermoids were the most type of these cysts in the present series followed by colloid and the arachnoid cysts.

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