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Research Article

CYSTS OF THE CENTRAL NERVOUS SYSTEM: A REVIEW OF 35 CASES WITH OUTCOMES

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ABSTRACT

Objective: Non-neoplastic, non-inflammatory cysts of the central nervous system (CNS) may cause symptoms as a result of changes in pressure, rupture or secondary inflammation. The cyst-related details, such as its age, site, cyst wall, and its contents, also provide an insight into their embryology and histogenesis. This paper reviews the clinic-pathological features of cysts of CNS.

Methods: In this study, a prospective analysis of the cysts of CNS diagnosed between November 2019 and October 2023 who presented to our outpatient department and managed at Command Hospital Lucknow and Base Hospital Delhi Cantt were reviewed. Written informed consent was taken from all patients. Cystic degeneration in tumors or inflammatory lesions was excluded from the study.

Results: There were 35 cysts in the study period of 04 years. Majority of intracranial cysts presented with signs of raised intracranial pressure. Patients with epidermoid cysts had additional cerebellar signs, visual disturbances and deafness. Cord compression is the main presentation of all spinal cysts.

Conclusion: Cysts of CNS are usually maldevelopment in nature. A few are acquired. The radiological identification of these cysts and pathological confirmation is necessary for prognostic purposes.

Keywords: Cysts, Clinical features, Histopathology.

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INTRODUCTION

Cysts of the central nervous system (CNS) are mostly benign and are an accidental finding on autopsy. They may show symptoms due to pressure changes, rupture or secondary inflammation. These cysts generally have a good prognosis although they show recurrence due to rupture or incomplete surgical removal. The cyst-related details, such as its age, site, cyst wall, and its contents, also provide an insight into their embryology and histogenesis. This paper reviews the clinic-pathological features of cysts of CNS.

METHODS

The cysts of CNS diagnosed between November 2019 and October 2023 who presented to our outpatient department and managed at Command Hospital Lucknow and Base Hospital Delhi Cantt were reviewed. Written informed consent was taken from all patients. Cystic degeneration in tumors or inflammatory lesions was excluded from the study. The age, site, magnetic resonance imaging (MRI) and computed tomography (CT) scan findings, cyst wall lining, and nature of cyst contents were noted in all cases. These included epidermoid and dermoid cysts of brain and spinal cord; colloid cysts of 3rd ventricle, arachnoid cysts of brain and spinal cord and Rathke's cleft cysts.

RESULTS

There were 35 cysts in the study period of 04 years. Tables 1 and 2 give details of age, sex, and site majority of intracranial cysts presented with signs of raised intracranial pressure. Patients with epidermoid cysts had additional cerebellar signs, visual disturbances and deafness. Seizures and increased head circumference were common in arachnoid cysts. Cord compression is the main presentation of all spinal cysts.

Epidermoid and dermoid cysts

While epidermoid cysts are more common in brain, dermoid cysts are more common in the spinal cord. The most common location of epidermoid cysts in the brain was cerebellopontine angle followed by suprasellar area and others. In the spinal cord, they were intradural in four and intramedullary in one. The spinal forms presented with cord compression and had associated anomalies in the form of spina bifida in four and kyphoscoliosis in another. Histology showed a cyst wall lined by stratified squamous epithelium and the cyst contents were keratinous flakes. One had intramedullary and four had intradural cysts. Cysts were lined by stratified squamous epithelium and hair follicles in all cases.

Colloid cysts

All colloid cysts were located in anterior 3rd ventricular region and all of them presented with raised intracranial pressure. The cyst wall was lined by cuboidal to columnar epithelium with or without cilia, supported by delicate collagenous stroma. Contents of cysts were mucoid in consistency.

Arachnoid cysts

These cysts, both in brain and spinal cord, presented with pressure symptoms. Histopathological findings of the cyst wall showed wall lined by flattened or cuboidal cells supported by collagen. (Figs 1 and 2) Contents were clear fluid like CSF.

Rathke's cleft cysts

Cysts were suprasellar in location. Two patients presented with headache and vision symptoms. The contents of the cyst were greenish in color and oil like in consistency.

Table 1: Clinical data

Type of Cyst	M: F	Age range	Mean age
Epidermoid cysts			
Brain	14:13	4–62 years	29.2 years
Spinal cord	3:5	5–65 years	26.7 years
Dermoid cysts			
Brain	1	32 years	32 years
Spinal cord	1:1	4.5-53 years	38.1 years
Arachnoid cysts			
Brain	5:3	9–65 years	24.3 years
Spinal cord	2:1	21–68 years	42 years
Colloid cysts	2:1	15–45 years	27.5 years
Rathke's cleft cysts	1:2	13–55 years	33.5 years

Table 2: Location of cysts of CNS

Type of Cyst	n	Location	n
Epidermoid			
Brain	8	Cerebellopontine angle	7
		3rd ventricle	1
Spinal cord	1	Dorsal	1
Dermoid cyst			
Brain	1	Vermian	1
Spinal cord	4	Dorsal and dorsolumbar	4
Colloid cysts	3	3rd ventricle	3
Arachnoid cysts			
Brain	12	Cerebellopontine angle	3
		Parenchymal	7
		Suprasellar	2
Spinal cord	3	Dorsal	1
		Lumbar	2
Rathke's cleft cysts	3	Suprasellar	3

CNS: Central nervous system

No malignant transformation was seen in any of the cyst. All of them were benign.

DISCUSSION

Cysts of CNS are usually maldevelopment in nature. A few are acquired. The radiological identification of these cysts and pathological confirmation is necessary for prognostic purposes. Epidermoid cysts account for 0.2–1.8% of all intracranial tumors and <1% of all intraspinal tumors. Cranial sites outweigh spinal sites by 14:1 [1-3]. The present series shows an approximate 8:1 ratio. Intracranial epidermoids usually occupy the cerebellopontine angle, parasellar/suprasellar region, cerebral and cerebellar hemispheres, ventricular region and pineal region [1-5]. Epidermoid cysts of the brainstem are extremely rare [2,6]. Intraspinal epidermoids are mostly intradural and extramedullary in location. Roux *et al*. [3] reviewed the published 47 cases of intramedullary epidermoid cysts. In our series, there was one intramedullary cyst in dorsolumbar location. Dermoid and epidermoid cysts due to the slowly enlarging lesion may present with mass effect [7]. Dermoids typically become symptomatic during the first 2 decades of life, and epidermoids, due to their slower production of compact keratin, tend to present between the ages of 20 and 50 years [7]. Cutaneous tissues misplaced during embryogenesis represent epidermoid and dermoid cysts and these are found along lines of otogenic neurocutaneous differentiation. These ectodermal inclusions occur between 3rd and 5th weeks of embryonic life and may result in heterotopia of these elements. The separation of neuroectoderm and its cutaneous counterpart which occurs dorsally along the midline may explain the median location of these cysts. Laterally situated lesions may result from the inclusion of ectoderm at a later stage of embryogenesis, especially during the formation of secondary otic and optic cerebral vesicles [2,3,7]. Colloid cysts of the third ventricle constitute 0.25 −0.5% of all intracranial tumors [8]. It is a well-established clinicopathologic entity as defined by its location, contents and histology. The most

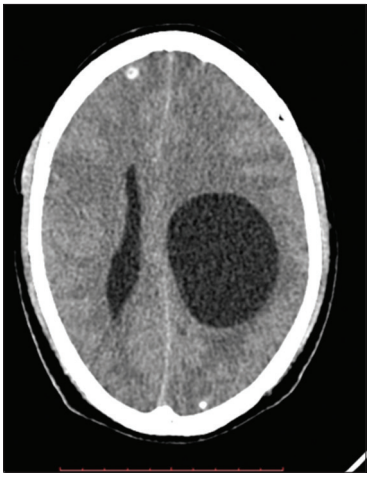

Fig. 1: Pre-operative non-contrast computed tomography head

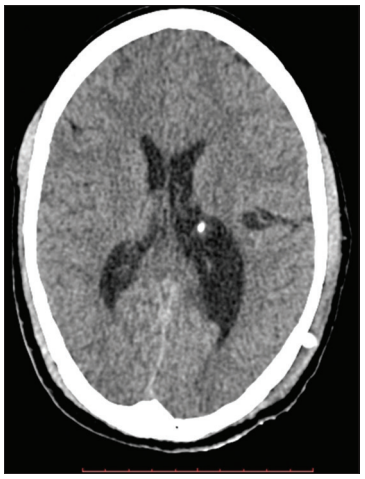

Fig. 2: Post-operative non-contrast computed tomography head showing a decrease in size of cyst

presentation in colloid cyst is acute hydrocephalus. The colloid cyst obstructs one or both foramina of Monro. With the advent of CT and MRI, it is more readily diagnosed and the characteristic radiologic appearance is due to its high cholesterol content [9].

Rathke's cleft cysts are small and asymptomatic and are found in 2–26% of routine autopsy series [10,11]. With the modern neuroimaging technology, they are being diagnosed much more frequently. Symptomatic Rathke's cleft cysts were reviewed by Voelker *et al.* [10]. These cysts are thought to originate from remnant of Rathke's pouch [11]. It is the persistence and enlargement of the cleft from the posterior wall of the pouch that produces Rathke's cleft cyst [10,11]. Immunohistochemical studies confirm the foregut derivation of Rathke's cleft cysts similar to colloid cysts and enteric cysts.

Benign developmental cysts that occur in the cerebrospinal axis in relation to the arachnoid membrane are the Arachnoid cysts. The lining epithelium, as seen in our cases, shows collagen and meningothelial cells. The cyst's contents were clear and liquid resembling normal CSF. All our cysts show the same. Despite several studies, the mechanism of formation of these cysts is not completely understood [12].

The cysts of the CNS are being identified more in a number due to modern neuroimaging. An understanding of the pathogenesis and embryology explains the location, presentation and histology in such cases. They are usually maldevelopment and malignant transformation of the lining epithelium is extremely rare.

CONCLUSION

Cysts of CNS are usually maldevelopment in nature. These cysts share many radiological characteristics with a variety of intracranial benign cysts. A few are acquired. The radiological identification of these cysts and pathological confirmation is necessary for prognostic purposes.

AUTHORS' CONTRIBUTION

All the cases presented in the current study were operated by AK. DS, NP and SC have contributed to final formatting and data analysis.

CONFLICTS OF INTEREST

There are no conflicts of interest.

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