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Original Research Article

Intracranial Cysts: A Morphological Spectrum

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Abstract

Introduction: Intracranial cysts are often seen in our daily routine practice. Most of the patients present with symptoms of raised intracranial pressure manifested as headache and vomitings. Most of these are benign in origin with defects in maldevelopment. Histopathological examination plays a major role in the diagnosis of these cases. We have presented a spectrum of lesions in our study period with emphasis on their origin, morphological features and differential diagnosis.

Materials and methods: A total of 11 cystic lesions were included in our study. Cystic degeneration of tumors and infective cysts were excluded from our study. Processing was done. Histopatholy slides were made and examined. Immunohistochemistry (IHC) was done wherever required.

Results: The lesions included epidermoid, dermoid, neuroenteric, colloid, arachnoid and rathke cleft cysts. Cysts of the neurospinal axis i.e Rathke cleft cyst, colloid cyst and neurenteric cyst had almost identical histological features and the site of the lesion played an important role in the diagnosis of these lesions.

Conclusion: Incranial cysts should be diagnosed in correlation with clinical features and imaging findings. These benign lesions can thus be managed appropriately sometimes in aconservative manner also. Histopathological diagnosis thus plays a major role in these intracranial cystic lesions.

Keywords: Incranial cysts; Morphological Spectrum; Histopathological diagnosis; Immunohistochemistry (IHC).

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Introduction

Cysts of the central nervous system and space occupying lesions which mimic cystic lesions are very common in routine practice especially in the recent years with advances in radiodiagnosis. They are a real diagnostic challenge to both the neurologist and the radiologist. Histopathological examination is very essential in these cases. We present a spectrum of cystic lesions of the central nervous system, their origin and their differential diagnosis which are very helpful in the final precise diagnosis.

Materials and Methods

We have done a prospective study over a period of 1 year at Narayana Medical College. Cysts of the brain received during this period were reviewed. Tumours with cystic degeneration or inflammatory lesions were excluded from the study. The clinical details with special reference to age and site were noted. All the specimens were processed and histopathological examination was done. Morphological aspects especially nature of cyst contents and cyst wall lining were noted in all cases. Based on these findings the various types included epidermoid, dermoid cysts of both brain and spinal cord, colloid cysts, arachnoid cysts, neurenteric cysts and rathke cleft cysts.

Results

In our study, during a period of one year we have encountered a total of 11 cases. The distribution of cases and the clinico-morphological details are provided in Table 1 and 2. Most of the cysts located in the brain presented with features of raised incranial pressure like headache and one case of dermoid cyst and another case of arachnoid cyst presented with additional features of status epilepticus. Lesions of the spinal cord presented with features of cord compression. Hydrocephalic attacks were seen in both cases of colloid cysts. One case of Rathke cleft cyst presented with endocrine disturbances.

Table 1.

Type of cyst	Number
Epidermoid cysts	2
Brain	2
Spinal cord	
Dermoid	3
Brain	3
Spinal cord	
Neurenteric	2
Rathke cleft	1
Colloid cyst	2
Arachnoid cyst	1

Table 2: Cysts of brain – Clinicomorphological details

S.No	C/O	Sex/Age	Type	Location	Lining	Contents
1	Head injury with loss of consciousness	46/F	Epidermoid	Right temparo-parietal	Squamous epithelium	Keratinous debris
2	Loss of consciousness	49/M	Epidermoid	Temporal	Squamous epithelium	Keratinous debris
3	Status epilepticus	50/M	Dermoid	Fronto-parietal	Squamous epithelium with skin appendages	Milky white fluid
4	Headache	32/F	Dermoid	4rth ventricle	Squamous epithelium with skin appendages	Milky white fluid
5	Headache with occasional vertigo	45/F	Dermoid	Midline parasagittal	Squamous epithelium with skin appendages	Milky white fluid
6	Neck pain with headache and occasional vertigo	17/F	Neurenteric	Cervical spinal region	pseudo stratified ciliated columnar epithelium	Clear to mucoid
7	Pain in the neck region	34/M	Neurenteric	Cervical spinal region	pseudo stratified ciliated columnar epithelium	Clear to mucoid
8	daytime somnolence with endocrine disturbances	56/M	Rakthe cleft	Rathke pouch - hypophysis	Non ciliated, single layer of flat cells or stratified columnar cells – mucin producing cells	Machine oil
9	Headache	43/F	Colloid cyst	Anterior 3rd ventricle	Cuboidal epithelium	Viscous gelatinous material
10	Headache	48/F	Colloid cyst	Anterior 3rd ventricle	Cuboidal epithelium	Viscous gelatinous material
11	Status epilepticus	25/M	Arachnoid cyst	Posterior cranial fossa	Flattened to cuboidal	Clear CSF like fluid

Dermoid and Epidermoid Cysts

All these cysts were located in the brain with complaints of raised intracranial pressure but an additional feature of status epilepticus was seen in one case of dermoid cyst. Microscopic examination of epidermoid cysts revealed stratified squamous epithelium lining and filled with keratin flakes. In dermoid cysts, skin appendages were also seen in addition and they were filled with milky white fluid.

Neurenteric Cysts

Both the cases presented with features of cord compression. Histopathological examination revealed cyst lined by ciliated columnar and filled with clear to mucoid material.

Rathke Cleft Cyst

A single case was located in the hypophysis with morphology showing single layer of non ciliated, flattened epithelium and at few places stratified columnar cells and mucin producing cells. Contents of the cyst revealed material with machine oil consistency.

Colloid Cyst

Both the cases presented with features of raised intracranial pressure with microscopy showing lining cuboidal epithelium.

Arachnoid Cyst

One case with presenting complaints of status epilepticus located in the posterior cranial fossa. Cyst wall was lined by flattened to cuboidal epithelium and filled with clear CSF like fluid (Fig. 1A-F).

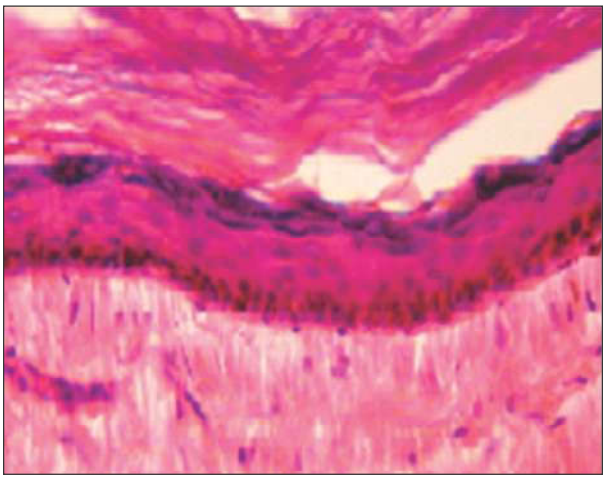

Fig. 1A: Dermoid cyst: wall lined by stratified squamous epithelium (H&E 40X)

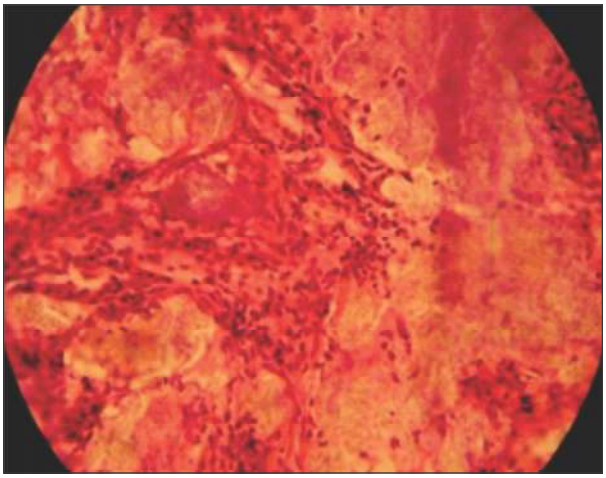

Fig. 1B: Epidermoid cyst: wall lined by stratified squamous epithelium with dermal appendages (H&E 40X)

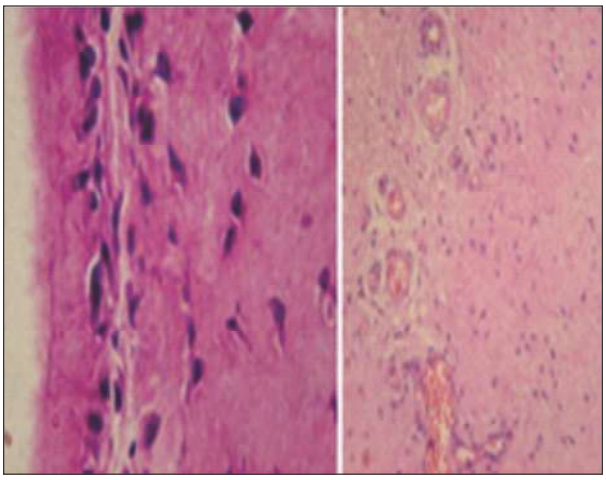

Fig. 1C: Neurenteric cyst: wall lined by pseudostratified epithelium with occasional mucin secreting cells (H&E 40X)

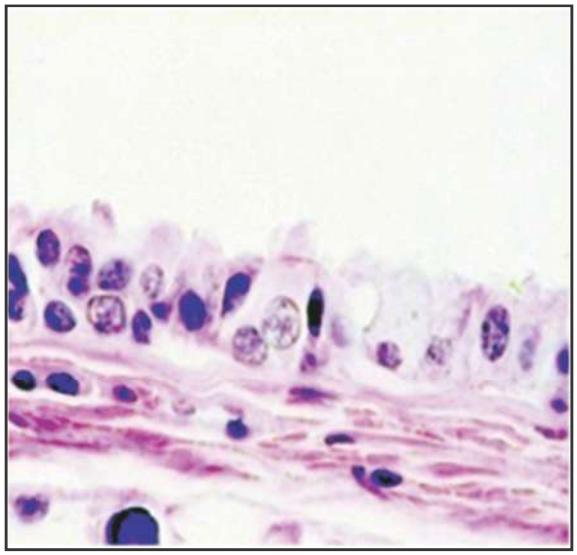

Fig. 1D: Colloid cyst: wall lined by columnar epithelium with mucin containing cells (H&E 40X)

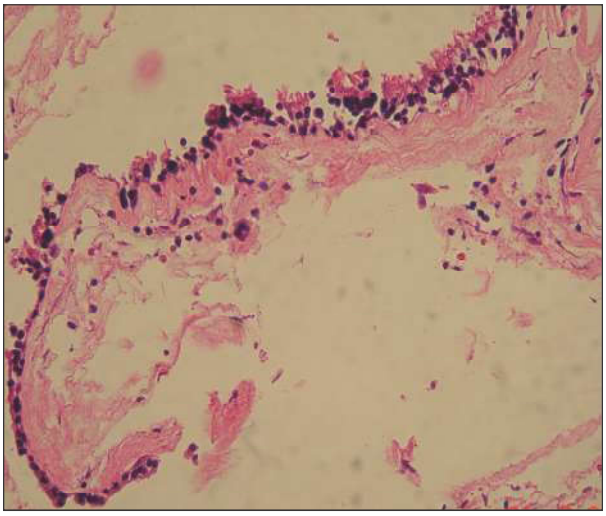

Fig. 1E: Rathke cleft cyst: wall lined by ciliated epithelial & mucous secreting cells (H&E 40X)

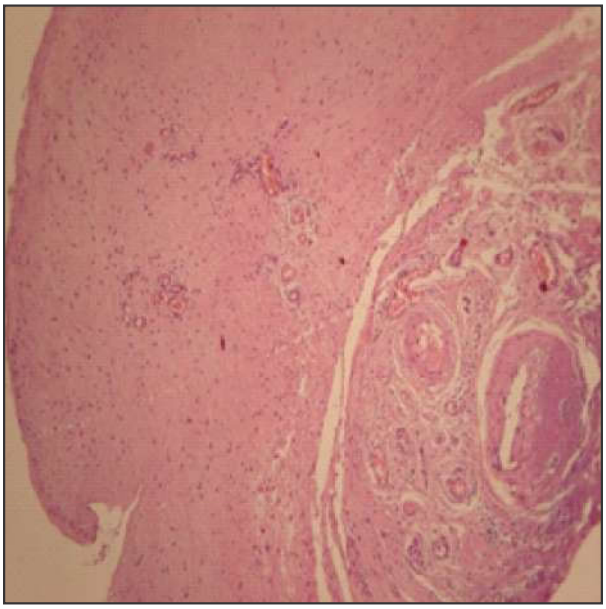

Fig. 1F: Arachnoid cyst: wall lined by flattened to cuboidal epithelium (H&E 40X)

Fig. 1A-F: Microscopy of CNS cysts

Discussion

The cysts of central nervous system are usually maldevelopmental in majority of the cases with a few of them being acquired. It is very essential to diagnose them radiologically and confirm them by histopathological examination. Differentiating them from cysts of various other etiology like infective or metastatic origin is also of paramount importance [1].

Epidermoid cysts account for 0.2 to 1.8% of all intracranial tumours and less than 1% that of all intraspinal tumours. Cranial sites outweigh spinal sites by 14:1. In our study all of them were located the brian. During embryogenesis, nests of cutaneous tissues are misplaced during 3rd to 5th week of embryonic life. Both epidermoid and dermoid cysts represent these tissues of ontogenic neurocutaneous differentiation [2,3]. Repeated lumbar punctures or incidental formation of a skin pocket by suturing leads to acquired epidermoid cysts in the lumbar region [4]. We have not encountered even a single case may be because of the expertise in doing the technique.

Neurenteric cysts are believed to result from embryonal dysgenesis. In the 3rd week of embryonal life, neurenteric canal gets closed and there is separation of notochord from the primitive gut. This process when disrupted may lead to the inclusion of endodermal tissue and cystic formation [5]. In the central nervous system (CNS) they have been reported to occur from the level of posterior fossa to the lumbosacral region [6]. Based on the histological features these cysts were classified into three groups by Wilkins and Odom [7]. Group A, the simplest type is lined by a layer of single cuboidal or columnar epithelial cells with or without cilia. Group B, the more complex type, has in addition elements of the gastrointestinal tract or tracheobronchial tree, for instance mucous glands and smooth muscle. Ependymal or glial tissue found along with Group B comprise the last group C.

The endodermal origin of these cysts is confirmed by immunohistochemical studies [8].

Rathke's cleft cysts are believed to originate from remnants of Rathke's pouch. The persistence and enlargement of the cleft from posterior wall of the pouch produces the cyst. Foregut derivation of Rathke's cleft cysts is confirmed by immunohistochemical studies [9-11].

Colloid Cyst

Colloid cysts of the third ventricle constitute 0.25 -0.5% of all intracranial tumours [12]. As defined by its location, contents and histology, it is a well established clinicopathologic entity. the commonest clinical presentation is acute hydrocephalus in view of its location leading to the obstruction of one or both foramina of Monro. Its high cholesterol content give a characteristic radiologic appearance make it readily diagnosed with the advent of CT and MRI [13]. Recent immunohistochemical studies showed a pattern identical to that of foregut derived epithelium and these findings indicate an

endodermal rather than neuroepithelial nature as was believed previously [14,15].

Arachnoid Cyst

Arachnoid cysts are developmental in origin and benign in nature. They occur in the cerebrospinal region in relation to the arachnoid membrane. The proposed theory regarding its formation is splitting of the arachnoid membrane at the margin of the cyst between the outer and inner layers of the arachnoid [16].

Differential Diagnosis

Rathke cleft cyst, colloid cyst and neurenteric cyst, all the three together constitute cysts along the neurospinal axis. All these are histologically identical except the location. Craniopharyngioma is most common lesion to be confused with these cysts because of the presence of squamous metaplasia in both. Presence of solid epithelial islands, absence of keratin, stellate reticulum and basal palisiding epithelium are the specific histological features for craniopharyngioma. Ependymal cyst can also be misdiagnosed histologically but IHC distinguishes this entity. GFAP and S100 are positive in Ependymal cyst but negative in the cysts of neurospinal axis. IHC markers specific to the cysts of neurospinal axis are cytokeratin, EMA and vimentin. These cysts are positive for cytokeratin and EMA whereas exhibit variable positivity with vimentin. Thereby IHC also plays a role in such difficult cases [17].

Conclusion

Most of the benign cysts of the CNS are a result of maldevelopment and transformation into malignancy is very rare. Understanding of these cysts with respect to pathogenesis and embryology explains the location, presentation and histology of the cysts. Diagnostic dilemma is seen in cases between abscess and tumours which have undergone cystic change with necrosis. This aspect is very important especially in life threatening cases wherein histopathological examination plays a key role.

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