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Guest Editorial

The Encephalic Wen- Neuro-Epithelial Cyst

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Preface

Neuro-epithelial cyst is an exceptional, benign, miniature, asymptomatic, intracranial cyst. Neuro-epithelial cyst is discerned in adults and cyst enlargement engenders specific clinical symptoms as hemiparesis or intracranial hypertension. Neuro-epithelial cyst can be non contiguous with the ventricular system or intracerebral spaces bathed in cerebrospinal fluid.

Neuro-epithelial cyst is categorized as ependymal cyst and glio-ependymal cyst, variants which can be distinguished with cogent histological examination. Additionally designated as neuroglial cyst, glio-ependymal cyst is a benign, infrequently discerned, neuro-epithelial lesion.

The exceptional, benign, intra-parenchymal or paraventricular neuro-epithelial cyst is layered by simple ciliated or non ciliated cuboidal epithelium or glial tissue.

Accurate determination of neuro-epithelial cyst can be achieved with surgical resection with a broad perimeter. Nevertheless, symptomatic neuro-epithelial or neuroglial cysts necessitate cogent treatment strategies.

Disease pathogenesis

Rather than a neoplastic phenomenon, neuro-epithelial cyst is configured and progresses on account of enhanced secretion from lining epithelial cells [1,2].

Trauma or intracranial inflammatory disorders may enhance secretory activity of the cyst. However, localized, miniature, intra-axial foci of inflammation may engender passive epithelial secretion which eventually induces pertinent clinical symptoms [1,2].

Alternatively, an ectopic choroid plexus may co-exist within the cyst and generate epithelial secretion [1,2].

It can be posited that a check-valve system with unilateral inflow into the cyst may induce cyst enlargement [1,2].

Additionally, vascular articulations situated upon the cyst wall or septum of cyst extensively perfuse layering epithelial cells which engender cyst secretion [1,2].

Neuroglial cyst is posited to originate from neural ectoderm. Besides, the congenital neuroglial cyst can emerge from rests of embryonic, neural tube segments sequestered within developing white matter.

The epithelium-layered cyst configured by ectopic cells is preponderantly situated within intra-cerebral or extra-cerebral region or along the neural axis. Contiguity of the cyst with ventricular system is absent. Consequently, the cyst exhibits a propensity for periventricular sites within supra-tentorial and infra-tentorial compartments [1,2].

Disease characteristics

The eccentrically located intracranial, neuro-epithelial cyst may incriminate adult cerebellum or appear within spinal cord. Besides, the lesion may represent as a 'burnt out' pilocytic astrocytoma [3,4].

Neuroglial cyst is a developmental lesion emerging within the central nervous system which represents an estimated < 1% of primary intracranial cysts. Generally, neuroglial cyst occurs as an intra-parenchymal, intraventricular, subarachnoid, intra-neural or intra-spinal lesion [3,4].

The intra-parenchymal neuroglial cyst can be embedded within cerebellar white matter and appear distinct from fourth ventricle. Besides, the lesion may infrequently arise within posterior cranial fossa, cerebellopontine angle, midbrain or cerebellum, abutting the fourth ventricle [3,4].

Neuroglial cyst is frequently discernible as an intraventricular mass confined to lateral ventricles. Intracerebral neuroglial cyst predominantly originates within centric, white matter of frontal lobe. Thus, documentation of the lesion may be challenging [3,4].

A female predominance is observed. Commonly, neuroglial cyst appears in adults within the fourth decade [3,4].

Clinical elucidation

Asymptomatic neuro-epithelial cyst may gradually progress and engender cogent clinical symptoms. Incriminated individuals manifest progressive symptoms as hemiparesis, transient diplopia, intracranial hypertension, occipital or non specific headache, progressive dizziness, papilledema or gait ataxia [4,5].

Clinically, neuroglial cyst is predominantly asymptomatic. Layering secretory epithelial cells engender considerable enlargement of cyst with progressive emergence of cogent clinical symptoms [4,5].

Generally, neurologic deficits pertain to location and expansion of neuroglial cyst [4,5].

Neuroglial cyst arising upon cerebellopontine angle cistern may manifest with hemi-facial spasms [4,5].

Supra-tentorial neuroglial cyst encountered in adults exhibits diverse clinical symptoms. Lesions arising within the posterior fossa induce dizziness, double vision, syncope, gait disturbances, cranial neuropathy, or hemifacial spasm. Exceptionally, the cyst may rupture and engender meningitis [4,5].

Characteristically, aforesaid manifestations are contingent to degree of obstruction to outflow of cerebrospinal fluid, severity of intracranial hypertension and proportionate compression of neural-vascular structures as the brain stem, cerebral aqueduct, cerebellum or the fourth ventricle [4,5].

Histological elucidation

Upon gross examination, the septum, daughter cysts and associated vascular articulations are sufficiently identifiable [5,6].

Cyst cavity is permeated with transparent fluid resembling cerebrospinal fluid. Neuroglial cyst is usually incorporated with cerebrospinal fluid-like clear fluid although contents of cyst cavity may be xanthochromic, opalescent, milky or turbid [5,6].

Upon microscopy, cyst surface and cyst wall are layered with ciliated or non ciliated cuboidal epithelium. The epithelial layer is superimposed upon connective tissue with prominent vascular articulations. A coat of glial tissue is usually absent. The cyst is devoid of infiltrating inflammatory cells [5,6].

Ependymal cyst exhibits a substratum of connective tissue with superimposed ciliated or non ciliated, mono-stratified, cuboidal epithelium [5,6].

Glio-ependymal cyst or neuroglial cyst exhibits a layer of glial tissue sandwiched between connective tissue and superficial epithelial layer. Epithelial cells appear propped up on neuroglia. However, the glial cell layer can be focal [5,6].

Neuroglial cyst is coated with partially ciliated, cuboidal or columnar epithelium with subjacent fascicles of astroglial tissue along with an absence of intervening basal membrane [5,6].

Cyst cavity is layered with an intrinsic sheath of cuboidal ependymal cells and an extraneous layer of glial tissue [5,6].

The cyst is coated with frequently ciliated, simple columnar or cuboidal epithelial cells with subjacent neuroglia. A circumscribing fibrous capsule is absent. Alternatively, cyst wall can be layered with gliosis [5,6].

Rosenthal fibres and variable deposits of hemosiderin pigment are observed. Occasionally, an epithelial layer can be absent [5,6].

Upon ultrastructural examination, layering epithelial cells simulate cells of neuro-epithelial origin [5,6].

Immunohistochemistry

Neuro-epithelial cyst is immune reactive to epithelial membrane antigen (EMA) and appears immune non reactive to glial fibrillary acidic protein (GFAP) [6,7].

Akin to ependymal cyst and choroid plexus cyst, cellular component of neuroglial cyst wall is immune reactive to glial fibrillary acidic protein (GFAP) and S100 protein [6,7].

Neuroglial cyst is immune non reactive to epithelial membrane antigen (EMA), cytokeratin and carcinoembryonic antigen (CEA) [6,7].

Differential diagnosis

Neuro-epithelial cyst requires a segregation from arachnoid cyst, epidermoid cyst, dermoid cyst or choroid plexus cyst, porencephalic cyst, enlarged perivascular spaces, infectious cyst as neurocystercercosis or cerebral hydatid cyst, neurenteric cyst and endodermal cyst which is immune reactive to cytokeratin [7,8].

Neuroglial cyst, unlike an ependymal cyst, exhibits a distinctive layer of glial tissue subjacent to superimposed epithelium. The infrequent, congenital, neuroglial cyst requires a segregation from midline cysts or cysts arising upon cerebellopontine angle within the posterior fossa [7,8].

Investigative assay

Upon imaging, neuroglial cyst may be devoid of scalloping of adjacent cranium [7,8].

Upon computerized tomography (CT), site-specific lesions depict a homogenous, minimally attenuated mass with smooth outline. Alternatively, computerized tomography (CT) can exhibit a unilocular cyst with an attenuated wall [7,8].

Magnetic resonance imaging (MRI) exemplifies an intra-parenchymal cyst of variable magnitude and smooth perimeter which may exude minimalistic pressure upon circumscribing structures. Upon sequential MRI, intra-cystic fluid appears isointense with cerebrospinal fluid. Following infusion of contrast medium, enhancement of cyst perimeter or nodular enhancement of lesion is absent [7,8].

Upon magnetic resonance imaging (MRI), a miniature, intra-axial, septate, gradually progressive, cystic lesion of magnitude around 5 centimetres can be delineated within the frontal lobe [7,8].

Upon T1 weighted imaging, cyst content exhibits a decimated signal intensity [7,8].

Upon T2 weighted imaging, contents of cystic cavity demonstrate an elevated signal intensity, simulating the signal intensity of normal cerebrospinal fluid. Upon T2 fluid attenuated inversion recovery (FLAIR) sequences, the cyst is suppressed [7,8].

Follow up with MRI enunciates a gradual decline of cyst volume. Diffusion weighted imaging lacks restriction of cyst contents [7,8].

Flow void and image enhancement upon administration of gadolinium contrast medium is absent [7,8].

Therapeutic option

Comprehensive or partial surgical eradication of the cyst is an optimal, preferred treatment strategy. Procedures such as sub-occipital craniotomy or a transcortical approach for surgical extraction of the lesion may be adopted [8,9].

Comprehensive cystectomy is appropriate for alleviation of cerebellar neuroglial cyst demonstrating cogent neurological symptoms. Besides, complete extirpation of the cyst may be employed. Eradication of functional lesions is to be circumvented [8,9].

Surgical extermination of neuroglial cyst is contingent to cyst location and proximity to subarachnoid space or ventricular space. Surgical procedures such as trephination, aspiration, craniotomy with excision, fenestration into the cistern, open or endoscopic cysto-ventriculostomy and cystoperitoneal, cystocisternal or cystoventricular shunting may be employed [8,9].

A contemporary manoeuver of endoscopic fenestration of cerebral hemispheric cyst through adjacent ventricle can be employed where craniotomy and shunt dependence is to be circumvented [8,9].

Simple puncture and open or stereotaxic cyst drainage is associated with enhanced proportionate cyst reoccurrence [8,9].

Comprehensive surgical excision of the cyst is an optimal technique for treating antecedent, symptomatic, cerebellar neuroglial cyst [8,9].

Neuroglial cyst occurring within cerebellopontine angle cistern can be treated with total excision or partial excision along with microvascular decompression wherein the procedure is associated with significant symptomatic resolution [8,9].

Cysts arising within dorsal midbrain with accompanying hydrocephalus can be subjected to third ventriculostomy, aqueductal stenting or ventriculoperitoneal shunt, procedures which decimate possible cyst reoccurrence or disorganized cerebrospinal fluid circulation [8,9].

Clinical symptoms may ameliorate following surgical excision with complete resolution of preoperative symptoms and lack of neurological morbidities [8,9].

Cyst reoccurrence is documented and may be circumvented with comprehensive surgical eradication of the cyst wall, employed in order to decimate secretory activity of layering epithelium. Nevertheless, comprehensive surgical resection may be challenging on account of possible injury to circumscribing cerebral structures [8,9].

Partial resection may be adopted in order to circumvent motor dysfunction. Placement of shunt catheter can be suitable although secondary infection may ensue [8,9].

Sequential cyst regression in the absence of comprehensive cyst eradication is indicative of concomitantly resected vascular articulations which may contribute to decimated epithelial secretion [8,9].

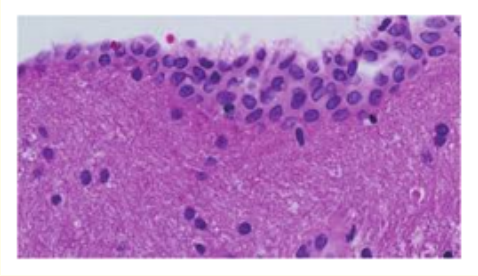

Figure 1: *Neuro-epithelial cyst depicting a mono-stratified layer of ciliated columnar epithelium abutting fibrous connective tissue [10].*

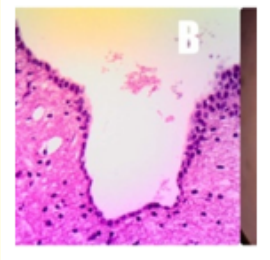

Figure 2: *Neuro-epithelial cyst demonstrating a layer of mono-stratified, ciliated columnar epithelium with subjacent fibrous connective tissue [11].*

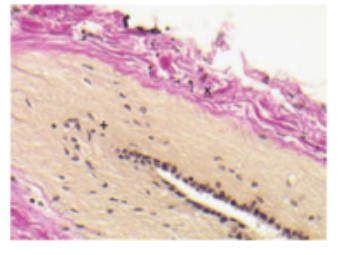

Figure 3: *Neuro-glial cyst delineating a layer of mono-stratified columnar epithelium superimposed upon neuroglial tissue and fibro-connective tissue [12].*

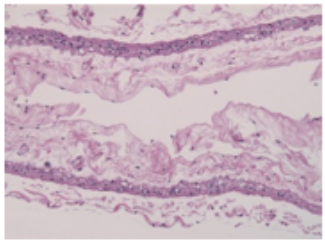

Figure 4: *Neuroglial cyst exemplifying a coat of ciliated columnar epithelium superimposed a layer of gliosis and fibrous connective tissue [13].*

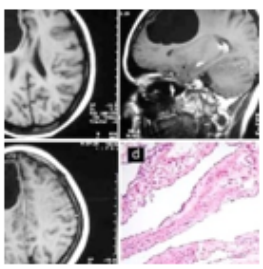

Figure 5: *Neuro-epithelial cyst enunciating an intra-axial cyst cavity with altered signal intensity lined by mono-stratified columnar epithelium with subjacent fibrous connective tissue [14].*

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10. Image 1 Courtesy: Brain tumour research and treatment.

11. Image 2 Courtesy: Research gate.

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14. Image 5 Courtesy: Semantic scholar.

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