

Research Article

Congenital Porencephalic Cavity in a Newborn: Scanographic Aspect and Differential Diagnosis - A Case Report at Medpark Clinic

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Abstract

Aim: The objective of this study is to clarify the contribution of CT scan in the diagnosis of porencephalic cavities among the range of congenital cerebral cavities. **Methods:** Congenital porencephalic cavities are rare conditions characterised by the formation of a cyst in the cerebral parenchyma, which causes certain physical symptoms. We present the case of an 18-month-old infant who was referred to our clinic for sudden onset of right spastic hypotonia. **Results:** The CT scan revealed the presence of a left fronto-parietal cerebral cystic cavity communicating with the ipsilateral lateral ventricle, pointing to the characteristics of a congenital porencephalic cavity. The differential diagnosis included: - Neuroglial cyst;- Schizencephaly; - Arachnoid cyst;- Holoprosencephaly. **Conclusion:** Due to its rarity and atypical presentation, porencephaly poses a challenge for clinicians. The pathogenesis and treatment of this condition are poorly understood. Imaging is essential for establishing a diagnosis and determining the best treatment option. MRI is the modality of choice for characterising cerebral cystic lesions. In countries with limited resources, CT scanning is a valuable tool for characterising lesions while ruling out other diagnoses.

Keywords

Porencephaly, Congenital, CT Scan

1. Introduction

Porencephaly is a rare disease characterised by the formation of a cyst in the cerebral parenchyma. This cavity must necessarily communicate with the lateral ventricles or the subarachnoid spaces to be called as such.

Porencephalic cysts can appear at any age (acquired form), but their discovery in early childhood often gives them a congenital character. [1].

Congenital porencephaly is thought to be caused by intra-

uterine vascular lesions leading to ischaemia or haemorrhage in the brain tissue. Inflammatory lesions caused by viruses such as cytomegalovirus have been cited in many textbooks [2, 3].

Excessive alcohol consumption during pregnancy has been implicated as the cause of a distortion of the foetal cerebral parenchyma through excessive sulcation formation. [2].

They may be asymptomatic or present with various symptoms depending on their size and location.

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The most common symptoms are often evident during the first year of life, with spasticity and seizures being common early manifestations.

Language disorders, intellectual disability, and motor deficits are also frequently observed.

Clinically, head circumference can range from normal to macrocephaly, due to the formation of synechiae that create a one-way valve and thus allow the ventricle responsible for hydrocephalus to fill. [4].

2. Case Report

We report the case of an 18-month-old infant brought by his parents to Medpark Clinic for right-sided spastic motor disorders with walking difficulties.

In the medical history, his mother's prenatal consultation requirements were met, resulting in a normal delivery and a high score of appearance-pulse-grimace-activity-respiration (APGAR score) for the child.

To date, the infant's vaccination schedule had been followed and the child showed no signs of infectious disease or deficiency until two weeks prior to our consultation, when he presented with abnormal spasticity of the right upper limb.

Upon arrival, a thorough consultation including a physical examination and extensive blood tests was carried out to rule out any infectious disease or blood circulation abnormalities. The blood tests were normal. The circumference of the skull was normal.

The neurologist did not deem it necessary to request an electroencephalogram (EEG), given the absence of signs of cortical irritation (convulsions).

3. Results

Presence of a left parietal cerebral cystic lesion with a density similar to CSF, well circumscribed, with polylobed contours, communicating with the subarachnoid space, exerting an attractive effect and dilation on the adjacent lateral ventricle. It is surrounded by cerebral parenchyma, which forms a hypodense ring compared to the rest of the parenchyma, possibly related to areas of gliosis (Figures 1 and 2).

There is no mass effect on the surrounding structures, which otherwise show no abnormalities in the supratentorial and infratentorial regions.

Contrast injection did not reveal any abnormal enhancement of the lesion or the rest of the parenchyma that could be related to meningoencephalitis.

Therefore, a diagnosis of porencephalic cyst was made.

4. Discussion

A porencephalic cyst or porencephaly is a rare condition. It is often congenital and discovered at birth. [2].

Its incidence is 3.5/100,000 births.

It can also be acquired in some cases and can vary in size, occupying both cerebral hemispheres, often respecting the territories supplied by the arteries of the circle of Willis. [1].

Congenital brain lesions include two types of porencephaly:

- 1). Genetic porencephaly: resulting from poor development during early neuronal migration;
- 2). Encephalophaloplastic porencephaly, which is a late prenatal or perinatal vascular lesion due to an arterial ischaemic stroke or venous thrombosis. [5].

Some authors believe that the genetic theory supports the hypothesis of a mutant gene, COL4A1, which is thought to cause vascular wall damage leading to porencephaly in human families. In addition to this, there is often an environmental factor, which is usually trauma. [6].

The clinical presentation of porencephalic cysts varies widely, ranging from simple tonic-clonic seizures to motor deficits. The EEG may be normal or show focal rhythmic epileptiform discharges. [7].

One study has claimed that the presence of a porencephalic cyst is not linked to recurrent episodes of epilepsy and that it is directly linked to hippocampal mesial sclerosis, which often coexists with congenital porencephalic cavities. They report the coexistence of amygdalo-hippocampal atrophy with unilateral or bilateral porencephalic cavities and that no cause-and-effect relationship has been established, but nevertheless recommend rigorous investigation for hippocampal atrophy whenever a porencephalic cyst is discovered. [1].

Apart from the spastic crisis experienced by our patient, the symptoms depend on the area affected by the lesion and may manifest as psychiatric disorders, but the latter are more common in schizencephaly because it often affects the temporal lobe. [5].

A case of otorrhoea was described following a dehiscence of the tympanic membrane caused by this cystic mass, which was believed to be responsible for CSF leakage through the outer ear. [8].

Another author reported the onset of diplopia and strabismus caused by a porencephalic cyst in the parieto-occipital region, the plausible mechanism being compression of the cerebral visual area. [9].

Another author reported a case of a porencephalic cyst with intracerebral haemorrhage and deep vein thrombosis despite a negative thrombophilic assessment. The cause-and-effect relationship was not clearly established, but it was noted that the patient tested positive for a heterozygous mutation of the COL4A1 gene. [10].

Given all this, we can see that porencephalic cysts can have a diverse clinical expression in the neurological sphere and that, in general, the treatment of porencephaly aims to relieve symptoms, as there is no treatment to induce brain growth in the missing sections; this may include anti-epileptic drugs, physiotherapy, or a shunt to remove excess cerebrospinal fluid.

The final diagnosis is made using imaging (CT or MRI).

The CT scan shows a cystic lesion within a vascular territory, surrounded by cerebral parenchyma and communicating with either the lateral ventricle or the subarachnoid spaces. [11].

Since the 1970s, it has long been considered the essential tool, supplanting ultrasound and scintigraphy until the advent of MRI. [12].

The advantage of MRI is that it provides the precision of the cerebral layer that delimits this cyst (white matter or grey matter), which distinguishes it from other entities that would constitute the range of differential diagnoses.

The differential diagnosis should be made with:

- 1). A neuroglial cyst: an intra- or extra-axial cystic formation in the central nervous system that does not communicate with the ventricle or subarachnoid spaces and is preferentially located in the frontal lobe. (Figure 3).
- 2). An arachnoid cyst: an extra-axial cyst that often exerts a mass effect on adjacent structures and is preferentially located in the temporal and occipital fossae. (Figure 4).
- 3). Schizencephaly: like porencephaly, it connects the subarachnoid space to the ventricle, which in this case is lined by the cerebral cortex or grey matter, unlike porencephaly, which is lined by white matter. (Figure 5).
- 4). Holoprosencephaly, which results from the failure of the hemispheres to separate, results in a single ventricle. (Figure 6) [11].

5. Conclusion



Figure 1. Well-defined left parietal porencephalic cavity communicating with the subarachnoid spaces.

Congenital porencephaly remains a little-known and rare condition in our environment, but it is responsible for a wide range of symptoms that can confuse clinicians and radiologists. Imaging (CT scan and MRI) remains essential for making a correct diagnosis. Prenatal morphological ultrasound should be rigorous and widely available in low-income

countries in order to make an accurate early diagnosis of the malformation and guide the obstetrician-gynaecologist in the aetiological assessment and management of the newborn.

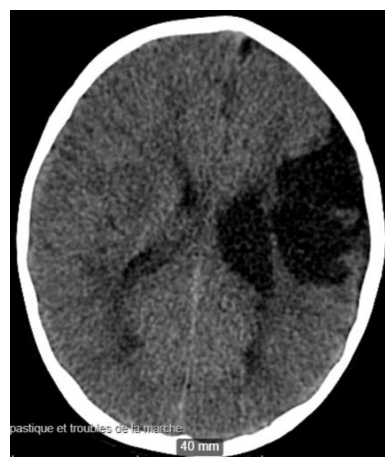


Figure 2. Porencephalic cavity with attractive effect on the ipsilateral VL.

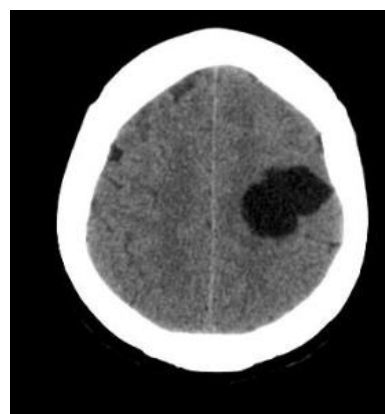


Figure 3. Left frontal neuroglial cyst. (Radiopaedia).



Figure 4. Arachnoid cyst of the right temporal fossa (Radiopaedia).

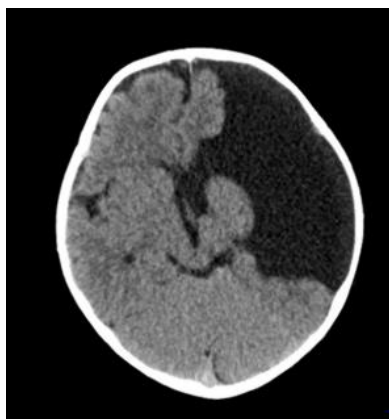


Figure 5. Open-lip schizencephaly bordered by grey matter and communicating with the ventricle (Radiopaedia).

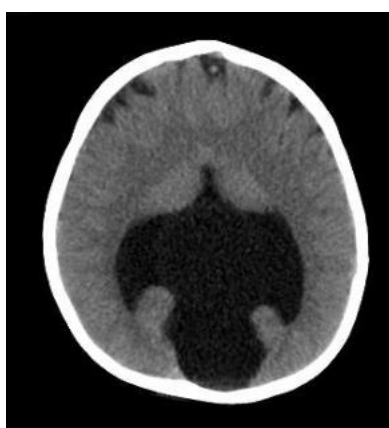


Figure 6. Semi-lobar holoprosencephaly (Radiopaedia).

Abbreviations

APGAR	Appearance, Pulse, Grimace, Activity, Respiration
EEG	Electroencephalogram
CSF	Cerebrospinal Fluid
CT	Computed Tomography
MRI	Magnetic Resonance Imaging

Patient Consent

Obtained from the patient's mother, as the patient was a minor and was not able to consent on her own account.

Conflicts of Interest

The authors declare no conflicts of interest.

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