

## Left Cerebellar Porencephalic Cysts with Ipsilateral Parieto-Occipital Encephalocoele: Antenatal and Post Natal Radiology

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**Abstract:** Porencephalic cyst is a rare congenital disorder which may cause a wide range of physiological, physical and neurological symptoms.

We present a case of three months old female child diagnosed in utero via sonography and perinatally using Magnetic Resonance Imaging (MRI) as a case of Porencephalic cyst associated with encaphalocoele. The mother presented at the Radiology Department of the University of Calabar Teaching Hospital for routine obstetrics scan at 40 weeks of gestation had a caesarean section at 40 weeks of gestation and was delivered of a live female neonate with APGAR Score of 8, body weight of 4.4kg and no obvious neurological deficits seen. Postnatal MRI of the baby on the 4<sup>th</sup> day of life also showed a dilated cystic partial replacement of the left cerebellar hemisphere with herniation of the same through a left parieto-occipital defect communicating with the 4<sup>th</sup> ventricle and pre-pontine cistern.

The existence of porencephaly with associated encaphalocoele is rare and an extensive literature search revealed no previous case report as depicted in this report, hence the need to highlight the role of radiology in the diagnosis.

**Key Words:** Porencephalic cyst, encephalocoele, ultrasonography, MRI.

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### I. Introduction

Porencephalic cyst is a rare congenital disorder that results in cystic degeneration and encephalomalacia and the formation of cysts.<sup>1</sup> The term is used variably among radiologists with its broadest definition being a cleft or cystic cavity within the brain,<sup>1</sup> and its more narrow definition being a focal cystic area of encephalomalacia that communicates with the ventricular system and/or the subarachnoid space.<sup>1</sup>

Porencephalic cystic lesions may be seen in-utero after focal necrosis as a result of an ischemic event involving the vascular distribution of a single major cerebral vessel, probably sequel to cerebrovascular accident or a motor vehicle accident or it could be due to infections or haemorrhages.<sup>2</sup>

The cysts and cavities cause a wide range of physiological, physical, and neurological symptoms. Depending on the patient, this disorder may cause only minor neurological problems without any disruption on intelligence, while others may be severely disabled or die before 20years of age. This disorder, however is far more common in infants, and can occur before or after birth.<sup>4</sup>

Patients with severe cases of porencephaly suffer epileptic seizures and developmental delays, whereas patients with a mild case of porencephaly display little to no seizures with healthy neurodevelopment. Infants with extensive defects show symptoms of the disorder shortly after birth, and the diagnosis is usually made before the age of one.<sup>4,5</sup>

The exact prevalence of porencephaly is not known; however, it has been reported that 6.8% of patients with cerebral palsy or 68% of patients with epilepsy and congenital vascular hemiparesis have porencephaly.

Encephalocoeles are congenital malformations in which the meninges and brain protrude out of the cranial cavity through a defect in the bone or soft tissue.<sup>7</sup> The incidence is about 1 to 3 per 10,000 live births. An encephalocoele results from failure of the surface ectoderm to separate from the neuroectoderm. This leads to a bony defect in the skull table, which allows herniation of the meninges (cranial meningocele) or of brain tissue.<sup>7,8,9</sup> The occiput is the most common site of this type of neural tube defect (75%) in the United States and Western Europe. They occur most frequently in the midline and may be subdivided based on their origin into: occipital, cranial vault, frontoethmoidal and basal subtypes. Females are more affected. The disease is associated with considerable mental and physical handicap, cosmetic problems and upper airway obstruction.<sup>8,9</sup>

The existence of porencephaly with encephalocoele is rare and an extensive literature search revealed no previous case report as depicted in this study, hence the need to highlight the role of radiology in the diagnosis.

## II. Case Presentation

Mrs. S.K. is a 26 year old full time house wife and primi-gravida who presented to the Radiology Department of our institution from the antenatal clinic for routine obstetrics scan at 40 weeks of gestation. There was no family history of still births or deliveries of grossly abnormal babies. No history of drug ingestion except for her haematinics. She gave no history of fever, abdominal pain, abnormal vaginal discharge or vaginal bleeding. She is not a known diabetic, smoker nor hypertensive. She had never been involved in a road traffic accident. Her husband is a 37year old textiles business man.

The physical examination of the mother was satisfactory and general condition stable. Baseline blood tests like full blood count, VDRL, urinalysis, electrolyte urea and creatinine were all within normal limits.

Obstetrics scan revealed a viable female fetus in longitudinal lie and cephalic presentation. The fetus was 40 weeks old based on femur length and abdominal circumference measurements. There was a defect in the left parieto –occipital cranium with herniation of a cyst (fluid containing) through it. The aforementioned cystic structure showed a connection with the temporal horn of the ipsilateral ventricle suggestive of a Porencephalic cyst with an associated encephalocoele. (Fig. 1)

Perinatal magnetic resonance imaging also showed the earlier described ultrasonographic features (Fig 2 and Fig.3).

A day later, through caesarean section the patient delivered a live female neonate with an APGAR score of 8 and body weight of 4.4kg.No obvious neurological deficits were seen. The other systems examined were normal. Biochemical, haematological and microbiological blood tests did not show any abnormality.

Magnetic resonance imaging was done for the baby on the 4<sup>th</sup> day of life which also showed a dilated cystic partial replacement of the left cerebellar hemisphere with herniation of the same through a left parieto-occipital defect. The above mentioned cystic dilatation was also seen to communicate with the fourth ventricle and pre-pontine cistern. Both cerebral hemispheres were preserved in their intensities, no focal lesion seen. The rest of the cranium and its content appeared within normal limit (Fig. 4 and Fig 5).

The child is now a three months old active and lively baby, awaiting corrective surgery by the neurosurgical team.



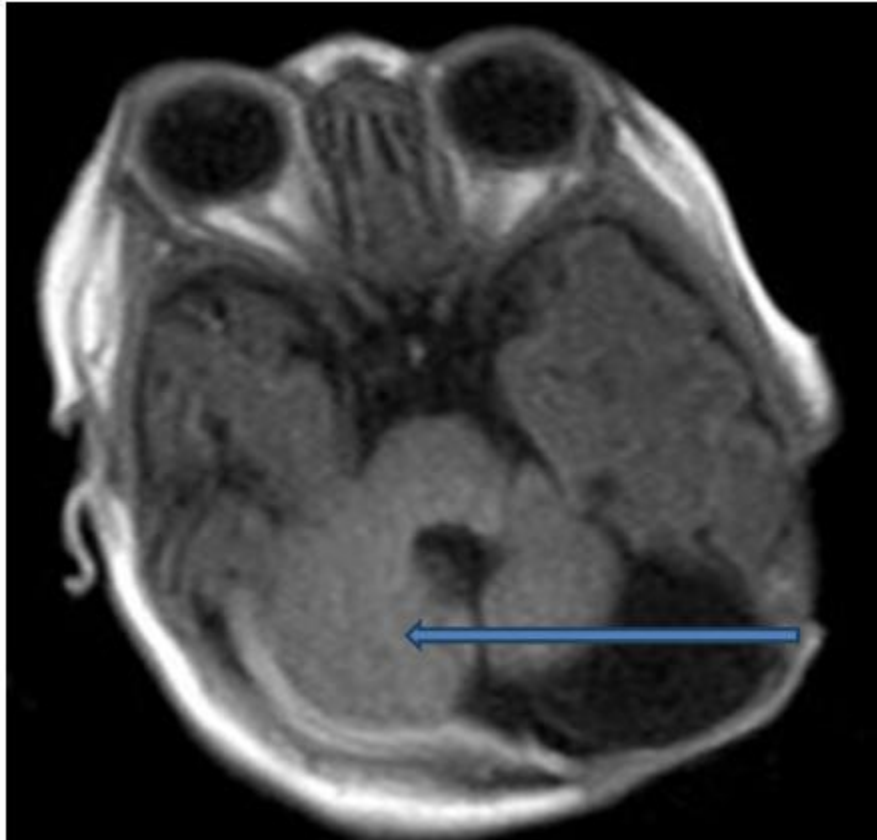
**Fig. 1: Prenatal sonogram showing a cranial defect in the parieto-occipital region with herniation of a cyst through the 3.85cm cranial defect (blue arrow).**



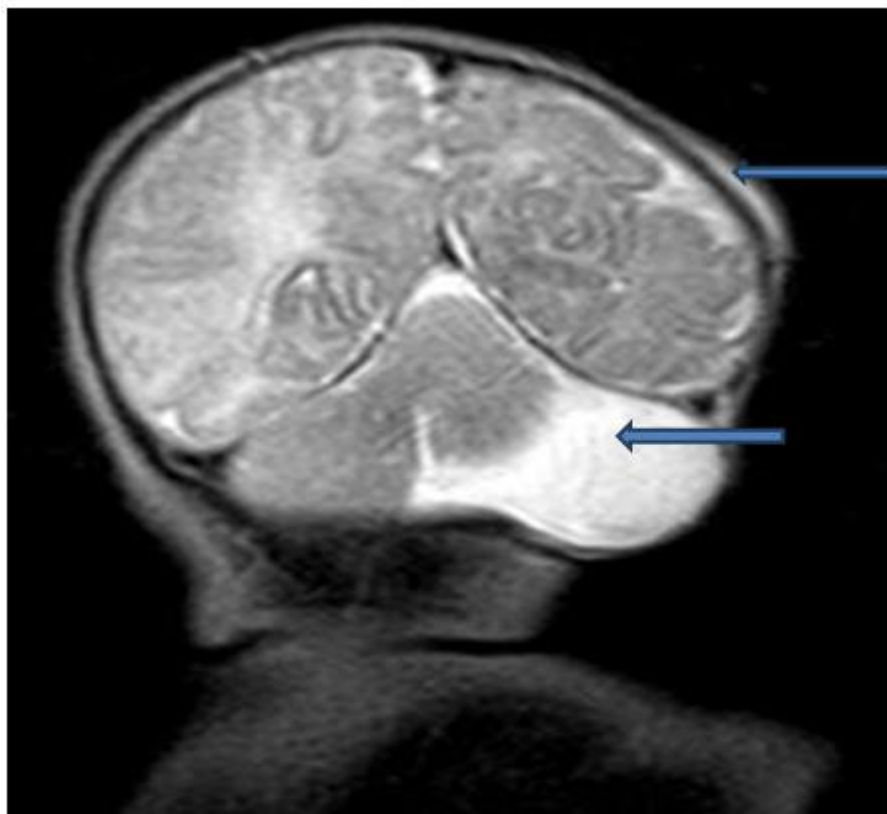
**Fig. 2:** Prenatal sonogram showing a parieto-occipital defect with cystic area communicating with the ventricular system (blue arrow)



**Fig 3:** Prenatal sonogram showing the cerebellum in relation to the parieto-occipital defect.



**Fig. 4:** T1W axial image showing hypoplastic left cerebellum, herniating Porencephalic cyst communicates with the 4<sup>th</sup> ventricle (blue arrow).



**Fig. 5:** Coronal T2 W image showing cystic herniation at the left parieto-occipital region (blue arrow).

### III. Discussion

There are many risk factors associated with development of Porencephalic cysts. These risk factors in the mother include cerebral ischemia, cardiac arrest, trauma during pregnancy, infections, vascular thrombosis, hemorrhage, chronic lung disease toxemia and drug abuse. Fetal risk factors include; cerebral hypoxia, cystic peri-ventricular leukomalacia, cerebral atrophy, male gender, encephalitis and meningitis. The index case was female and negative for the above listed risk factors.

Cysts can develop in the frontal lobe, parietal lobe, forebrain, hindbrain, temporal lobe, or virtually anywhere in the cerebral hemisphere.<sup>5</sup>

Patients with Porencephalic cysts range from asymptomatic, to profoundly impaired.

Often signs and symptoms become evident in the first year of life, with spasticity and seizures being common early manifestations. Language impairment, mental retardation, and motor deficits are also frequently encountered.<sup>5,6</sup> The index case is three months old and this may explain why the clinical features are yet to be evident.

Head circumference is variable. It may be normal or small, or alternatively synechia can create a one-way valve effect with progressive enlargement of the cyst and skull expansion or hydrocephalus, resulting in an enlarged head.<sup>4,6</sup>

In antenatal ultrasound scan, one or more intracranial cysts may be seen to communicate with the ventricular system and/or subarachnoid space. Porencephalic cysts appear as an intracranial cyst which has a well-defined border and central attenuation the same as CSF. There is usually no mass effect on the adjacent parenchyma, although occasionally the cysts are enlarging and do result in local mass effect. There is no enhancement with contrast and no solid component.<sup>1,4,7</sup>

In computerized tomography (CT), the cyst appears well defined and often corresponds to a vascular territory. The cyst is lined by white matter, which may or may not demonstrate evidence of gliosis (this depends on the age at which the insult occurred). Importantly the cyst is not lined by grey matter, helpful in distinguishing them from arachnoid cysts and schizencephaly. The content of the cyst follows CSF signal on all sequences: **T1:** low signal intensity, **T2:** high signal intensity, **FLAIR:** suppression of fluid signal intensity, **DWI:** no restricted diffusion

Many malformations and/or chromosomal anomalies are noted in at least 60% of patients with encephalocoele,<sup>8,9</sup> such as hydrocephalus, microcephaly, Chiari I malformation and tetrasomy 20p

The combined presentation of a porencephaly and an encephalocoele is rare. Currently, most cases are diagnosed prenatally<sup>8,9</sup> as in the index case. Maternal serum alpha-fetoprotein levels are elevated in only 3% of patients, because most encephalocoele are covered with skin. This however was not done in this case. Postnatally, infants may have associated cerebrospinal fluid (CSF) rhinorrhea and recurrent meningitis.<sup>7,8,9</sup>

Encephalocoele can also appear along the cranial vault due to abnormal closure of cranial bones, the most common location for encephalocoele is the occipital region. If the bulging portion contains only cerebrospinal fluid and the overlying membrane, it may be called a meningocele. If brain tissue is present, it may be referred to as a meningoencephalocoele.<sup>7,8</sup>

There are various radiologic modalities that could be used to investigate this patient namely; plain radiography, ultrasonography (prenatal perinatal and post natal), computerized tomography and magnetic resonance imaging.

A skull radiograph will show the soft tissue mass over the skull vaults.<sup>7,9</sup>

Sonography will poorly show the site of bony defect but vividly reveal its content (fluid, solid or mixed). Computerized tomography and Magnetic resonance imaging will elegantly show the defect, its content and the relationship with adjacent structures. Extreme care should be taken when using the head coil as excessive manipulation during MRI may rupture the encephalocoele.<sup>10,11,12</sup>

Treatment of the encephalocoele involves resection of some of the neural tissue and repairing of the defect<sup>8,9</sup>. However, for porencephalic cysts, treatment is essentially supportive as the degree of impairment is dependent on the location.<sup>3,4,5</sup>

In the index case, the clinical and radiological findings of Porencephalic cysts were in consonance with the findings in other literature. The combination of both pathologies is a managerial dilemma for the neurosurgeons.

### IV. Conclusion

This is a case of a three months old female child diagnosed in utero via sonography and peri-natally using magnetic resonance imaging as a case of Porencephalic cyst, currently awaiting corrective surgery, however clinically stable.

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