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Neurosurgery

# PORENCEPHALIC CYST: A RARE AND INTERESTING CASE

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## BACKGROUND

Porencephaly is a rare congenital disorder that results in cystic degeneration and encephalomalacia and the formation of porencephalic cysts which are cysts filled with cerebrospinal fluid. Bilateral involvement is rare. As such, there is no association with congenital rubella syndrome and porencephaly. Clinical symptoms and signs are usually apparent within the first year of life. Prognosis of this condition is very poor and management generally involves supportive care after diagnosis. Here, we present a case of a 6 month old child with congenital rubella syndrome presenting with hydrocephalus.

### Case Presentation

### History and examination

We present a case of a 6 month old female child, known case of congenital rubella syndrome, born to non-consanguineous couple who was referred to our neurosurgical unit with increased head circumference. At birth she was placed in the neonatal intensive care unit for 6 days for reduced birth weight. A CT scan taken at 3 months for a history of seizures revealed tetraventriculomegaly with right exophthalmos and microphthalmia and has since been under phenobarbitone cover. The patient underwent ophthalmological examination and was then referred here for further evaluation. There was posturing of upper limbs. The patient did not have a history of fever, vomiting, or incessant cough. Developmental milestones were delayed.

The child was conscious and irritable. She was active and crying well. Head circumference was 45cm. Right microphthalmia and corneal opacities were noted. Left eye showed minimal corneal opacities, however was reactive to light. Dilated veins were noted on the scalp. Anterior fontanelle was tense and sutural diastasis was noted. Head holding was absent. Social smile or spinal dysraphism was absent.

CT brain taken showed bilateral porencephalic cysts, larger on the left side with connection to grossly dilated ventricles. MRI brain revealed periventral calcification with features suggestive of congenital rubella syndrome. An EEG done showed continuous left hemispheric discharges with increased delta waves. Patient was optimized and taken up for left ventriculo-peritoneal shunt placement. Post-operative period was uneventful. The patient was managed on IV fluids, antibiotics, analgesics, antiepileptics and other supportive measures.

The patient was alert and active on discharge.

## DISCUSSION

Porencephaly is a rare congenital disorder that results in cystic degeneration and encephalomalacia and the formation of porencephalic cysts which are cysts filled with cerebrospinal fluid. Common causes include ischemia, hemorrhage and abnormal development, usually involving mutations in COL4A1 gene.[1,2]

An infant is suspected to have congenital rubella syndrome if the child meets the criteria for probable or confirmed case having one or more of the constellation of symptoms, including but not limited to cataracts, pigmentary retinopathy and microcephaly. Diagnosis is confirmed on laboratory evidence including isolation of rubella virus, detection of rubella specific IgM antibody, rubella titer that does not drop at the expected rate of a 2-fold decline per month or a specimen that is PCR-positive for rubella virus. There has however been no definitive direct link between congenital rubella syndrome and porencephaly.

Porencephaly may be internal or external depending on whether the communication lies with ventricle or subarachnoid space.[2,3] Porencephalic cysts may involve any lobe. Bilateral involvement is however rare.[2]Gliosis develops in late insults.

Clinical symptoms and signs are usually apparent within the first year of life and include seizures, motor impairment, developmental deficits and increasing head circumference. Hydrocephalus develops in the presence of synechiae creating a one-way valve mechanism leading to progressive enlargement of the cyst, hydrocephalus and skull expansion.[4]

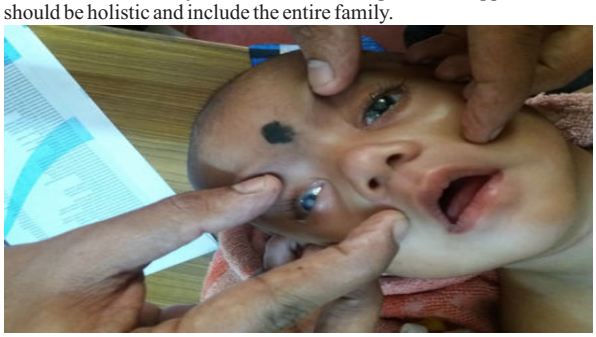
Antenatal diagnosis using ultrasound has been described in the case of congenital cysts. In CT scan, the cysts appear as an intracranial cyst with a well-defined border and central attenuation, with no contrast enhancement or solid components. There usually is no mass effect but large cysts may occasionally result in a local mass effect. An MRI shows a cyst, lined with white matter communicating with ventricles or sub-arachnoid space. In areas surrounding the lesion sites, increased theta and delta waves may be seen. [5]

Differential diagnoses include neuroglial cysts, arachnoid cysts and schizencephaly. These are differentiated on the fact that there are no communication with the ventricles or subarachnoid space, are extra-axial with underlying gray matter and are not lined with white matter. [6]

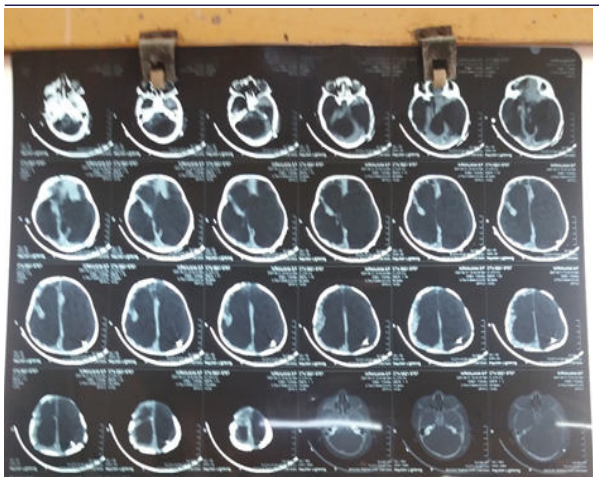
Prognosis of this condition is very poor, with mortality recorded within weeks or months. Management include symptomatic relief and supportive care to both the patient and family.

## CONCLUSION

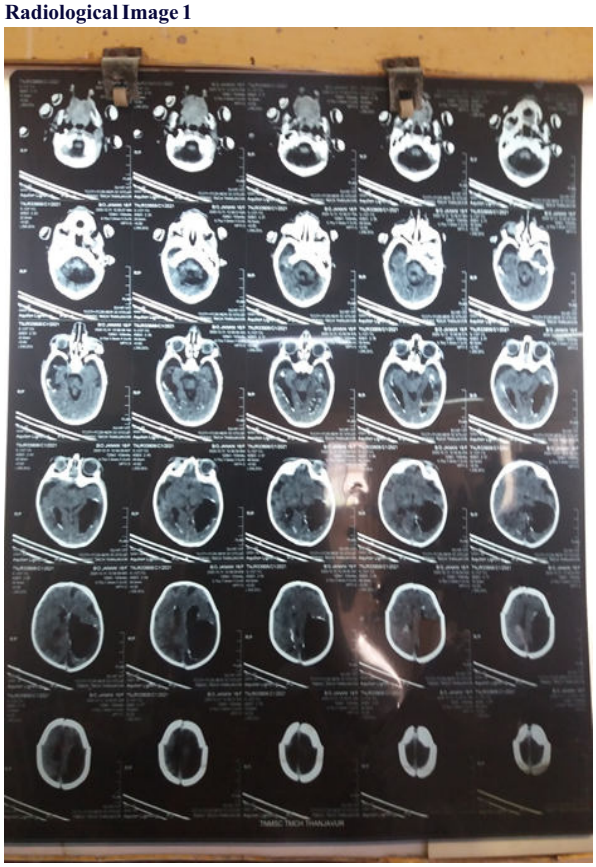
Porencephaly is a rare condition affecting infants with a very poor clinical course. A link with congenital rubella syndrome, while may be coincidental in this case has not been proven. It is thus prudent to deal with this clinical entity with as much care as possible. Supportive care should be holistic and include the entire family.



**Clinical Image 1**



**Radiological Image 1**



**Radiological Image 2**

## ABBREVIATIONS

Not Applicable

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