

Congenital Externally Communicating Porencephaly Presenting as Hemiplegic Cerebral Palsy: Imaging Study of a Rare Condition

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1. Abstract

Congenital porencephaly is a very rare condition characterized by cystic degeneration encephalomalacia and cysts or cavities within the brain. Porencephalic cysts have a variable size and site and therefore it result in a variable clinical presentations including asymptomatic, various forms of cerebral palsy, seizures and cognitive impairment. The disorder is heterogeneous in nature and the brain lesions can be caused by developmental abnormalities, infection, perinatal brain ischemia, trauma and hemorrhage. Genetic factors have been suggested and familial cases have been reported. Congenital porencephaly is generally classified into, internally communicating with the ventricle and externally communicating with the subarachnoid space. The aim of this paper is to report the rare finding of externally communicating porencephaly in a child with hemiplegic cerebral palsy.

2. Keywords: Congenital porencephaly; CT-scan; Hemiplegic cerebral palsy

3. Introduction

Congenital porencephaly is a very rare condition characterized by cystic degeneration encephalomalacia and cysts or cavities within the brain. Porencephalic cysts have a variable size and site and therefore it results in a variable clinical

presentation including asymptomatic, various forms of cerebral palsy, seizures and cognitive impairment. The disorder is heterogeneous in nature and the brain lesions can be caused by developmental abnormalities, infection, perinatal brain ischemia, trauma and hemorrhage. Genetic factors have been suggested and familial cases have been reported. Congenital porencephaly is generally classified into, internally communicating with the ventricle and externally communicating with the subarachnoid space [1-7]. The aim of this paper is to report the rare finding of externally communicating porencephaly in a child with hemiplegic cerebral palsy.

4. Patients and Methods

The case of a five-year old girl with hemiplegic cerebral palsy caused by porencephaly is described and CT-scan images are presented.

5. Results

There was no history of birth asphyxia and family history was negative for a similar condition. The girl had spastic weakness on the left side of the body. She had gait abnormality mostly in form of dragging her left leg. She could take and carry weight with her right

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arm for some time, but she could not take the same weight with her left arm and she could not prevent herself from using her right arm when she was encouraged to keep trying. The girl was able to take a pen to try copying a line and a circle, but she couldn't (Figure 1).



Figure 1: A five-year old girl with left hemiplegic cerebral palsy. She could take and carry weight with her right arm for some time, but she could not take the same weight with her left arm and she could not prevent herself from using her right arm when she was encouraged to keep trying. The girl was able to take a pen to try copying a line and a circle, but she couldn't.

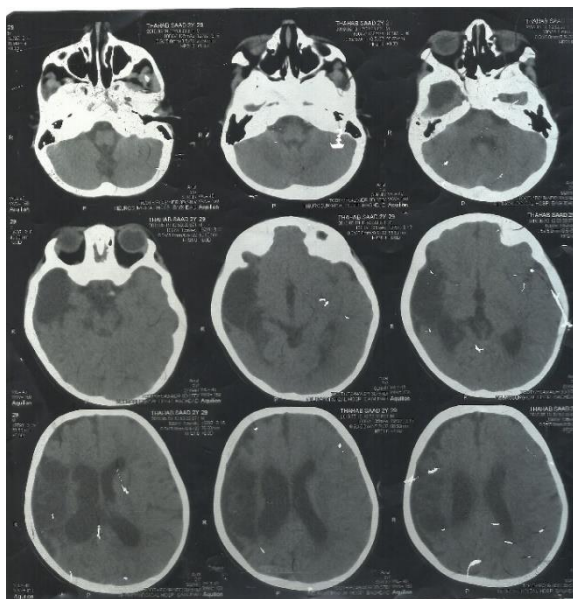


Figure 2A: Brain CT scan showed large right sided temporo-parietal gliotic changes with large communicating porencephaly cyst.

Brain CT scan (Figure 2A-2D) showed large right sided temporo-parietal gliotic changes with large communicating porencephaly cyst.

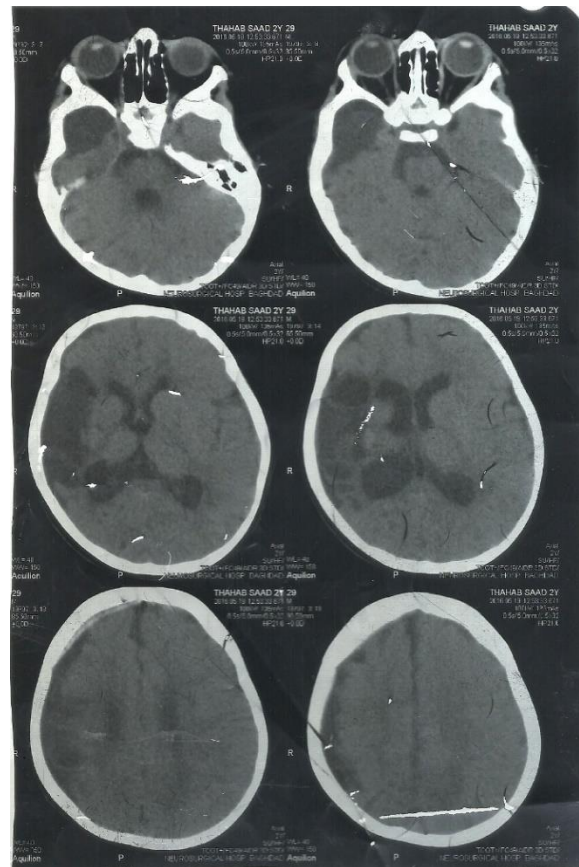


Figure 2B: Brain CT scan showed large right sided temporo-parietal gliotic changes with large communicating porencephaly cyst.

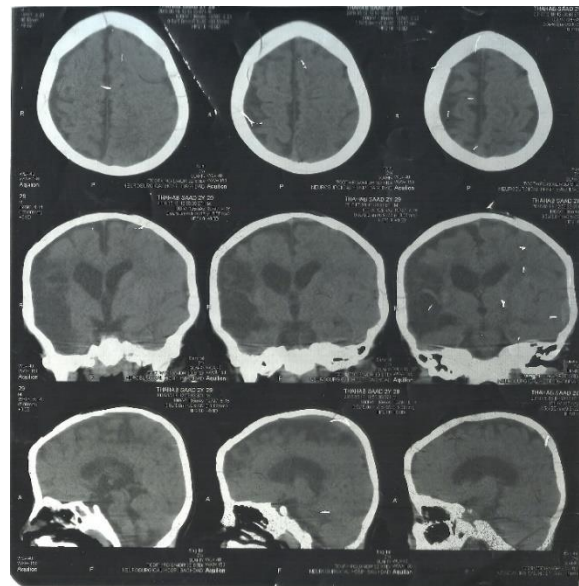


Figure 2C: Brain CT scan showed large right sided temporo-parietal gliotic changes with large communicating porencephaly cyst.

The decision was made to follow up the girl and

encouraging physiotherapy as her disability was not regarded to be significantly serious.

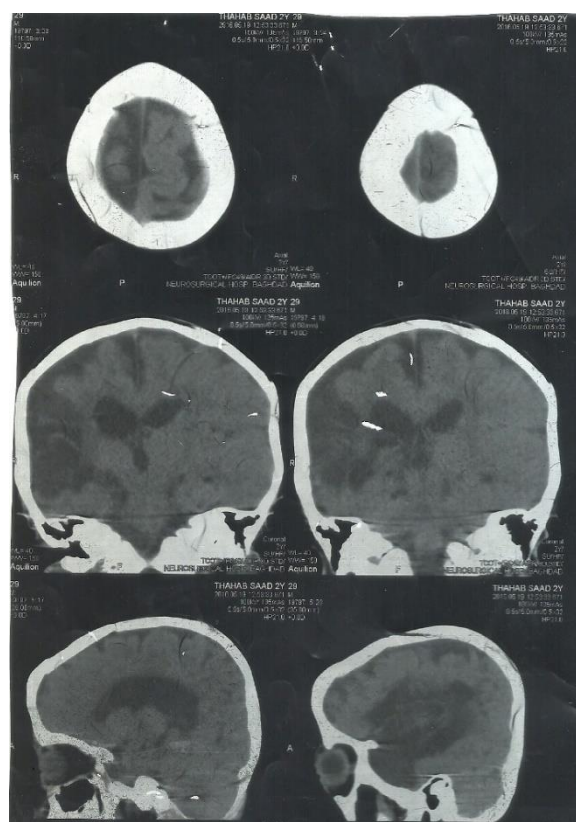


Figure 2D: Brain CT scan showed large right sided temporoparietal gliotic changes with large communicating porencephaly cyst.

6. Discussion

In 1859, was first reported by Heschl who derived the term “porencephaly” from Greek roots meaning holes or cavities in the brain [1]. Before the 1970s, the diagnosis of congenital porencephaly was largely made using pneumoencephalography [2,6]. However, Ramsey RG, Huckman (1977) emphasized that CT scan is often the only imaging study necessary to make the diagnosis of Porencephaly. Porencephalic areas usually have a well-defined border, have the density of cerebrospinal fluid and do not change in density following the use of contrast medium enhancement [8].

Several authors emphasized the association of congenital porencephaly with hemiplegic cerebral palsy [9-12].

Claeys, Deonna and Chrzanowski (1983) 37 children who had congenital hemiparesis with brain CT-scan. They reported that four patients (11%) had normal

CT-scan, nine patients (24%) had unilateral ventricular enlargement and 24 patients (63%) had a variety of cortical including cystic porencephaly in two patients [9].

7. Conclusion

The rare finding of externally communicating porencephaly in a child with hemiplegic cerebral palsy is documented.

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