Academic Journal of
Pediatrics & Neonatology
ISSN 2474-7521

Juniper Publishers
key to the Researchers

Case Report
Volume 12 Issue 4 - March 2023
DOI: 10.19080/AJPN.2023.12.555897

Acad J Ped Neonatol

Rapid Increase in Volume of a Neuroepithelial Cyst: An Unusual Case with High Proliferative Index

Alick Durao[1], Giacomo Piaser Guerrato[2], Marco Zemella[2] and Luca Massimi[2,3]*

[1]*Department of Neurosurgery, Hospital de Força Aérea do Galeão, Rio de Janeiro, Brazil*

[2]*Pediatric Neurosurgery, Fondazione Policlinico Universitario A. Gemelli IRCCS, Rome, Italy*

[3]*Catholic University Medical School, Rome, Italy*

Submission: February 12, 2022; **Published:** March 08, 2023

***Corresponding author:** Luca Massimi, M.D., Institute of Neurosurgery, Catholic University Medical School, Largo A. Gemelli, 8, 00168, Rome, Italy; Tel: +393397584217

Abstract

Neuroepithelial cysts (NC) are rare cystic lesions of neuroectodermal origin, usually differentiated into ependymal and choroidal plexus variant. They can be found at any age and in any region of the central nervous system, although young age and intracranial space are the most commonly involved. In spite of their possible incidental diagnosis and their frequently benign course (with indolent or slow growth over the time), NC may show a rapid growth causing symptoms due to the mass effect. Herein, we present a rare case of quickly growing NC characterized by hyperactive cystic wall secretion.

Keywords: neuroepithelial cysts, choroid plexus cysts, ependymal cysts, microsurgery, proliferative index

Introduction

Neuroepithelial cysts (NC) are rare cystic lesions with ependymal or epithelial lining, which include ependymal and choroidal plexus cysts [1]. NC correspond to 0.4-1% of all intracranial cysts [2,3]. Both the choroidal and the ependymal variant have a neuroectodermal origin that leads to the development of a cyst starting from ectopic ependymal cells [4]. They can be found in any region of the central nervous system, but the intracranial space is the most common location [5]. The clinical course is usually benign, most of NC often remaining asymptomatic or showing an indolent growth over the time [6]. Herein, we present a case with an unusually rapid clinical course and with apparently "aggressive" pathological features.

Case presentation

This three-month old girl was admitted to our department fifteen days after the birth with a fetal ultrasounds and MRI diagnosis of intracranial cystic lesions (Figure 1). The child was in good general condition, with regular head circumference and slightly bulging anterior fontanel. No neurological or ophthalmological deficits were detected at physical examination. Postnatal brain MRI showed a large posterior midline cystic lesion with a bulky multi-lobulated left temporo-occipital component and a small right occipito-mesial cyst, associated to left temporal and occipital lobes hypoplasia (Figure 2-A,B). The cysts appeared as hypointense on T1-weighted and hyperintense on T2-weighted images and showed an enhancement of their inner wall following Gadolinium administration. The child was discharged with planned, serial clinical and radiological follow-up.

However, in the first days following the discharge, an abnormally quick head growth was noticed. The child remained in good clinical condition apart from an isolated epileptic seizure. A new MRI, performed ten days after the previous one, showed a remarkable enlargement of left cystic components with dislocation and compression of the homo-lateral cerebral peduncle (Figure 2-C,D). Therefore, the child underwent a left temporo-parieto-occipital craniotomy with wide removal of the left-sided cyst wall and marsupialisation into the sylvian and midline cisterns. At operation, a clear cleavage with brain surface was evident; the cyst wall appeared thick, tenacious and opaque (Figure 3). The post-operative course was uneventful and following neuroimaging investigation confirmed a remarkable reduction of the cyst volume, stable over the time. At current follow-up (6 years after the operation), the child shows good clinical condition and mild psychomotor delay. No recurrence of the cyst has been detected (Figure 4).