

Case lesson 26

Neuroepithelial cyst regression in watchful waiting follow-up

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Introduction: Intracranial neuroepithelial cysts are rare benign lesions thought to be of developmental origin, generally seen in the third and lateral ventricles. They form during embryogenesis from simple folding of neuroepithelium into or out of the primitive ventricular system and represent less than 0.4% of all cyst-like lesions [1, 2]. Although there is ongoing debate on their etiopathogenesis, a number of theories have been put forth throughout the years. They may remain asymptomatic throughout the life or can cause symptoms at any age. Regarding ventricular or subarachnoid spaces, there is no clear communication between them [3].

We report a case of a boy with asymptomatic epithelial cyst located in the left lateral ventricle, diagnosed at birth, whose cystic lesion resolved itself without the need of performing surgery.

Case presentation: we report a case of a 8-years-old boy who was diagnosed at birth with a massive cystic lesion of the left lateral ventricle causing enlargement of the ventricle and obstruction of left foramen Monro. Immediately after birth he was transferred to neonatal intensive care unit due to a raised suspicion of hydrocephalus since the prenatal ultrasound check in the 37th week of pregnancy. Normal Apgar score was noted, as well as normal reflexes with no signs nor symptoms of hydrocephalus. Immediate Ultrasound and brain CT scan as well as one month MRI confirmed the presence of the left lateral ventricle cyst associated with occipital horn exclusion, resembling an epithelial cyst. Wait and watch was suggested with regular follow ups every month for the first year, and MRI every 6 months in the upcoming years. One year later, to our surprise, the brain MRI showed a significant reduction of cyst size. There was a normal physical and neurological development of the child, and no neurological

signs were seen during these observations. Periodic fundoscopic exams every six months were normal. The last neurological and psychological evaluation showed normal activity at the age of 8. The brain MRI showed a self-retracted cyst compared to the first MRI at birth.

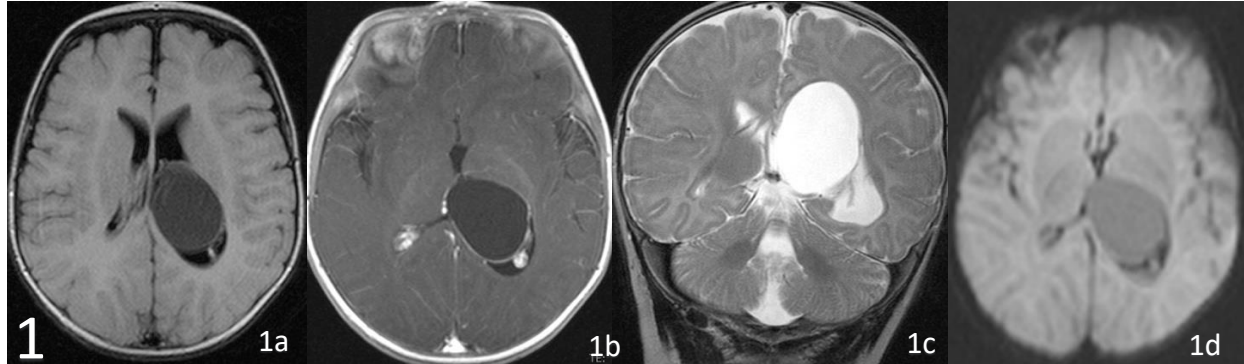


Fig 1: MRI of the brain performed 3 months after birth showing a left lateral Intraventricular neuroepithelial cyst hypointense on T1/FLAIR and hyperintense on T2 weighted sequence with no restriction on DWI.

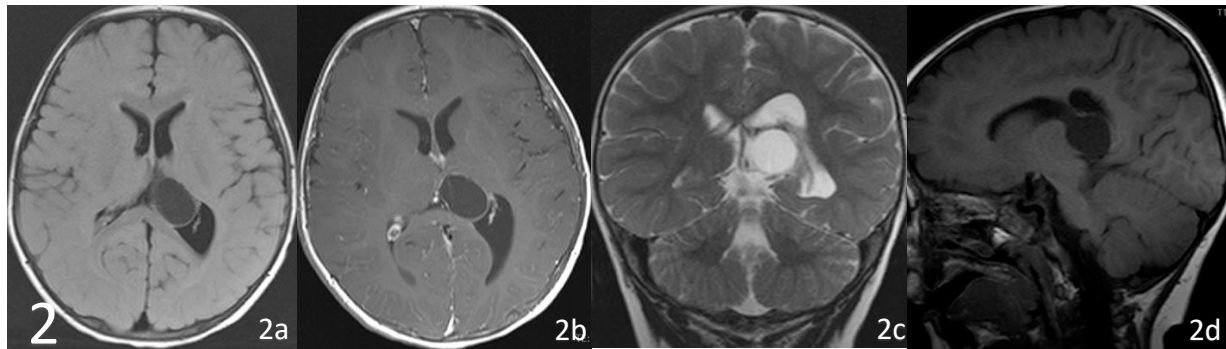


Fig 2: MRI performed one year later showing noted shrinking in size of the cyst

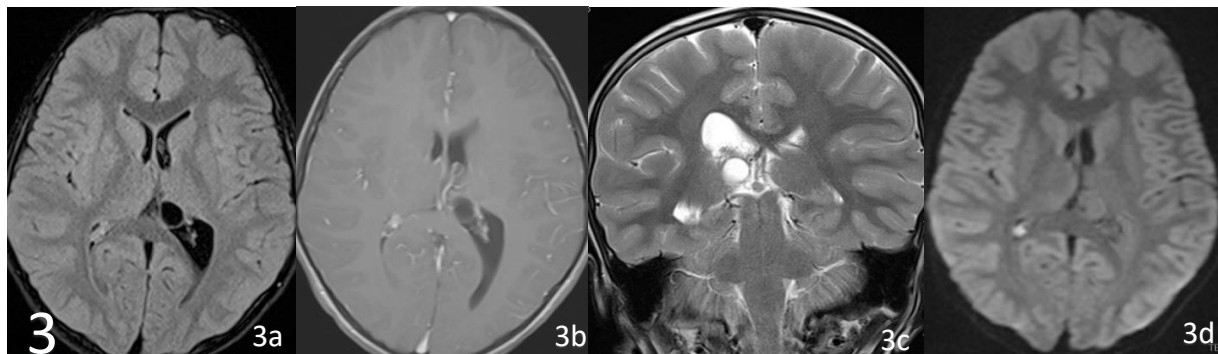


Fig 3: Last MRI 8 years later revealing further reduction in size of the cyst

Discussion: Neuroepithelial cysts are nonneoplastic cysts lined by cells morphologically resembling epithelium that develop in the central nervous system. Lateral and third ventricle is the location of choice of intracranial epithelial cysts [4]. Gliependymal cysts, ependymal cysts, and choroid plexus cysts are lesions subclassified in the group of neuroepithelial cysts because they all derive from the same embryological origin, the neuroepithelium [5]. They generally appear hypointense on T1 sequence in MRI and iso or hyperintense on T2 [6]. They rarely are asymptomatic, because they commonly present with symptoms related to hydrocephalus and elevated intracranial pressure (ICP) such as headache, nausea, vomiting, altered level of consciousness and even death [7]. Tange observed that neuroepithelial cysts in infants are generally associated with other central nervous system abnormalities such as microgyria or corpus callosum agenesis [8]. In contrast, our case did not have other abnormality. Symptomatic lateral ventricle cysts predominantly arise in the trigone, which is the most likely location to lead to obstruction between the body and the horn, causing episodic headache, which is a very common and non-specific symptom. Surgery is indicated in such cases [9]. The exact natural history of these cysts if left untreated is unknown. Recently [10], the symptomatic neuroepithelial cysts with progressive growing evidence were recommended for surgery [10]. A conservative approach is accepted for the asymptomatic cases, as we did in our case where no changes have been clinically observed over the years. Czervionke [6] reported the only case in which the cyst size diminished significantly over a 3-year period without treatment, similar to ours. For this reason, in an asymptomatic case, careful clinical and imaging follow-ups are needed before any decision.

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