

# Case lessons 24

## Neuroepithelial cyst of fourth ventricle and secondary Chiari I: a case report

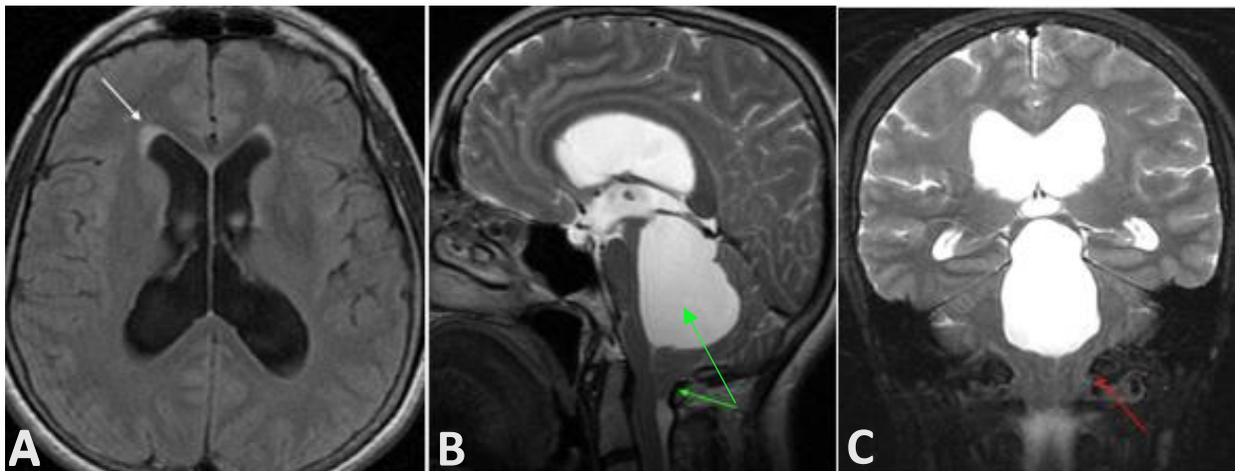
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**Introduction:** Neuroepithelial cysts (NECs) of the central nervous system are rare congenital benign growths with ectodermal origin [1]. Their etiopathogenesis still remains controversial, however several hypotheses have been proposed during the years [2]. Intracranial neuroepithelial cysts are generally found in the lateral ventricles, less frequently in the third ventricle, and among the sporadic cases in the posterior fossa, the fourth ventricular location is an extremely rare occurrence [1, 3]. Very few cases located in the fourth ventricle have been reported throughout the years [1,2, 4-7]. Their symptoms vary according to the localization and size, and are mostly due to mass effect. Signs of raised intracranial pressure (ICP) because of obstructive hydrocephalus are also observed, typically in fourth ventricle cysts, which can also be associated with vertigo and ataxia [8]. Acquired Chiari Malformation type I (CMI) is never reported.

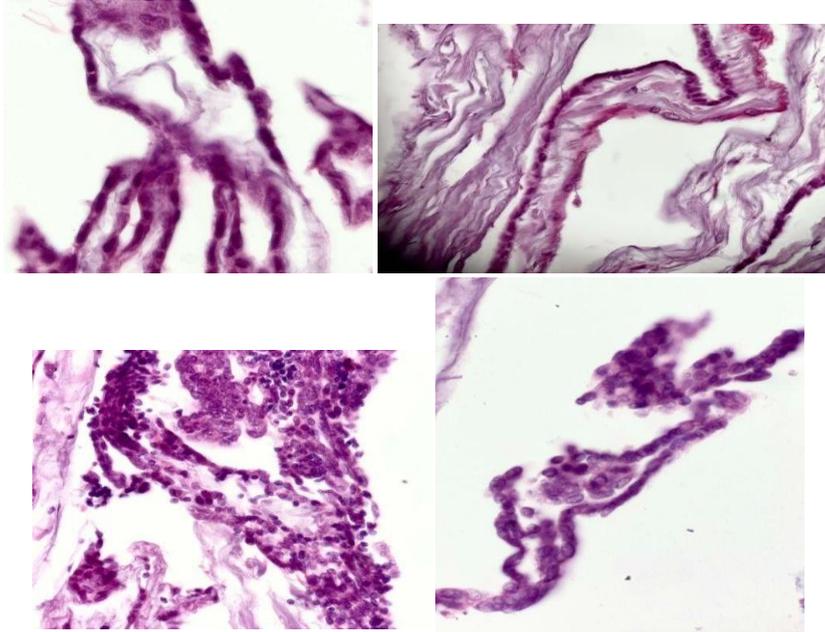
A case of a patient with epithelial cyst located in the fourth ventricle is presented with hydrocephalus and CMI.

**Case presentation:** a 56-year-old female with 5 years history of progressive gait ataxia, nausea, cephalgia, and persistent Valsava-induced headache located in occipital and frontal region that didn't respond to non-steroidal anti-inflammatory agents. She was a heavy smoker, and had before an untreated post-traumatic stress disorder. Neurological examination revealed left cerebellar syndrome with dysmetria, dysdiadochokinesia, scanned speech, wide-base gait, severe static and dynamic ataxia, hypotonic reflexes of inferior limbs with bilateral indifferent plantar reflex. Brain Magnetic Resonance Imaging (MRI) revealed a cystic lesion in fourth

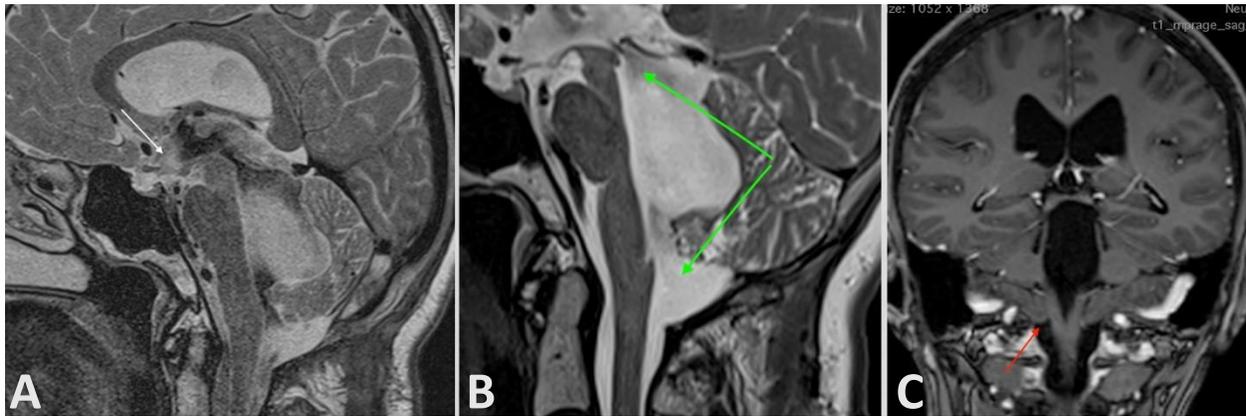
ventricle, tetraventricular obstructive hydrocephalus with acquired CMI. (Figure 1) Ocular fundus examination revealed on the right a stage I papilledema. The strategy was to perform Endoscopic third ventriculostomy (ETV) to release hydrocephalus, followed by suboccipital craniotomy for Chiari decompression and total removal of the fourth intraventricular cyst, all three procedures in one anesthesia. Surgery was realized in the sitting position and total removal of the cyst from the walls of the fourth ventricle was possible through telovelar approach (video on request Dr. Grada). Histological findings confirmed the diagnosis of a Neuroepithelial cyst lined by simple columnar epithelium (Dr. Latifaj, London) (Figure 2). Three months postoperative MRI showed reduction of the size of all ventricles and the level of both cerebellar tonsils was above Foramen Magnum (Figure 3). Headache and vertigo resolved, and there was an improvement of ataxia.



**Figure 1. Pre-operative images** (A) Periventricular hyperintensities due to obstructive hydrocephalus (white arrow) on T2/FLAIR- weighted axial MRI sequence, (B) cystic lesion which fills the fourth ventricle causing mass effect in the posterior fossa and tonsillar herniation through Foramen Magnum (green arrows) on T2-weighted sagittal MRI sequence, (C) T2-weighted coronal MRI sequence showing cerebellar tonsils below the foramen magnum (red arrow)



**Figure 2.** A cyst lined by simple columnar epithelium, neuroepithelial cyst



**Figure 3. Post-operative images** (A) T2-weighted sagittal MRI showing CSF fluid void after successful ETV (white arrow), (B) T2-weighted sagittal MRI showing clear passage of CSF into aqueduct following total cyst removal and free cisterna magna after tonsillar aspiration (green arrows). (C) Cerebellar tonsils above the level of Foramen Magnum (red arrow) on T1-weighted coronal MRI

**Discussion:** The majority of NECs never occur beyond the lateral and third ventricle [6]. Only a few reports have been described and to our knowledge this is the 7<sup>th</sup> case in the literature [1, 2, 4-7]. Typically incidental with no symptoms, they can show a variety of clinical presentations when symptomatic, from minor focal neurological symptoms to life-threatening ones,

determined by their location. Similar to our case [1, 2, 4-7], fourth ventricle NECs with ataxia and vertigo, presented with symptoms of increased intracranial pressure as well, with severe headache, blurred vision, nausea and vomiting, due to obstructive hydrocephalus. Headache is a common symptom, but is often neglected by patients and physicians [4, 9].

Diagnosis of NECs is based on histological findings [10]. Radiological features of NECs typically appear hypointense on T1-weighted images and iso or hyperintense on T2-weighted images, with no contrast enhancement [3].

In symptomatic cases, total resection of the cyst should be the goal of treatment in order to prevent recurrence. It is reported that if cyst walls are preserved during surgery, which might happen due to adjacent neurovascular adhesions or fragile locations, recurrence can occur from 2 months to 32 years after surgery [11]. Surgery is indicated for posterior fossa NECs, as in our case, because it allows direct inspection of the cyst wall, confirmation of the diagnosis by biopsy, fenestration and excision of the cyst [12]. Telovelar approach is suggested for fourth ventricle NECs [6]. However, GTR may not always be possible. Generally, in other locations when cyst wall removal was impossible or recurrence occurs, it was recommended the usage of an external drainage device, cysto-ventricular or cysto-subarachnoid shunt [12].

Fourth ventricular tumors fill and obstruct the ventricle cavity and the foraminal outlets resulting in tetra-ventricular hydrocephalus [13]. In hydrocephalus with acquired CMI, fluid resolving is important, as we did in our case. Not only it resolves elevated ICP, but it may resolve the acquired CMI as well [14]. ETV efficiency is confirmed for the treatment of hydrocephalus associated with secondary CMI [15]. In our patient, ETV, total removal of fourth ventricular cyst, and aspiration of both tonsils were performed.

**Conclusion:** Neuroepithelial cyst of the fourth ventricle is an extremely rare occurrence, often asymptomatic, but can complicate with important hydrocephalus and secondary CMI as it is reported for the first time. Attentive evaluation is needed before surgery. Total resection of the cyst is the accepted treatment when possible. ETV for hydrocephalus with cerebellar tonsils aspiration for CMI is recommended.

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