A dorsally located giant posterior fossa neurenteric cyst in a Chinese woman

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Abstract
Neurenteric cysts (NC) are rare, endodermal-derived intracranial lesions, commonly located anteriorly in the posterior cranial fossa. We describe a rare case of a giant posterior fossa NC (6.5 × 5.9 × 4.3 cm) located dorsal to the brain stem in a Chinese woman with a 1 week history of cerebellar symptoms. To our knowledge, this is the largest documented cyst of this type and the third dorsally located NC in the posterior fossa.

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1. Introduction
Neurenteric cysts (NC) are endodermally-derived lesions in the central nervous system (CNS), most commonly found in the spinal canal [1]. Intracranial NC are rare and are typically located in the posterior fossa, anterior to the brainstem [1,2]. Here we present an unusual case of a giant NC in the dorsal posterior fossa of a 39-year-old Chinese woman.

2. Case report
2.1. History and examination
A 39-year-old Chinese woman presented with a 1 month history of intermittent vertigo and occipital headaches, and a 1 week history of poor coordination in the lower extremities and frequent falls. Upon neurological examination, there were no abnormal findings except for mild deep tendon hyperreflexia, signs of cerebellar dysfunction including gait disturbance, and positive Romberg’s sign.

2.2. Neuroimaging
CT scan showed a hyperdense (95–105 Hounsfield units) homogeneous rounded lesion with smooth borders in the dorsal posterior cranial fossa. The fourth ventricle was compressed and obstructive hydrocephalus was present. MRI showed a well-demarcated, extra-axial cyst measuring 6.5 × 5.9 × 4.3 cm in size. The lesion had heterogeneous signal intensity on T1-weighted imaging, low signal intensity on T2-weighted imaging, and showed no enhancement with gadolinium administration (Fig. 1).

2.3. Operation
A suboccipital craniectomy was performed. The cyst was noted to be extra-axial and thin-walled, with brown, machine oil-like contents (Fig. 2). After a complete excision, the patient made an uneventful post-operative recovery and was subsequently discharged home.

2.4. Pathological findings
Light microscopic examination revealed a cyst wall lined by a pseudostratified, ciliated, columnar to cuboidal, mucin-producing cell-poor epithelium, which was surrounded by a thin layer of connective tissue stroma. These features are consistent with a NC (Fig. 2).

3. Discussion
NC are developmental cysts of endodermal origin, which represent approximately 16% of cysts of the CNS [3]. Most NC occur in the spinal canal and are associated with bony anomalies in 50% of patients [4]. In contrast, intracranial NC are rare and most commonly located in the posterior fossa. They are usually midline, anterior to the brain stem, or in the cerebellopontine angle [1]. They have also been found in the fourth ventricle [1,4]. To our knowledge only two other NC have been reported to be located dorsally in the posterior fossa [5]. Unlike spinal neurenteric cysts, NC in the posterior fossa are almost never associated with a bony anomaly of the clivus or skull base [4]. These extra-axial posterior fossa cysts are usually small, ranging in size from 1 × 0.5 to 3 × 3 cm [4]. The cyst we report here is approximately two times larger, and is the largest posterior fossa NC documented to our knowledge. The average age of patients diagnosed with intracranial NC is 34 years [1], which is similar to our patient’s age.

On gross examination, yellow, white, gray, and red cyst walls have all been reported [6]. The fluid of the cyst, however, is often clear or yellowish. A brown oily substance, similar to that seen in our patient, has also been reported [3,6]. Xanthomatous changes also can occur [7]. NC have two major histologic patterns [4]. One type of NC is composed of pseudostratified, ciliated, columnar to cuboidal, mucin-producing cell-poor epithelium, and the other of simple, nonciliated, mucin-producing cell-rich epithelium. Although the precise embryologic development of intracranial NC is not clear [1,3], these two histologic patterns correspond, respectively, to the epithelia of respiratory and gastrointestinal origin.

NC typically appear to be hypodense on CT scans, but are occasionally hyperdense [1]. Similarly, they may be hypo- to hyperintense on T1- and T2-weighted MRI sequences, or may appear to have mixed or compartmentalized densities [4,8].
These discrepancies are thought to result from variations in the protein and lipid content of the cyst [1]. Complete surgical excision has thus far been the only effective treatment option for symptomatic intracranial NC. Patients must be followed with subsequent MRI, as recurrence is possible even with apparent gross total resection [9].

Conflicts of Interest/Disclosures

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

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References

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