Neurenteric cyst of the 4th ventricle. Case report and short review of the literature

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Abstract

Intracranial neurenteric cysts are uncommon and occur usually in the posterior fossa. We report one case of a neurenteric cyst that was situated in the 4th ventricle. Total surgical removal was performed. One hundred cases having been published in the English language literature. We report on the imaging features of the tumor on several modalities as well as its histopathology. We further review the literature regarding this rare benign tumor entity.

Introduction

Neurenteric cysts (NCs) are rare congenital lesions that belong to the group of notochordodysraphisms. They are known to occur in the posterior mediastinum and abdomen. Occurrence adjacent to the CNS is rare but is predominantly in the cervical and thoracic spinal canal, located intradurally and extramedullarily. They compose 0.3–0.5% of all spinal tumours [1] and 16% of spinal cysts [2]. In 12–92% of cases, NCs are associated with other dysraphic malformations [3]: spina bifida, diastematomyelia, diplomyelia, dermal sinus, etc [1, 4-6]. Puusepp et al. first reported an intraspinal endodermal cyst in 1934, calling it ‘intestinoma’. Since then, nomenclature has varied widely, with the term ‘neurenteric cyst’ being coined by Holcomb and Matson in 1954 [7, 8]. NCs have had several names, including enterogenous cyst, enteric cyst, endodermal cyst, gastroenterogenous cyst, gastrocytoma, intestinoma, and archenteric cyst [9].

Intracranial NCs are uncommon and have been the subject of isolated case reports. The majority of these cysts are located in the posterior fossa [10-28] such as the preoptic cistern [29-31], cerebellopontine angle [28,29,32-37] and craniovertebral junction [38-58]. Supratentorial NCs are exceptionally rare, only 26 have been reported [2, 28, 59-73]. Also, rare condition is neurenteric cyst in the subarachnoid or intraventricular space of the central nervous system. To our knowledge, at least ten case reports of an intra- fourth ventricular neurenteric cyst have been reported in the literature (Table 1).

We report a case of an intra- fourth ventricular neurenteric cyst and review the literature for imaging characteristics and also the radiology literature is reviewed.

Case report

A 37 year-old woman presented with headache and vertigo. Her symptoms had become worse in the last 6 months. Physical and neurologic examinations revealed no abnormalities.

A contrast CT scan demonstrated a large well circumscribed non-enhancing hypodense midline mass lesion in the posterior fossa (Figure 1). The lesion extended from the midline to the adjacent brain parenchyma. The MRI demonstrated a well-circumscribed lobulated lesion in the posterior fossa with superior displacement of the vermis and fourth ventricle, significant compression of the brain stem was seen as well. On T1-weighted images the cyst was slightly hypointense and on T2-weighted images was homogeneously hyperintense. After contrast administration no enhancement was seen (Figure 2a, b, c).

The cyst was removed after craniotomy. The patient was operated in 3/4 prone position through a midline skin incision from the inion until 5/4 prone position through a midline skin incision from the inion until 5th cervical spine. A suboccipital craniectomy was performed that was caudally extended to a laminectomy of the 1st cervical spine. The 5th cervical spine. A suboccipital craniectomy was performed that was caudally extended to a laminectomy of the 1st cervical spine.

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1. Parkinson et al. (1952) [10] M 28Y 4th ventricle + _
2. Shuangshoti et al. (1966) [11] M 48Y 4th ventricle + _
3. Shuangshoti et al. M 60Y 4th ventricle + _
4. Shuangshoti et al. M 50Y 4th ventricle + _
5. Hoening et al. (1967) [12] M 68Y 4th ventricle + _
6. Afshar et al. (1981) [15] M 48Y 4th ventricle + _
7. Wells et al. (1986) [18] F 40Y Multiple, 4th (1 post.cr.fossa) + _
8. Okabe et al. (1995) [21] M 53Y 4th ventricle + +
9. Preece et al. (2006) [28] F 57Y 4th ventricle _ +
10. Yasuda et al. (2008) [13] e F 46Y 4th ventricle + -
11. Potsi et al. (2009) F 37Y 4th ventricle + +

Table 1. Summary of reported intra-fourth ventricular neurenteric cyst (-) no exist, (+) exist

| Sex | Age | Location | CT | MRI |
|-----|-----|----------|----|-----|
| M   | 28Y | 4th ventricle | +  | _  |
| M   | 48Y | 4th ventricle  | +  | _  |
| M   | 60Y | 4th ventricle  | +  | _  |
| M   | 50Y | 4th ventricle  | +  | _  |
| M   | 68Y | 4th ventricle  | +  | _  |
| M   | 48Y | 4th ventricle  | +  | _  |
| F   | 40Y | Multiple, 4th (1 post.cr.fossa) | + | _  |
| M   | 53Y | 4th ventricle  | +  | +  |
| F   | 57Y | 4th ventricle  | _  | +  |
| F   | 46Y | 4th ventricle  | +  | -  |
| F   | 37Y | 4th ventricle  | +  | +  |
dura was opened in a “U” shape manner and the content of the cyst was aspirated (Figure 3). Several milliliters of viscous fluid were removed and after the cyst was gradually collapsed, the margins were coagulated and detached from the surrounding tissue using bipolar coagulation and small strippers. Only the cervical portion of the left glossopharyngeal nerve was closely attached to the capsule and was damaged during the excision. Finally, after cutting all the surrounding adhesions, the collapsed cyst was excised en bloc. The patient was transferred in an intensive care unit and woke up the same day without any neurology. Damage to the left glossopharyngeal nerve was incomplete and didn’t compromise the patient who had a full and uneventful recovery. The patient was discharged one week later.

The histopathologic examination revealed endodermal cyst composed of pseudostratified, ciliated, columnar to cuboidal, mucin-producing cell poor epithelium (Figure 4).

**Discussion**

Neurenteric cysts of the central nervous system have been described in regions from the orbital apex to the sacrum. They are usually found in the spinal canal at the lower cervical and upper thoracic segments. Neurenteric cysts may be intraspinal, extraspinal or both, although most often they are intradural and extramedullary in location.

In contrast, intracranial NCs are extremely rare, with less than 100 cases being reported. They occur in all age groups. It would seem that actual prevalence of NCs is likely to be independent of gender (Table 2). With rare exceptions, intracranial NCs have not been associated with skeletal anomalies. Exact mechanism of formation of enterogenous cysts is not well known and many hypotheses have been suggested. Neurenteric cysts may occur because of an incomplete separation of the primitive gut from the neural groove, due to the persistence of the Giebler-Kuhn channel which connects endoderm and ectoderm in the third week of embryonic life.

The patients may present neurologically with signs of spinal cord compression and meningitis, but the clinical symptoms associated with neurenteric cysts depend on the site of the lesion.

The cysts consist of mucin-producing, nonciliated epithelium that is simple or pseudostratified and either cuboidal or columnar. The cyst can be ciliated, or it can have a mixture of gastrointestinal, pancreatic, and/or squamous epithelium. Some authors described a possible relation to the respiratory tract.

Almost half of intracranial cysts reported in the literature were located within the posterior fossa (47%) [10-21,23-36,45,49,62]. They are typically midline, anterior to the brain stem, or in the cerebellopontine angle [28,29,32,34-36]. Cysts have also been described in the fourth ventricle which are only 10 cases have been reported so far [10-13,15,21,28]. The case presented here bring the total to 11. Other locations include the parasellar/suprasellar region [60,63] septum pellucidum [65], third ventricle [63], orbital apex [59], the optic nerve [58] and extraxial anterior cranial fossa [71].

They are rare at the craniovertebral junction, with only 28 reported cases [1,22,8,37-44,46-48,50-57]. Supratentorial NCs are rare, and 25%
of the cysts in those cases reported to date have been located within the brain parenchyma [2,28,58-73].

The classic NC, according to the literature, is an ovoid, lobulated mass in front of the medulla. The majority of them are small in size: less than 2cm.

Forty-nine of the one hundred patients had CT scans. Although CT is superior in demonstrating calcification NCs are extremely rarely calcified with only one case being described [69].

Magnetic resonance imaging is the imaging investigation of choice with 73 of 100 patients having MR images. Most NCs appear ovoid or lobulated and appear sharply demarcated. Signal intensity on MR is variable, depending on cyst contents and reflecting the amount of protein content. The protein content of cystic fluid shortens both T1 and T2 relaxation times. Most are proteinaceous with a T1 signal intensity that is isointense to slightly hyperintense compared with CSF and typically very hyperintense on T2, unless inspissated.

Thirty-four of the seventy-three patients (46,5%) are hyperintense, seventeen (23,2%) are hypointense and twenty (27,3%) are isointense and typically very hyperintense on T2, unless inspissated.

Contrast-enhanced scans are available in 54 of 73 cases (73,9%). Typically, no enhancement of the cyst occurs following contrast administration. Rim enhancement is seen in only 15 out of 73 cases at the site where the cyst adhered to the brain [21,28,31,35,47,53-56,65,67].

The differential diagnosis of intracranial NCs include epidermoid cyst, arachnoid cyst, and other endodermal cysts (Rathke and colloid). The rare “white” epidermoid is most similar to the NC in that it is hyperintense on T1 and can be difficult to distinguish if midline. Diffusion weighted imaging is useful to differentiate between endodermal cysts and epidermoids. Epidermoids lesions show moderate to striking diffusion restriction. Arachnoid cysts follow CSF on all sequences. Other endodermal-derived cysts, such as Rathke and colloid, can be differentiated from NCs based on location. A Rathke cleft cyst is sellar or suprasellar, while a neurenteric cyst is usually in the posterior fossa, and a colloid cyst typically is in the region of the foramen of Monro along the anterior superior wall of the third ventricle. Differentials in children should probably include juvenile and pilocytic astrocytoma. Also, in patients of all ages should probably include parasitic cysts.

In conclusion we suggested that the neurenteric cyst should also be included in differential diagnosis of the intracranial cystic mass bearing in mind that in rare cases NCs may present rim enhancement and more rarely calcification.

Conflict of interest
No

Presentation at a conference
No

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