Spinal Neurenteric Cysts

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History and Nomenclature

Neurenteric cysts (NECs) are rare benign cystic lesions of the central nervous system (CNS) that are derived from the endoderm. These lesions were first described by Puusepp in 1934 as "intestinomas" [1]. They have been given various alternative names in the literature such as enterogenous cysts, enteric cysts, endodermal cysts, bronchogenic cysts, gastroenterogenous cysts, gastrocytomas, and archenteric cysts [2–6]. Although the World Health Organization (WHO) prefers the term "enterogenous" in the revised classification of CNS tumors, the term "neurenteric cyst" is the most widely accepted in the neuropathology literature [5–7].

Epidemiology and Location

These cystic lesions occur among all age groups with no gender predilection.

A case of dorsal neurenteric cyst has been reported in a 2-year-old patient who also had an infection within the cyst, with evidence of paraspinal extension [8]. NECs can occur anywhere along the neuraxis, but spinal NECs are about three times more frequent than intracranial ones [2, 9]. They represent about 0.7–0.3% of all spinal tumors [10]. They are usually found ventral to the cord. Spinal NECs are most often found at the lower cervical or upper thoracic level [11], although a few authors have reported rare occurrences at the craniovertebral junction and the lumbar spine [12, 13]. Most intraspinal NECs are located in the intradural extramedullary (IDEM)

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compartment of the cervicothoracic junction and are often associated with bony, soft tissue, and visceral abnormalities. They occur in the intramedullary compartment in only <5% of cases [12–14]. About 50% of cases are associated with vertebral body anomalies such as hemivertebrae and kyphoscoliosis [10, 15, 16]. Associations with bony defects such as spina bifida, split cord malformation, and Klippel-Feil syndrome are not uncommon [10].

Intracranial NECs are rare (about 0.01–0.35% of all CNS lesions) and only about 100 cases have been reported in the literature to date [2, 17, 18]. The posterior fossa is the most common intracranial location, accounting for approximately 70–90% of intracranial NECs [18, 19]. They are typically found in the midline, often anterior to the brain stem (23.1–51%), the cerebellopontine angle (17–51%), or inside the ventricles [20]. Only a few cases of NECs have been reported in rare supratentorial locations such as the suprasellar [21, 22] or the parasellar [18, 23] regions, along the optic [24] or the oculomotor [25] nerve, in the superior orbital fissure, [26] or as intraparenchymal- or dural-based lesions [27, 28]. In contrast to their spinal counterparts, intracranial NECs are seldom associated with bony anomalies, only one case report showing erosion of the petrous temporal bone adjacent to the cyst [18, 20, 29].

Various Embryological Explanations for the Development of NECs

The exact etiology of NECs remains unknown [3, 30], but several hypotheses have been proposed [3, 9, 18, 31–33]. These cystic lesions are thought to arise from faulty development of the neurenteric canal. During early embryogenesis, there is transient communication between the primitive neuroectoderm and endoderm. Around the 3rd week of gestation, if the notochord and foregut fail to separate during the process of escalation, primitive endodermal cells are incorporated into the notochord [3, 4, 29]. These displaced nests of alimentary tissue ultimately generate the components of the cyst. Smith proposed a classification system depending upon the extent of persistence of the neurenteric canal. The spectrum of variability in the composition of an NEC ranges from the most innocuous congenital dorsal enteric sinus to a dorsal enteric fistula, where the entire neurenteric canal remains patent [34]. The clivus forms the cranial margin of the endoderm in an embryo; this can account for the origin of spinal NECs caudal to the clivus, but the origin of supratentorial NECs remains unexplained [29].

Some authors believe that supratentorial NECs arise from the endodermally derived Seesel pouch, a midline diverticulum located behind the oropharyngeal membrane. This hypothesis is supported by the fact that NECs have similar immunohistochemical staining properties to those of Rathke's cleft cyst and colloid cysts, positive for epithelial membrane antigen (EMA), carcinoembryonic antigen

(CEA), and cytokeratin and negative for glial fibrillary acidic protein (GFAP). However, this still fails to explain the development of off-midline cysts [17, 18, 21, 35, 36].

Clinical Presentation

Spinal NECs occur among a wide range of age groups from the neonatal period to the eighth decade. There is a bimodal age distribution with a small peak found in the first decade and a larger second peak in the third to fourth decade with no gender predilection [37].

The most common overall location of NECs is the posterior mediastinum [22, 37, 38]. They can connect the CNS to the mediastinum or abdominal viscera [22, 38]. The spinal canal is the second most common site. Spinal NECs are usually located ventral to the cord; dorsally placed ones are very rare [8]. About 50% of them are associated with such anomalies as dysraphism, hemivertebrae, and VACTERL anomalies (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities). NECs constitute less than 1% of all spinal tumors [11, 37]. Sometimes they occur near the lower clivus, in the medulla oblongata, at the parietal convexity, and within the optic nerve; they can even be extradural [24, 37, 39–42].

NECs can present as incidental space-occupying lesions on magnetic resonance imaging (MRI) (Figs. 10.1, 10.2, 10.3, 10.4, 10.5, 10.6, and 10.7). When symptomatic, the onset of symptoms can range from a week to 10 years prior to diagnosis [38]. They commonly present with nonspecific symptoms predominantly depending upon the anatomical location of the lesion, such as focal neurological deficits, seizures, cranial nerve palsies, gait disturbance, ataxia, myelopathy, radicular pain, and incontinence. They can manifest as memory disturbances, obstructive hydrocephalus with features of raised intracranial pressure (ICP) [43], and rarely with atypical presentations such as an acute onset of symptoms, recurrent episodes of myelo-radiculopathy, or aseptic meningitis resulting from microleakage of cyst contents [13]. The most common presenting symptom is headache. These symptoms can result from chronic inflammation or irritation of the surrounding structures or to the local mass effect [22, 44]. They are usually benign lesions with a slow growth potential; however, rapid expansion can result from hemorrhage, inflammation, or increased secretion of fluid content within the cyst [37]. Sharma et al. observed acute-onset, rapidly progressive quadriparesis with respiratory failure in a ventrally placed foramen magnum NEC, which required emergency life-saving surgical decompression [12]. Malignant transformation of a NEC is very rare, with only six cases reported in the literature [45–47]. NECs with rare presentations and with nonspecific symptoms pose a diagnostic challenge.

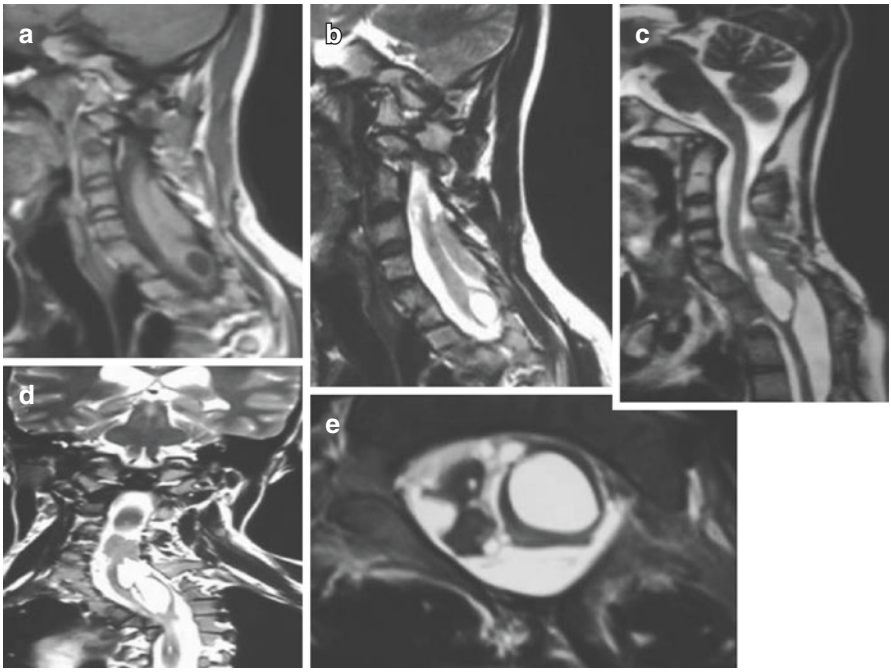

Fig. 10.1 Typical radiological findings in a spinal neurenteric cyst. A well-defined intramedullary cystic lesion can be seen extending from C6 to D3. The cystic lesion is (**a**) hypointense on T1-weighted image; (**b**) hyperintense on T2-weighted image. (**c**, **d**) It causes fusiform enlargement of the spinal cord. The C5 and C6 hemivertebrae exhibit scoliosis toward the right side. (**e**) A large intramedullary cyst is visible in the axial section

Histopathological Classification

NECs usually appear as thin-walled transparent cysts containing a gelatinous fluid, yellow or milky-white in color on gross inspection. They have been described as having a variable appearance under light microscopy. There are two predominant histological variants: [3, 5] (1) pseudostratified, ciliated, columnar-to-cuboidal epithelium with scant mucin-producing cells (17%); and, (2) simple, nonciliated epithelium with abundant mucin-producing cells (50%). The only consistent radiological finding with a pathological correlate is T2 hypointensity associated with elevated protein concentration in the cyst fluid [29]. The cyst fluid is often clear, yellowish, or brown; it is oily and contains histiocytes, cholesterol crystals, and keratin debris. Various other pathological features such as foamy histiocytes with chronic inflammatory cells [48], amyloid deposits in the cyst wall [37], and a xanthogranulomatous change [49] have been reported. Occasionally, there is squamous metaplasia.

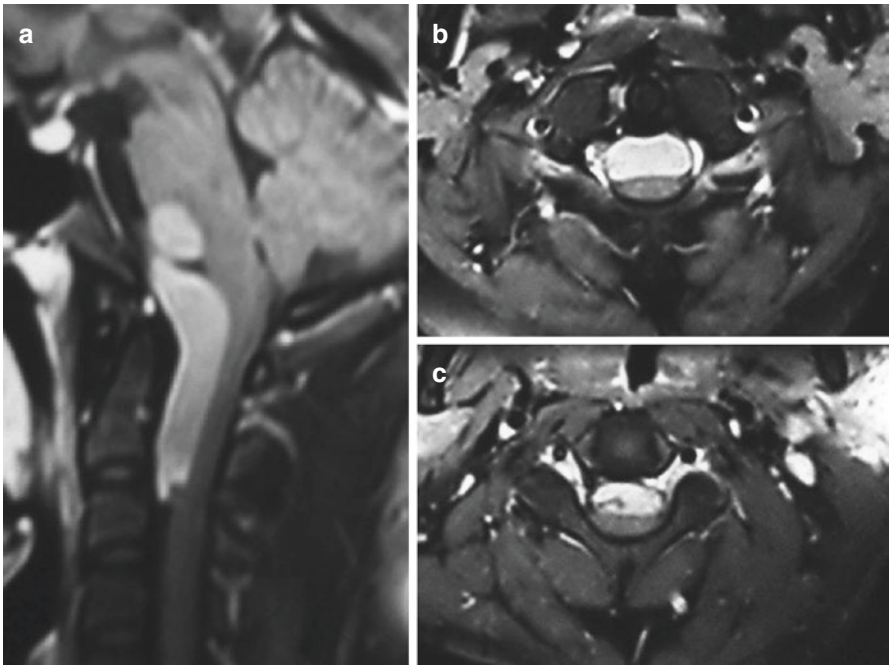

Fig. 10.2 (**a**) Contrast T1 sagittal MR image of the craniovertebral junction (CVJ) showing an anteriorly placed neurenteric cyst with an indentation caused by the traversing vertebral artery. (**b**, **c**) Contrast-enhanced axial T1 MR images showing the lesion situated anterior to the cervical cord. (From Shukla et al. [14]; with permission)

Three histological types of spinal NEC can be distinguished according to the Wilkins and Odom classification (Table 10.1) [50]. Type 1 cysts resemble respiratory or gastrointestinal epithelium and are covered with a single or pseudostratified layer of ciliated or nonciliated cuboidal or columnar epithelium with a basement membrane overlying the fibro-connective tissue. Type 2 cysts are richer in connective tissue and can include smooth muscles, glandular and lymphoid tissues, and rarely nerve ganglia, in addition with all the features of type 1 cysts. Type 3 cysts are similar to type 2 with the addition of ependymal or glial elements. Anteriorly placed intracranial cysts are usually type 1, while posteriorly placed ones are often type 2 or type 3 and are more likely to be associated with congenital anomalies (Fig. 10.8) [16].

NECs are typically benign; however, de novo malignancy, and even malignant transformation to mucinous low-grade adenocarcinoma [51], invasive mucinous papillary cystadenocarcinoma [45], or a well-differentiated papillary adenocarcinoma [52], can occur. A supratentorial NEC can coexist with an intraparenchymal subependymoma [53].

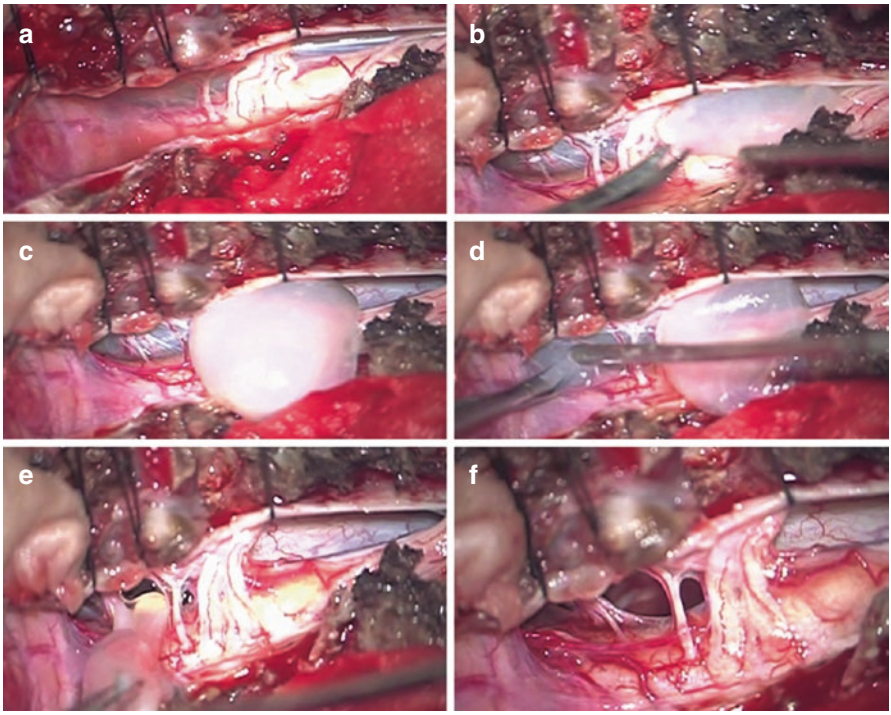

Fig. 10.3 Intraoperative images showing the lesion undergoing a right-sided far lateral approach without vertebral artery mobilization. (**a**) The medulla and upper cervical cord are compressed by an anteriorly placed cyst covered with arachnoid membrane. The cyst lies both superior and inferior to the origin of the lower cranial nerves. (**b**, **c**) The arachnoid anterior to the upper cervical cord is opened to reveal the inferior part of the neurenteric cyst. (**d**) Opening the arachnoid above the lower cranial nerves reveals the upper part of the neurenteric cyst. (**e**) The lower part of the cyst is excised. The capsule of the upper part is delivered, and the fibrotic band adherent to the anterior cord is visible; (**f**) the entire cyst has been excised, and the vertebral artery can be seen in the depth anterior to the cervicomedullary junction. (From Shukla et al. [14]; with permission)

Use of Immunohistochemistry (IHC) in Diagnosis

Immunohistochemistry is important in establishing the diagnosis, particularly if the radiological findings are nonspecific. The epithelia of NECs stain positive for anti-EMA antibodies, anti-cytokeratin monoclonal antibodies, and anti-carcinoembryonic antigen antibodies [8], suggesting an endodermal origin. They stain negative for neuroectodermal markers such as GFAP, S100, neuron-specific enolase, and vimentin [22]. There is no NEC-specific marker. Periodic acid Schiff (PAS), mucicarmine, and Alcian blue stains can also be used to demonstrate secretory granules in the goblet cells, again signifying an endodermal origin. Some authors have reported carbohydrate antigen (CA) 19-9 positivity in benign cysts as well as in those that have undergone a malignant transformation. The titers of CA19-9 in the cerebrospinal fluid (CSF) increase in recurrent cysts, so this could prove to be an important biochemical surveillance marker [54, 55].

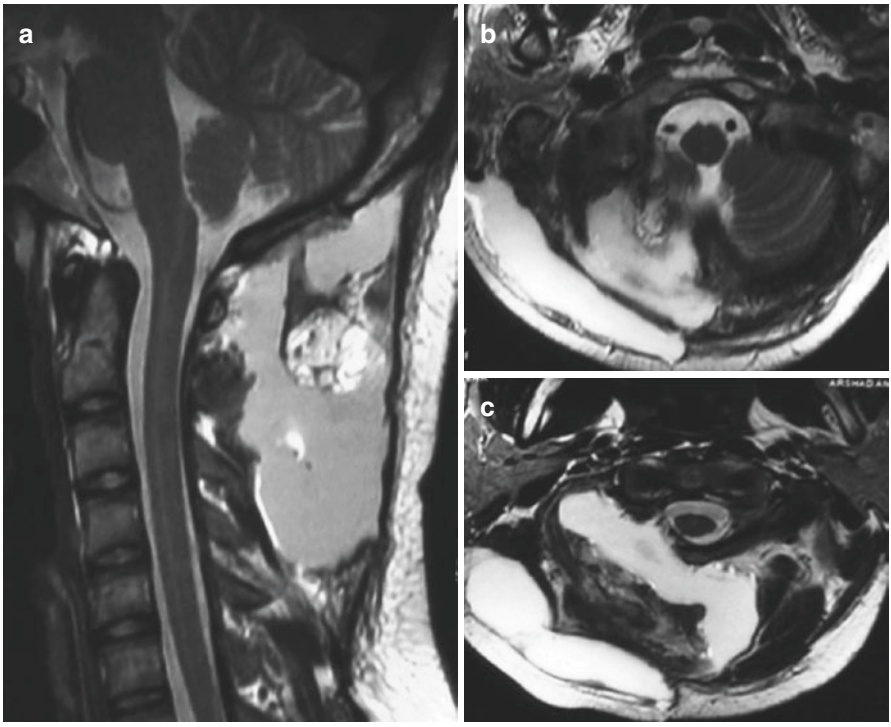

Fig. 10.4 (**a**) Postoperative T2 sagittal and (**b**, **c**) axial MR images showing total cyst excision by the far lateral approach. (From Shukla et al. [14]; with permission)

Diagnostic Imaging

Detailed radiological examination of the vertebral anatomy is essential in the treatment plan. The classic NEC, according to the literature, is usually less than 2 cm in size, a well-demarcated, lobulated cystic lesion with smooth borders and without contrast enhancement, found in extra-axial, intraparenchymal [35], or intraventricular locations [35, 56–58]. NECs most commonly appear as hypodense non-enhancing lesions on computed tomographic (CT) imaging. However, they can appear hyperdense or sometimes isodense to the surrounding brain parenchyma, posing a diagnostic dilemma. Some authors have reported that these cysts show peripheral wall enhancement [18, 29].

MRI is the radiological imaging method of choice. The signal intensity on MRI varies with the protein content of the cyst. It is typically T1 isointense to slightly hyperintense and hyperintense on T2-weighted imaging (WI) (Fig. 10.1). A case series has reported T1 hyperintensity (hyperintense to CSF) and T2 hyperintensity in 88.9% of cases. T2 hypointensity is described in the other 11.1%. High protein and xanthogranulomatous change can result in very high signal intensity on T1WI. T2 hypointensity in NECs has been reported in a few cases with squamous metaplasia (Figs. 10.1, 10.2, 10.3, 10.4, 10.5, 10.6, and 10.7) [29]. The cysts appear as

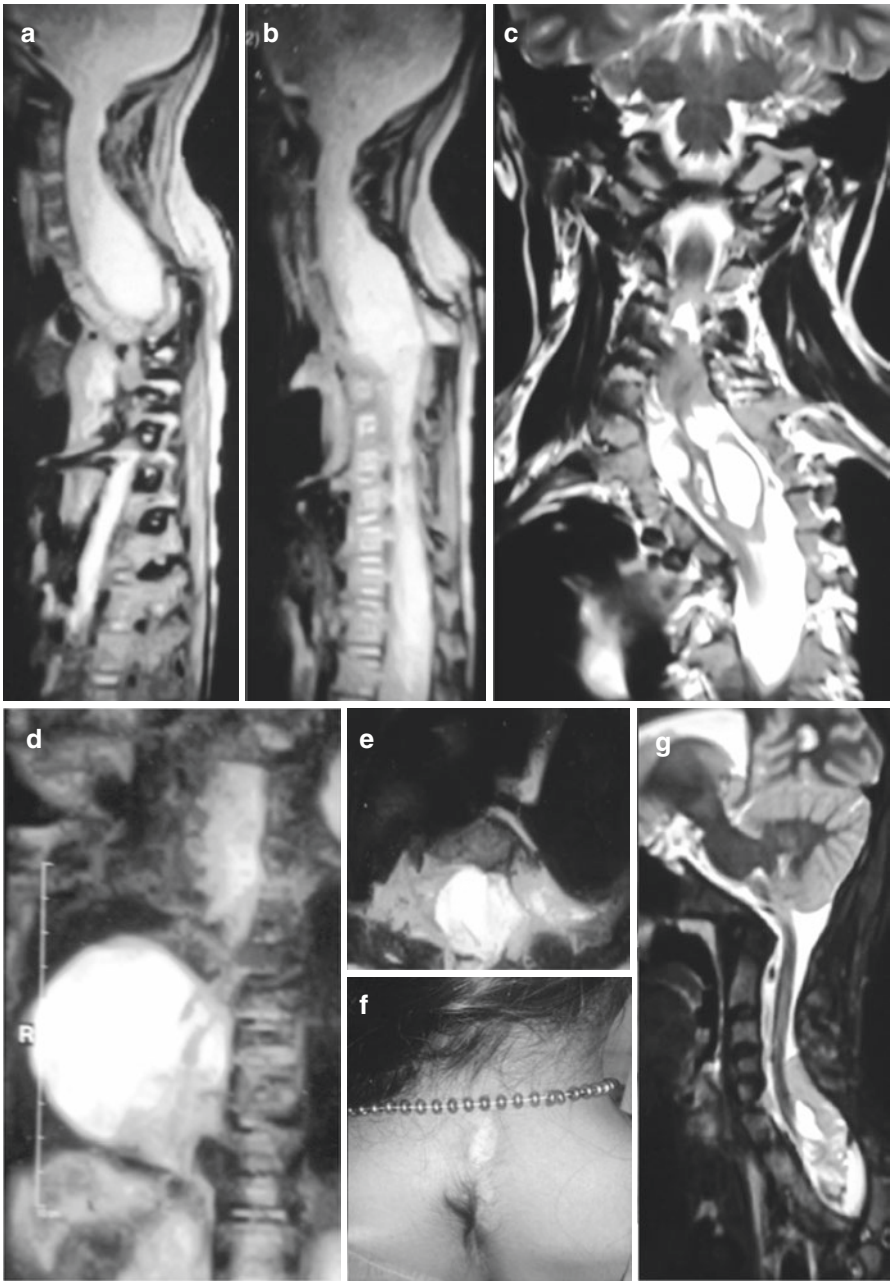

Fig. 10.5 An intradural extramedullary neurenteric cyst at the cervicothoracic region. (**a**, **b**) T2 sagittal and (**c**) T2 coronal image showing the cyst with cervicothoracic scoliosis with split cord malformation and abnormal dilatation of the spinal canal at the same level. (**d**) T2 coronal MR image of the thoracic spine showing a right-sided posterior mediastinal cyst. (**e**) T2 axial MR image showing asymmetrical dilatation of the cervical canal and an anteriorly placed neurenteric cyst. (**f**) Hypertrichosis with spina bifida. (**g**) T2 sagittal postoperative image shows excision of the cyst. There is persistent syringomyelia. (From Shukla et al. [14]; with permission)

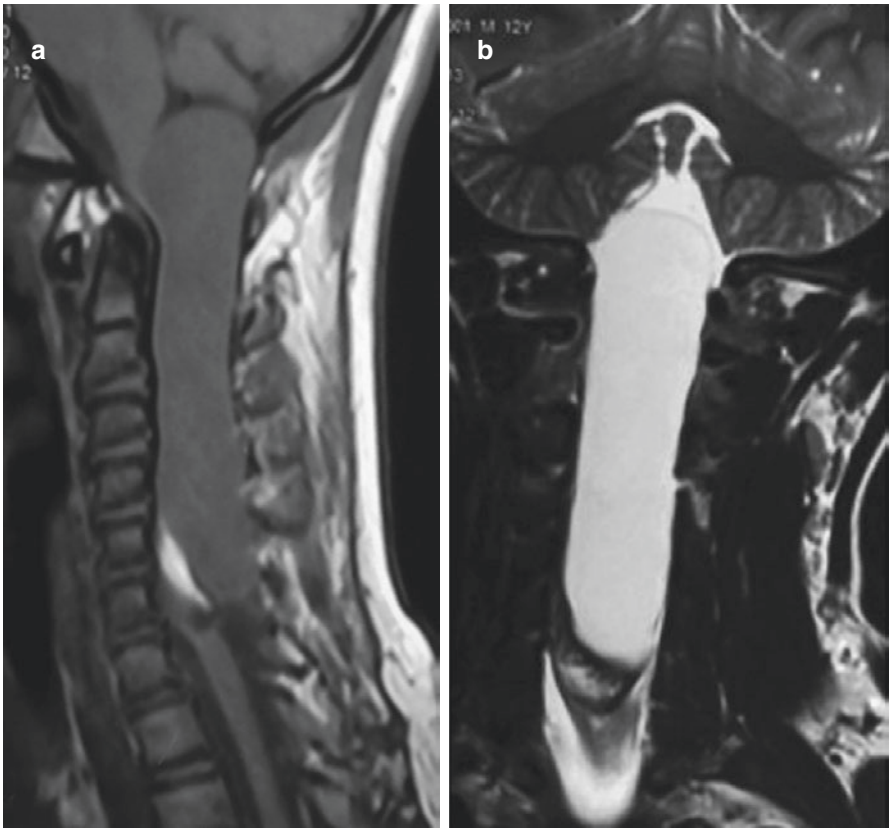

Fig. 10.6 (**a**) T1 sagittal and (**b**) T2 coronal MR images showing a long segment neurenteric cyst extending from the cervicomedullary junction to the cervicothoracic junction posterior to the cord with an abnormally shaped vertebral body and an anteroinferiorly placed fibrolipoma, causing adhesion to the spinal cord. (From Shukla et al. [14]; with permission)

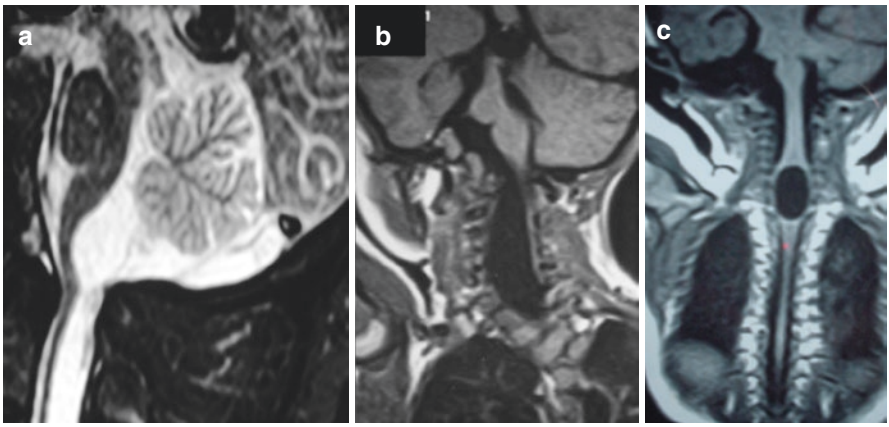

Fig. 10.7 (**a**) T2 sagittal MR image of a posteriorly situated cyst compressing the medulla and cervical cord. (**b**) T1 coronal image showing a patient with cervical scoliosis and a cervicothoracic hemivertebra with an anterolateral cyst causing cervical cord thinning. (**c**) An intramedullary neurenteric cyst. (From Shukla et al. [14]; with permission)

Table 10.1 Wilkins and Odom histopathological classification of spinal neurenteric cysts

Type	Characteristic features
Type A	Single layered pseudostratified ciliated columnar epithelium with basement membrane similar to respiratory or gastrointestinal epithelium
Type B	In addition to features of Type A – complex invaginations and organized glands producing mucinous or serous fluid, nerve ganglion, mesenchymal elements like muscle tissue, fat, cartilage, or bone
Type C	In addition to features of Type B – glial or ependymal tissue is also found

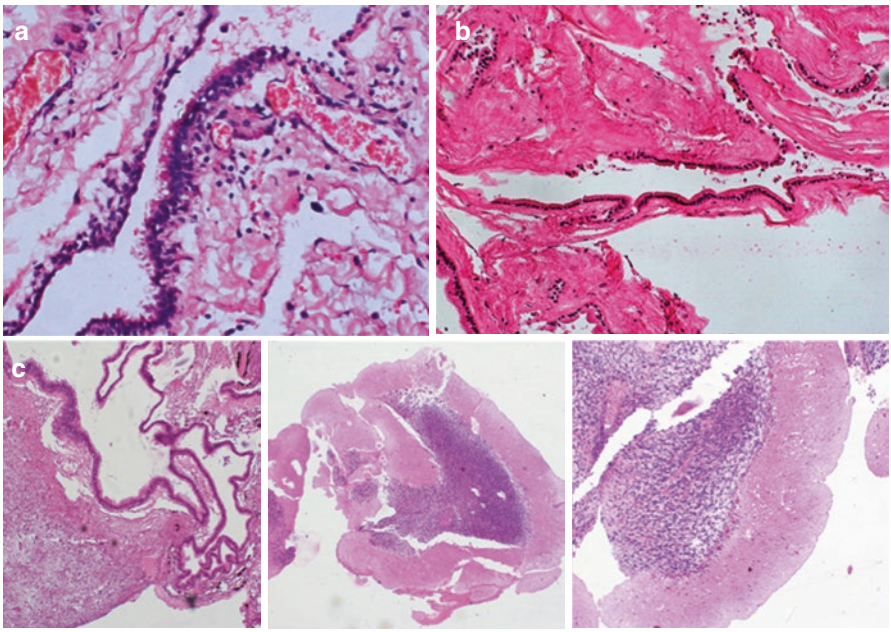

Fig. 10.8 Micrograph of a neurenteric cyst. (**a**) A cystic structure with the wall comprising fibrocollagenous tissue and lined with pseudostratified ciliated columnar epithelium and mucinous cells. The subepithelium shows congested vessels and mild mononuclear inflammatory cell infiltrate (hematoxylin and eosin X 400). (**b**) A type A cyst with pseudostratified ciliated columnar lining (hematoxylin and eosin X 100). (**c**) A neurenteric cyst containing cerebellar tissue (hematoxylin and eosin X 100). (From Shukla et al. [14]; with permission)

hyperintense lesions on fluid-attenuated inversion recovery (FLAIR) sequences with no perilesional edema. They can show mild restriction on diffusion-weighted imaging (DWI); however, frank restriction of diffusion has never been reported [59].

There is a characteristic large peak similar to N-acetylaspartate (NAA) on magnetic resonance spectroscopy (MRS) at 2.03 ppm despite the presence of neuronal contents in the cyst. The classical MRS peak is due to secreted contents and helps to differentiate the NEC from other cystic lesions [60]. A mural nodule is typically not seen in NECs; however, a "false" mural nodule can form as a result of epithelial cell secretion and epithelial cell desquamation [61, 62]. Other radiological features

such as rim enhancement [2, 29, 63–65], calcification [29, 31], and rapid expansion [37] of the cyst have also been reported. Supratentorial cysts are larger than their posterior fossa counterparts [29]. Hydrocephalus and other features of mass effect can also occur depending upon the location of the cyst. NECs can also be diagnosed in the prenatal period using MR imaging [66].

Differential Diagnosis

Owing to recent advances in neuroimaging, the reported incidence of NEC is rising. NECs need to be differentiated radiologically from other cystic lesions such as the epidermoid cyst, dermoid cyst, parasitic cyst (e.g., the hydatid cyst or neurocysticercosis), tumoral cyst, lipoma, neuroepithelial cyst, arachnoid cyst, other endodermal cysts (Rathke's and colloid cysts), cystic schwannoma, and craniopharyngioma [2, 16, 29, 67–69].

Epidermoid and dermoid cysts usually have irregular shapes, often encasing the adjacent neurovascular structures and infiltrating into contiguous cisterns. In comparison, NECs expand and displace the surrounding nerves and vessels. Dermoids can also be differentiated on the basis of fat suppression MRI imaging. Spinal epidermoid and dermoid cysts are usually located at the lumbosacral and cauda equina level and can have a (posterior) dermal sinus tract. Arachnoid cysts follow CSF intensity on all sequences. Other endodermal-derived cysts such as Rathke's and colloid cysts can be differentiated from NECs on the basis of their characteristic locations in the suprasellar region and the foramen of Monro region, respectively [70].

Craniopharyngiomas are hyperintense in T2-weighted images with bright contrast enhancement. NECs can be differentiated from neuroepithelial cysts by histopathological findings such as the presence of a basement membrane, cilia, and goblet cells with secretory granules. Neoplastic intramedullary lesions can be differentiated from these cysts by lack of contrast enhancement of the cyst wall and the absence of a mural nodule.

Surgical Management: Different Surgical Approaches

The goal of surgery for NECs is to alleviate the mass effect without causing dissemination, so complete excision of cyst is recommended. However, total excision is not always possible as the cyst wall is usually adherent to important structures such as the brainstem, spinal cord, cranial nerves and spinal nerve roots, and blood vessels. Small parts of the tumor capsule can persist and cause a recurrence with increased patient morbidity [14, 47, 71, 72]. There is a considerable recurrence rate from residual tumors, with the possibility of cranio-spinal dissemination and malignant transformation [14, 72].

The location of the cyst determines the operability, operative approach, and prognosis for the patient. For spinal NECs, there are three basic types of approaches – posterior, anterior, and lateral (Table 10.2). Each has its own merits and limitations.

Table 10.2 Different surgical approaches used for surgical excision of spinal neurenteric cysts in some of the major articles in literature

Serial no.	Studies	Location of spinal neurenteric cysts	Surgical treatment and approach
1	Song et al. (n = 1) [81]	Cervical spine	Lateral cervical approach
2	Abhishek et al. (n = 1) [82]	Intradural extramedullary ventrally located cystic lesion in cervical spine	Extreme lateral approach
3	Sasani et al. (n = 2) [83]	Cervical spine	Anterior approach
4	Laidlaw et al. (n = 1) [84]	Lower cervical spine	Ventral resection and anterior fusion utilizing sternal notch exposure
5	Tuzun et al. (n = 1) [85]	Upper cervical spine	Posterior approach
6	Liu et al. (n = 2) [86]	Cervicomedullary junction	Far lateral transcondylar approach
7.	Shukla et al. (n = 16) [14]	Cervicomedullary, cervical, thoracic, intradural extramedullary and intramedullary	Far lateral and posterior approaches

There is no consensus regarding the best approach, but the posterior approach is used most commonly worldwide. Even though most NECs are ventrally placed, the posterior approach provides an adequate surgical corridor. Various surgical options have been exercised such as cyst aspiration, cyst fenestration, partial excision, marsupialization, and cyst-subarachnoid shunting [73]. Owing to the high recurrence rate (0–37%), simple aspiration is not preferred.

Maneuvers such as cyst aspiration and cord manipulation can help in the surgery. Complications associated with this approach include cord or nerve root injury, epidural venous hemorrhage resulting in hematoma formation, and chances of cyst rupture leading to meningitis. The anterior approach using a corpectomy gives an anatomical advantage in the ease of accessing ventrally placed cysts, but additional instrumentation is usually required to provide stabilization. The anterolateral approach, which includes the far lateral and extreme lateral transcondylar approach at the foramen magnum (Fig. 10.3), the transpedicular and transfacetal approach with vertebral artery mobilization in the cervical spine, the costotransversectomy, extradural transpedicular approach, or the anterolateral approach at the thoracic spine is also reported in the literature. To access anteriorly placed lesions, the maneuvers adopted include turning the operating table obliquely to the contralateral side, a partial facetectomy along with lateral laminectomy, opening the dura in a T-shaped manner to connect the extradural and intradural portions anterolateral to the cord, sectioning of denticulate ligament at several levels, a good arachnoidal dissection, adequate cyst decompression from between the corridors provided by the nerve roots emerging from the spinal cord, and ensuring that the fibrous attachment of the tumor pedicle linking the last part of the tumor capsule to the spinal cord is divided carefully under vision. The cyst-cord boundary can easily be seen using this technique without cord manipulation or corpectomy.

Complete excision of a spinal NEC results in improvement of neurological symptoms in most (71%) cases [10]. Unfortunately, complete excision is not always achievable for intramedullary cysts, where the cyst wall is adherent to the spinal cord and there is no clear plane of dissection [74–76]. The extent of surgical resection must be weighed against the morbidity associated with the procedure. De Oliveira et al. have reported worsening of symptoms in 11% of their cases and failure to regain premorbid neurological function in 18% [38].

Surgical Outcome and Recurrence

Total resection of a spinal NEC often yields better improvement in associated motor and sensory deficits. After reviewing the case series of surgical outcomes of spine NECs, we found a worsening of symptoms in 11% of patients after surgical excision and failure to regain premorbid neurological function in 18% [76]. Although the literature on long-term outcomes is sparse, leading to a poor estimation of the true recurrence rate, the postsurgical recurrence mentioned ranges between 0% and 37%. Kim et al. and Cai et al. observed no recurrences in their case series of eight and seven patients, respectively [6, 77]. Holmes et al. observed recurrence in only 4% of their patients [38]. Chavda et al. reported the longest follow-up of 30 years and observed a 37% recurrence rate among their eight patients. Notably, all cases of recurrence in this study were in patients who had undergone partial surgical resection [74]. To date, the actual recurrence rate after partial resection of spinal NECs is unknown. Kimura et al. reviewed 18 cases with recurrent cysts. In 16 of them, only a partial cyst resection was achieved [71]. Paleologos et al. reported a case series in which there was no recurrence in patients who underwent gross total excision; in contrast, 11% of patients who underwent partial excision had a recurrence [78]. A similar trend was reported by Garg et al. No recurrence was observed in patients with gross total resection, whereas recurrence was seen in five out of eight (63%) patients with partial resection [13]. Menezes et al. found no correlation between recurrence and factors such as age, sex, cyst size, and level [79]. The estimated time gap for recurrent cysts to appear ranges from 2 months to 14 years after the initial partial resection. Therefore, a long-term clinical and neuroradiological follow-up is highly recommended [80]. There is no effective role for conventional radiotherapy and chemotherapy at present [80].

Consequently, the surgeon must try to achieve the primary treatment goal of gross total excision of the cyst contents and cyst wall while appreciating the possible morbidity associated with such a resection.

Conclusions

Spinal NECs occupy a particular part of the clinical spectrum of spinal diseases. These lesions display a characteristic histopathology, which includes a lining of well-differentiated columnar or cuboidal epithelium with or without cilia and mucus

globules. Patients with these heterotypic lesions of endodermal origin often present with myelopathy and/or radiculopathy. MRI is the gold standard for characterizing NECs and excluding other close differential diagnoses. CT scans are important for defining bony abnormalities that often coexist with displaced remnants of the developing gastrointestinal/respiratory tract. Complete surgical resection is the goal of treatment. However, the surgical plan must consider the merits and demerits of complete and partial resection in cases with no well-defined plane of cleavage between the cord and the cyst. With the documented recurrence rate as high as 37%, patients with partial resection of spinal NEC require regular clinical and radiological follow-up to test for re-accumulation of cyst contents.

References

1. Puusepp M. Variete rare de teratome sous-dural de la region cervicale (intestinome). Rev Neurol Paris. 1934;2:879–86.
2. Osborn A, Blaser S, Salzman K, et al. Neurenteric cyst. Diagn Imaging Brain Amirsys. 2004;I–7:40–1.
3. Harris CP, Dias MS, Brockmeyer DL, Townsend JJ, Willis BK, Apfelbaum RI. Neurenteric cysts of the posterior fossa: recognition, management, and embryogenesis. Neurosurgery. 1991;29(6):893–8.
4. Brooks B, Duvall E, ElGammal T, et al. Neuroimaging features of neurenteric cysts: analysis of nine cases and review of the literature. AJNR Am J Neuroradiol. 1993;14:735–46.
5. Eynon-Lewis N, Kitchen N, Scaravilli F, et al. Neurenteric cyst of the cerebellopontine angle: case report. Neurosurgery. 1998;42:655–8.
6. Kim CY, Wang KC, Choe G, et al. Neurenteric cyst: its various presentations. Childs Nerv Syst. 1999;15:333–41.
7. Kleihues P, Burger PC, Scheithauer BW. Histological typing of tumors of the central nervous system: World Health Organization. Berlin: Springer; 1993.
8. Savardekar A, Salunke P, Rane S, Chhabra R. Dorsally placed extradural infected neurenteric cyst in a two-year old with paraspinal extension. Neurol India. 2012;60:129–31.
9. Gumerlock MK. Epidermoid, dermoid and neurenteric cysts. In: Winn RH, editor. Youmans neurological surgery. 5th ed. Philadelphia: WB Saunders; 2004. p. 1223–30.
10. Can A, Dos Santos Rubio EJ, Jasperse B, Verdijk RM, Harhangi BS. Spinal neurenteric cyst in association with Klippel-Feil syndrome: case report and literature review. World Neurosurg. 2015;84:592.e9–14.
11. Singhal BS, Parekh HN, Ursekar M, Deopujari CE, Manghani DK. Intramedullary neurenteric cyst in mid thoracic spine in an adult: a case report. Neurol India. 2001;49:302.
12. Sharma R, Chandramouli TC, Rao RM. Ventral foramen magnum neurenteric cyst presenting as acute rapidly progressive quadriparesis and respiratory compromise: a case report and review of literature. Neurol India. 2013;61:187.
13. Garg N, Sampath S, Yasha TC, Chandramouli BA, Devi BI, Kovoor JM. Is total excision of spinal neurenteric cysts possible? Br J Neurosurg. 2008;22:241–51.
14. Shukla M, Behari S, Guruprasadh B, et al. Spinal neurenteric cysts: associated developmental anomalies and rationale of surgical approaches. Acta Neurochir. 2015;157:1601–10.
15. Gao P, Osborn AG, Smirniotopoulos JG, et al. Neurenteric cysts: pathology, imaging spectrum, and differential diagnosis. Int J Neuroradiol. 1995;1:17.
16. Simon JA, Olan WJ, Santi M. Intracranial neurenteric cysts: a differential diagnosis and review. Radiographics. 1997;17:1587–93.

17. Christov C, Chretien F, Brugieres P, et al. Giant supratentorial enterogenous cyst: report of a case, literature review, and discussion of pathogenesis. Neurosurgery. 2004;54:759–63.
18. Bejjani GK, Wright DC, Schessel D, et al. Endodermal cysts of the posterior fossa: report of three cases and review of the literature. J Neurosurg. 1998;89:326–35.
19. Gauden AJ, Khurana VG, Tsui AE, Kaye AH. Intracranial neuroenteric cysts: a concise review including an illustrative patient. J Clin Neurosci. 2012;19:352–9.
20. Wang L, Zhang J, Wu Z, et al. Diagnosis and management of adult intracranial neurenteric cysts. Neurosurgery. 2011;68:44–52.
21. Graziani N, Dufour H, Figarella-Branger D, Donnet A, Bouillot P, Grisoli F. Do the suprasellar neurenteric cyst, the Rathke cleft cyst and the colloid cyst constitute a same entity? Acta Neurochir. 1995;133(3–4):174–80.
22. Tucker A, Miyake H, Tsuji M, et al. Neurenteric cyst of the lower clivus. Neurosurgery. 2010;66(1):E224–5.
23. Sampath S, Yasha TC, Shetty S, et al. Parasellar neurenteric cyst: unusual site and histology: case report. Neurosurgery. 1999;44:1335–7.
24. Scaravilli F, Lidov H, Spalton DJ, Symon L. Neuroenteric cyst of the optic nerve: case report with immunohistochemical study. J Neurol Neurosurg Psychiatry. 1992;55:1197–9.
25. Morgan MA, Enterline DS, Fukushima T, et al. Endodermal cyst of the oculomotor nerve. Neuroradiology. 2001;43:1063–6.
26. Leventer DB, Merriam JC, Defendini R, et al. Enterogenous cyst of the orbital apex and superior orbital fissure. Ophthalmology. 1994;101:1614–21.
27. Bavetta S, El-Shunnar K, Hamlyn PJ. Neurenteric cyst of the anterior cranial fossa. Br J Neurosurg. 1996;10:225–7.
28. Cheng JS, Cusick JF, Ho KC, et al. Lateral supratentorial endodermal cyst: case report and review of literature. Neurosurgery. 2002;51:493–9.
29. Preece MT, Osborn AG, Chin SS, Smirniotopoulos JG. Intracranial neurenteric cysts: imaging and pathology spectrum. AJNR Am J Neuroradiol. 2006;27:1211–6.
30. Rauzzino MJ, Tubbs RS, Alexander E, et al. Spinal neurenteric cysts and their relation to more common aspects of occult spinal dysraphism. Neurosurg Focus. 2001;10:Article 2.
31. Tan GS, Hortobagyi T, Al-Sarraj S, et al. Intracranial laterally based supratentorial neurenteric cyst: case report. Br J Radiol. 2004;77:963–5.
32. Tubbs RS, Salter EG, Oakes WJ. Neurenteric cyst: case report and a review of the potential dysembryology. Clin Anat. 2006;19:669–72.
33. Kulkarni V, Daniel RT, Haran RP. Extradural endodermal cyst of posterior fossa: case report, review of the literature, and embryogenesis. Neurosurgery. 2000;47:764–7.
34. Gimeno A. Arachnoid, neurenteric and other cysts. In: Vinken PJ, Bruyn GW, editors. Congenital malformations of the spine and spinal cord. Handbook of clinical neurology, vol. 32. Amsterdam: North Holland Publishing Company; 1978. p. 421–46.
35. Jhawar SS, Nadkarni T, Goel A. Intraparenchymal temporal neurenteric cyst. J Clin Neurosci. 2011;18:415–7.
36. Teufack S, Campbell P, Moshel YA. Intracranial neuroenteric cysts: two atypical cases and review of the literature. JHN J. 2011;6:18–21.
37. Priamo FA, Jimenez ED, Benardete EA. Posterior fossa neurenteric cysts can expand rapidly: case report. Skull Base Rep. 2011;1:115–24.
38. de Oliveira RS, Cinalli G, Roujeau T, Sainte-Rose C, Pierre-Kahn A, Zerah M. Neurenteric cysts in children: 16 consecutive cases and review of the literature. J Neurosurg. 2005;103(6, Suppl):512–23.
39. Inoue T, Kawahara N, Shibahara J, et al. Extradural neurenteric cyst of the cerebellopontine angle: case report. J Neurosurg. 2004;100:1091–3.
40. Zalatnai A. Neurenteric cyst of medulla oblongata—a curiosity. Neuropediatrics. 1987;18:40–1.
41. Miyagi AMD, Katayama YMD. Neurenteric cyst arising in the high convexity parietal lesion: case report. Neurosurgery. 2007;60:E203–4.

42. Basheer N, Kasliwal MK, Suri A, Sharma MC, Arora A, Sharma BS. Lateral extradural, supratentorial neurenteric cyst. J Clin Neurosci. 2010;1:639–41.
43. Dadlani R, Ghosal N, Khetan R, Hegde AS. Neurenteric cyst of the cerebello-pontine angle in an infant presenting with obstructive hydrocephalus. Neurol India. 2012;60:247–9.
44. Adams RD, Wegner W. Congenital cyst of the spinal meninges as cause of intermittent compression of the spinal cord. Arch Neurol Psychiatr. 1947;58:57–69.
45. Dunham CP, Curry B, Hamilton M. Malignant transformation of an intraaxial-supratentorial neurenteric cyst – case report and review of the literature. Clin Neuropathol. 2009;28:460–6.
46. Monaco R, Boscaino A, Di Blasi A, et al. Intraepithelial carcinoma arising in an endodermal cyst of the posterior fossa. Neuropathology. 2003;23:219–24.
47. Wang W, Piao YS, Gui QP, Zhang XH, Lu DH. Cerebellopontine angle neurenteric cyst with focal malignant features. Neuropathology. 2009;29:91–5.
48. Ko BS, Jung S, Jung TY, Moon KS, Kim IY, Kang SS. Neurenteric cyst with xanthomatous changes in the pre-pontine area: unusual radiological findings. J Neurosurg Pediatr. 2008;2:351–4.
49. Schwartz AM, Jensen ME, Saks DA, Ghatak NR. Epithelial cyst in cerebellopontine angle with xanthogranulomatous changes simulating cholesterol granuloma. Surg Neurol. 1989;31:454–8.
50. Wilkins RH, Odom GL. Spinal intradural cysts. In: Handbook of clinical neurology. New York: Wiley; 1976. p. 55–102.
51. Gessi M, Legnani FG, Maderna E, Casali C, Solero CL, Pollo B, et al. Mucinous low-grade adenocarcinoma arising in an intracranial enterogenous cyst: case report. Neurosurgery. 2008;62:E972–3.
52. Ho LC, Olivi A, Cho CH, Burger PC, Simeone F, Tihan T. Well-differentiated papillary adenocarcinoma arising in a supratentorial enterogenous cyst: case report. Neurosurgery. 1998;43:1474–7.
53. Natrella F, Mariottini A, Rocchi R, Miracco C. Supratentorial neurenteric cyst associated with a intraparenchymal subependymoma. BMJ Case Rep. 2012;2012.bcr0120125566.
54. Fujita T, Kayama T, Saito S, et al. Immunohistochemical detection of tumor marker in recurrent clivus enterogenous cyst – case report. Neurol Med Chir (Tokyo). 1997;37:479–82.
55. Rivierez M, Buisson G, Kujas M, et al. Intramedullary neurenteric cyst without any associated malformation. One case evaluated by RMI and electron microscopic study. Acta Neurochir. 1997;139:887–90.
56. Ormond DR, Omeis I, Mohan A, Murali R, Narayan P. Obstructive hydrocephalus due to a third ventricular neuroepithelial cyst. J Neurosurg Pediatr. 2008;1:481–4.
57. Nakase H, Ishida Y, Tada T, Sakaki T, Goda K, Tunoda S, et al. Neuroepithelial cyst of the lateral ventricle: clinical features and treatment. Surg Neurol. 1992;37:94–100.
58. Czervionke LF, Daniels DL, Meyer GA, Pojunas KW, Williams AL, Haughton VM. Neuroepithelial cysts of the lateral ventricles: MR appearance. AJNR Am J Neuroradiol. 1987;8:609–13.
59. Medhi G, Saini J, Pandey P, Mahadevan A, Prasad C. T1 hyperintense prepontine mass with restricted diffusion – a white epidermoid or a neuroenteric cyst? J Neuroimaging. 2015;25:841–3.
60. Candiota AP, Majós C, Bassols A, Cabanas ME, Acebes JJ, Quintero MR, et al. Assignment of the 2.03 ppm resonance in in vivo 1H MRS of human brain tumour cystic fluid: contribution of macromolecules. MAGMA. 2004;17:36–46.
61. Paolini S, Ciappetta P, Domenicucci M, Guiducci A. Intramedullary neurenteric cyst with a false mural nodule: case report. Neurosurgery. 2003;52:243–5.
62. Cho JM, Ahn JY, Kim SH, Lee KS, Chang JH. An endodermal cyst mimicking an intra-axial tumor in the medulla oblongata. Childs Nerv Syst. 2010;26:853–6.
63. Shakudo M, Inoue Y, Ohata K, et al. Neurenteric cyst with alteration of signal intensity on follow-up MR images. AJNR Am J Neuroradiol. 2001;22:496–8.
64. Evans A, Stoodley N, Halpin S. Magnetic resonance imaging of intraspinal cystic lesions: a pictorial review. Curr Probl Diagn Radiol. 2002;31:79–94.

65. Hayashi Y, Tachibana O, Muramatsu N, et al. Rathke cleft cyst: MR and biomedical analysis of cyst content. J Comput Assist Tomogr. 1999;23:34–8.
66. Gulrajani M, David K, Sy W, Braithwaite A. Prenatal diagnosis of a neurenteric cyst by magnetic resonance imaging. Am J Perinatol. 1993;10:304–6.
67. Hingwala DR, Radhakrishnan N, Kesavadas C, Thomas B, Kapilamoorthy TR, Radhakrishnan VV. Neuroenteric cysts of the brain-comprehensive magnetic resonance imaging. Indian J Radiol Imaging. 2013;23:155–63.
68. Lonjon M, Paquis P, Michiels JF, et al. Endodermal cyst of the foramen magnum: case report and review of the literature. Childs Nerv Syst. 1998;14:100–3.
69. Chaynes P, Thorn-Kany M, Sol JC, et al. Imaging in neurenteric cysts of the posterior cranial fossa. Neuroradiology. 1998;40:374–6.
70. Afshar F, Sholtz CL. Enterogenous cyst of the fourth ventricle: case report. J Neurosurg. 1981;54:836–8.
71. Kimura H, Nagatomi A, Ochi M, Kurisu K. Intracranial neurenteric cyst with recurrence and extensive craniospinal dissemination. Acta Neurochir. 2006;148:347–52.
72. Perry A, Scheithauer BW, Zaias BW, Minassian HV. Aggressive enterogenous cyst with extensive craniospinal spread: case report. Neurosurgery. 1999;44:401–5.
73. Muzumdar D, Bhatt Y, Sheth J. Intramedullary cervical neurenteric cyst mimicking an abscess. Pediatr Neurosurg. 2008;44:55–61.
74. Chavda SV, Davies AM, Cassar-Pullicino VN. Enterogenous cysts of the central nervous system: a report of eight cases. Clin Radiol. 1985;36:245–51.
75. Savage JJ, Casey JN, McNeill IT, Sherman JH. Neurenteric cysts of the spine. J Craniovertebr Junction Spine. 2010;1:58–63.
76. Holmes GL, Trader S, Ignatiadis P. Intraspinal enterogenous cysts. A case report and review of pediatric cases in the literature. Am J Dis Child. 1978;132:906–8.
77. Cai C, Shen C, Yang W, Zhang Q, Hu X. Intraspinal neurenteric cysts in children. Can J Neurol Sci. 2008;35:609–15.
78. Paleologos TS, Thom M, Thomas DG. Spinal neurenteric cysts without associated malformations. Are they the same as those presenting in spinal dysraphism? Br J Neurosurg. 2000;14:185–94.
79. Menezes AH, Traynelis VC. Spinal neurenteric cysts in the magnetic resonance imaging era. Neurosurgery. 2006;58:97–105.
80. Menezes AH, Ryken TC. Craniocervical intradural neurenteric cysts. Pediatr Neurosurg. 1995;22:88–95.
81. Song JK, Burkey BB, Konrad PE. Lateral approach to a neurenteric cyst of the cervical spine: case presentation and review of surgical technique. Spine (PhilaPa 1976). 2003;28:E81–5.
82. Abhishek A, Anushree A, Patir R, Sehgal AD. Extreme lateral approach in a case of acute-onset quadriplegia due to high cervical neurenteric cyst. Pediatr Neurosurg. 2007;43:134–6.
83. Sasani M, Ozer AF, Oktenoglu BT, Peker K, Bozkus MH, Sarioglu AC. Excision of an asymptomatic cervical intradural neurenteric cyst through the anterior approach: a study of two cases and a review of the literature. Spine J. 2007;7:720–7.
84. Laidlaw JD. Iso-intense neuroenteric cyst in the lower cervical spine treated with ventral resection and anterior fusion utilising sternal notch exposure: case report, technical note and literature review. J Clin Neurosci. 2003;10:606–12.
85. Tuzun Y, Izci Y, Sengul G, Erdogan F, Suma S. Neurenteric cyst of the upper cervical spine: excision via posterior approach. Pediatr Neurosurg. 2006;42:54–6.
86. Liu JK, Couldwell WT. Far-lateral transcondylar approach: surgical technique and its application in neurenteric cysts of the cervicomedullary junction. Report of two cases. Neurosurg Focus. 2005;19:E9.