Neurenteric cysts are rare congenital lesions, and they are even rarer when located intramedullarily. MR images are very helpful for a diagnosis of neurenteric cysts and preoperative planning of treatment. Complete recovery of neurological functions is usually possible after surgical decompression, but the chance of recurrence still remains to be determined. The authors present 2 cases with cervical neurenteric cysts: one was intradural extramedullary and the other was intramedullary in location. The clinical features, MR image characteristics, and management of neurenteric cysts are discussed. Neurenteric cysts must be considered whenever a cystic lesion is present within the spinal canal.

Case 1

A 10-year-old girl complained of left forearm and shoulder pain 3 months prior to admission. These symptoms resolved after medical treatment. Approximately 2 months after the initial symptoms, the girl presented with neck stiffness and ataxia. Examination on admission revealed a positive Babinski’s sign on the right side. Muscle power was generally normal in all extremities, and the right supinator and knee reflexes showed hyperreflexia. All her sensory functions were intact. Other physical examinations were unremarkable.

A non-enhanced brain CT and subsequent plain radiograph of the thoracic spine were normal. Magnetic resonance images (MRIs) of the cervicothoracic spine showed an intradural extramedullary cystic lesion anterior to the cervical spinal cord which was isointense to cerebrospinal fluid (CSF) on T1-weighted images.
(T1WIs) (Figure 1a) and slightly hyperintense to CSF on T2-weighted images (T2WIs) (Figure 1b). Compression of the spinal cord was seen. The bony vertebrae and intervertebral discs appeared normal. The patient was then transferred to another hospital for surgery. After decompression of the cyst with a fine needle inserted between the lamina of C6 and C7, laminectomy from C4 to C7 was performed with gross resection of the intradural extramedullary cystic lesion. A neurenteric cyst lined by cuboidal to ciliated columnar epithelium was proven by pathology. The girl had no complaints, and her neurologic functioning was normal 6 months after surgery.

Case 2

A 5-year-old boy was first admitted to our hospital because of a 2-day history of intermittent fever, headaches, neck stiffness, and unstable gait. Physical examination at the time of admission was unremarkable. The neurological examination revealed negative Kernig’s and Brudzinski’s signs. The deep tendon reflex was 2+ in the bilateral upper extremities and 3+ in both lower extremities. Meningitis and an intracranial lesion were the first 2 considerations. CSF examination showed no significant abnormality, and no growth of microorganisms was seen in CSF and urine cultures. Brain CT was normal. An MRI of the cervical spine revealed an oval, sharply demarcated intramedullary cystic lesion with low signal intensity at the level of C4 to C6 on coronal and sagittal T1WIs (Figure 2a), which became bright on T2WIs (Figure 2b). No enhancement was seen after intravenous injection of contrast medium. Under the impression of an intramedullary cystic lesion, the boy was scheduled to have an operation. After total laminectomy of C4 to C5 with intradural exploration, a cystic mass was found in the cord from the level of C3 to C5. The cyst was then punctured and partially removed under a microscope. Histology showed only dense collagenous tissue that may have belonged to the dura or ligaments. No definitive diagnosis was achieved.

About 14 months later, the boy was admitted to our hospital again due to progressive neck pain and stiffness. His muscle power was generally normal and sensations were intact. A repeat MRI of the cervical spine showed a focal area of an intramedullary lesion at the level of C4 and C5 with predominant iso-signal intensity and stippling of high signal intensity spots on T1WIs (Figure 3a). There was evidence of homogeneous and focal lobulated high signal intensity of this lesion demonstrated on a gradient echo proton density image and T2WI (Figure 3b). After

Figure 1. Cervicothoracic MR images of Case 1. a. Sagittal T1-weighted image (TR/TE=500/25) showing a well-demarcated intradural extramedullary cystic lesion with anterior compression on the cord. The lesion is hypointense to the cord and isointense to CSF. b. The lesion having become significantly hyperintense to the cord and only slightly hyperintense to CSF on a sagittal T2-weighted image (3000/100). The bony vertebrae of the cervicothoracic spine appear intact.
intravenous contrast medium administration, this lesion showed no significant abnormal enhancement. During operation, the specimen was sent to the lab for freeze-sectioning. Microscopic examination revealed that a portion of the cystic wall was composed of a thin fibrous or gliotic wall lined by pseudostratified ciliated columnar epithelium with abundant vacuolar cytoplasm, suggesting a single layer of intestinal-type epithelium (Figure 4). Focal incomplete squamous metaplasia was present. The subsequent permanent pathologic specimen confirmed the result. On the basis of these histologic features, the definitive diagnosis was a neurenteric cyst.

Figure 2. MR images of Case 2 obtained before the first operation. a. Sagittal T1-weighted image (450/20) revealing a well-demarcated cystic lesion within the cervical spinal cord with low signal intensity similar to CSF. The cord is expanded. b. Sagittal T2-weighted image (2800/100) showing that the cyst is hyperintense to the cord and is almost the same as CSF.

Figure 3. MR images of Case 2 obtained before the second operation. a. Sagittal T1-weighted image (500/25) showing that the previous cystic lesion has become isointense with peripheral high signal intensity spots after the first operation (arrowhead). b. On a sagittal T2-weighted image (4000/100), the lesion, clearly identifiable, is hyperintense to the cord and slightly hyperintense to CSF.
The postoperative course was uneventful, and the patient’s condition gradually improved during the 5 months after surgery.

**DISCUSSION**

Although various theories have been proposed to explain the formation of neurenteric cysts, it is widely accepted that these cysts originate from abnormal separation of the endoderm and neuroectoderm in the third week of embryonic life, resulting in the inclusion of endodermal elements in the spinal canal. They are also thought to be components of the split notochord syndrome [1]. Woo and Sharr concluded that there seemed to be 2 types of cysts: “developmental cysts” and “teratomatous cysts”. “Developmental cysts” are typically found in the cervical segment lying anterior to the spinal cord with no associated spinal defects. For this type, symptoms usually appear after the second decade or later. By contrast, “teratomatous cysts” are typically found at the level of the conus lying posterior to the cord or roots and are frequently associated with spinal defects. Generally, symptoms of the latter type appear early in life probably due to major abnormalities [2].

The clinical presentations of neurenteric cysts are mainly the results of spinal cord and nerve root compression. Pain may be prominent, and is usually related to the area where the cysts are located. According to the previous literature, many patients with neurenteric cysts have a long and intermittent clinical course resembling multiple sclerosis [3]. Our Case 2 had symptoms of fever, headaches, and neck stiffness with prodromal flu-like illness typical of aseptic meningitis despite the absence of lymphocytes in CSF. This is presumably caused by spontaneous rupture of the cyst with leakage of its contents into the subarachnoid space, which results in meningeal irritation [4, 5].

Neurenteric cysts are most frequently located in the lower cervical and upper thoracic regions followed by the lumbar area, with very few case reports located at the craniovertebral junction [1, 6, 7]. They are largely intradural extramedullary, and intramedullary neurenteric cysts are rare. According to Agnoli’s review of 29 cases of intraspinal neurenteric cysts, 24 were intradural extramedullary, 4 were intramedullary, and 1 was extradural in location [8].

On CT, these cysts may be hypo-, iso-, or hyperdense depending on their content. MRI is superior to CT because it can not only disclose the precise location and extent of the cystic lesions useful for surgical approach, but also can characterize these lesions on the basis of the intensity pattern of the cyst contents, thereby narrowing the differential diagnosis and improving diagnostic accuracy. The sensitivity of MRI is also higher than CT because of its excellent contrast resolution. In general, neurenteric cysts can be differentiated from solid tumors because they usually have distinct margins and uniform signal intensity iso- or hyperintense to CSF on both T1- and T2WIs. By contrast, solid tumors frequently possess indistinct margins and nonuniform signal intensity with non-isointensity to CSF [9]. Absence of contrast enhancement within a lesion further confirms its cystic nature. Kjos et al. categorized intracranial cystic lesions into 3 groups on the basis of the intensity pattern of cyst contents: 1. Arachnoid and postoperative cysts have an intensity pattern identical to CSF. 2. More proteinaceous cysts, such as inflammatory cysts or nonhemorrhagic tumoral cysts, have an intermediate intensity pattern with characteristically low intensity (but often slightly more intense than CSF) on the short TR sequence (0.5 second), but have clearly higher intensity than CSF on long TR sequences (2 second). 3. Hemorrhagic cysts in certain stages have a distinctly different pattern of high intensity on all pulse sequences like colloid cysts (TR/TE=500/28, 2000/28, and 2000/56) [10].
our Case 1, the cyst content had a signal intensity almost identical to CSF despite T2WI showing a slightly higher signal intensity than CSF. A similar condition was observed on the MR images of Case 2 obtained before the first operation. Such an intensity pattern suggested that the cyst contents were CSF-like or contained low protein. Interestingly, subsequent MR images of Case 2 taken 14 months following the first operation disclosed that the signal intensity of the lesion was predominantly isointense to the spinal cord on T1WI, probably due to heavy protein content or to hemorrhaging. The signal intensity of neurenteric cysts may be CSF-like due to low protein content, or they may be hyperintense to CSF on T1WIs with or without fluid-fluid level due to high protein content or to viscid mucus.

An intramedullary neurenteric cyst should be distinguished from other types of intramedullary cysts, cystic cord tumors such as astrocytoma and hemangioblastoma, syringomyelia, intradural extramedullary masses with an associated syrinx, and myelomalacia. Although MRI may help narrow the differential diagnosis, diagnosis of a neurenteric cyst is sometimes difficult to achieve. The concomitant presence of vertebral defects with or without extraspinal lesions would suggest a diagnosis of neurenteric cyst.

The management of neurenteric cysts is still controversial. Some authors emphasize that complete excision of the cyst is needed to prevent recurrence [11, 12], but others consider that biopsy with decompression is adequate [13]. In our Case 2, neurologic functions improved after partial removal and decompression of the cyst in the first operation, but the symptoms recurred after 14 months. Although the risk of recurrence remains uncertain, we would suggest complete excision of the cysts if technically feasible. As for intramedullary cysts, complete excision is usually impossible. Evacuation of the cyst contents with partial removal of the cyst wall and shunting of the cyst cavity into the subarachnoid space for the prevention of recurrence has been reported [4]. Posterior laminectomy is a widely performed technique for neurenteric cysts, but sometimes the total removal of the anteriorly located cysts is doubtful because of lack of direct visualization of the lesions. Devkota et al. [12] and Geremia et al. [14] removed the cyst wall completely with corpectomy via an anterior approach, but long-term follow-up for degenerative diseases of the spine is important in these patients. Generally speaking, the outcome of neurenteric cysts after decompression is satisfactory, and complete recovery of neurological functions is often possible according to the literature.

REFERENCES

頸部神經腸囊腫之磁振造影特徵：二例病例報告

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神經腸囊腫是造成幼兒及小孩脊髓壓迫罕見的原因。這些先天性的異常是因為在胚胎發育時，內胚層與神經外胚層不正常的分離所造成的。神經腸囊腫的術前診斷並不容易。我們在此報告二例患有神經腸囊腫的病例，並回顧相關文獻。磁振造影對神經腸囊腫的術前診斷及手術計畫，均能提供相當有用之訊息。神經腸囊腫在手術切除之後，神經系統之功能往往能完全恢復，但是其術後復發率仍有待商榷。當脊椎管中出現囊腫狀病灶時，我們必須將神經腸囊腫的可能性列入考慮。

關鍵詞：磁振造影，頸部脊椎，脊髓壓迫