CASE IN POINT

CRANIOCERVICAL NEURENTERIC CYST

Yashodeep P. Jadhav, MD, Pankaj Singh, MD, and Sanjay V. Bhat, MD

This patient's lesion was unusual in that it was both intraspinal and intracranial—and it was not associated with other spinal abnormalities.

Neurenteric cysts are relatively rare developmental malformations resulting from incomplete separation of the developing notochord and foregut in the embryo.[1–5] They may be purely spinal, mediastinal, or part of a complex malformation of the gut and the spine.[6–10]

Typically, neurenteric cysts are located near the cervicothoracic junction.[11,12] Vertebral anomalies often are present and include hemivertebrae, spina bifida, diastematomyelia, or fibrous remnant of the connection to the foregut.[13–15] Intraspinal neurenteric cysts are usually extramedullary and intradural.[1,5,7]

We report an atypical case of a neurenteric cyst at the craniocervical junction. In addition to its location, the cyst was unusual in that it was not associated with other spinal anomalies.

Dr. Jadhav is a staff radiologist, **Dr. Singh** is a staff gastroenterologist, and **Dr. Bhat** is a staff radiologist, all at the Olin E. Teague Veterans Center, Central Texas Veterans Health Care System, Temple.

INITIAL EXAM

A 25-year-old man, whose job involves operating a computer, came to the emergency department of a tertiary care center with weakness and inability to move his arms. He had begun to note pain in the posterior part of the neck about four weeks before, with increasing severity. Two weeks earlier, he had experienced an electric shock sensation in his arms and down his legs, known as Lhermitte sign,[16] which was exacerbated by neck flexion and certain other neck movements. The intermittent electric sensations recurred four days prior to presentation, and approximately three days before presentation, the patient developed urinary retention that lasted three days.

On neurologic examination, his motor tone was increased with sustained clonus in both lower extremities at the knees and ankles. His motor power in both upper extremities was 1/5 at the deltoid, biceps, and grip; in the lower extremities it was 3/5. The flexors of the feet were 3/5. He had an extensor plantar response bilaterally to plantar reflex testing and his superficial abdominal reflexes were absent. Sensory examination revealed diminished pinprick sensation from the mid pinna line to the level of the shoulders. There was an absence of position and vibratory sensation in the hands. In the lower extremities the pinprick, position, and vibratory sensations were intact.

Magnetic resonance imaging (MRI) showed a sausage-shaped lesion anterior to the medulla and upper cervical cord, extending from just above the foramen magnum to mid C2 level. It showed high signal intensity on T1-weighted, T2-weighted, and proton density MRI (Figures 1 and 2), which suggested a high protein content.

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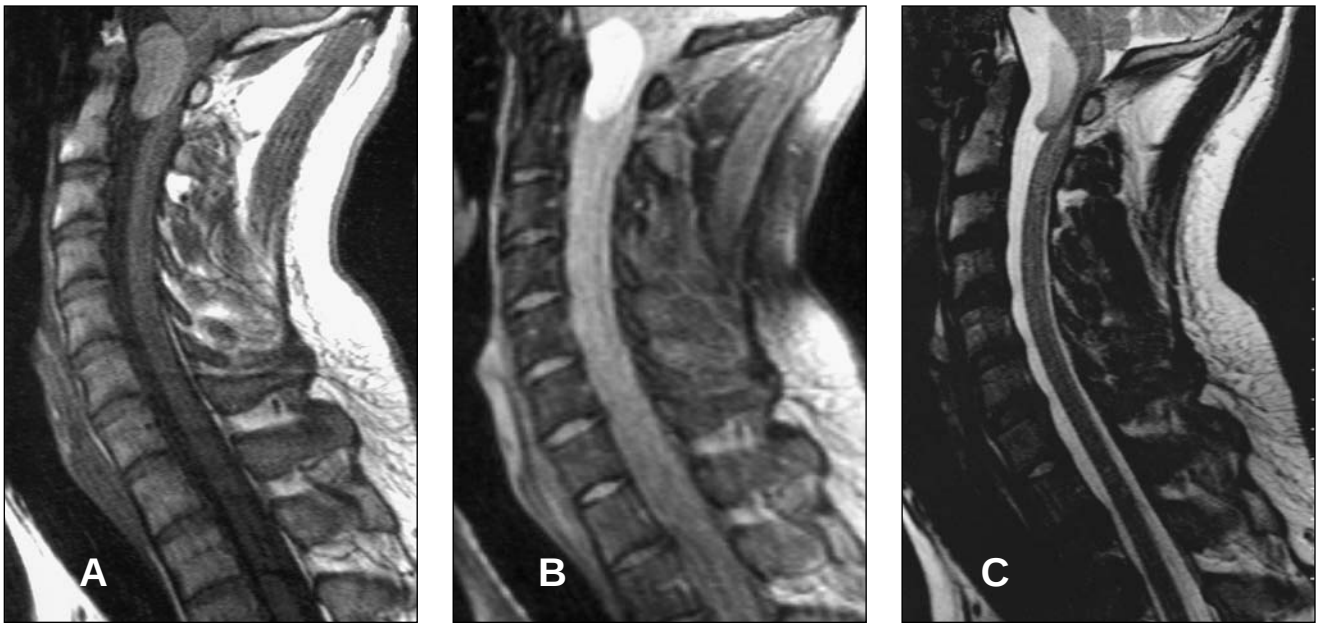

Figure 1. (A) Sagittal T1-weighted magnetic resonance imaging (MRI) scan (TR/TE, 500/15), showing a high signal intensity neurenteric cyst at the craniocervical junction with fluid-fluid level. (B) Sagittal proton density MRI scan (TR/TE, 2500/36), showing entire lesion to be hyperintense. (C) Sagittal T2-weighted MRI scan (TR/TE, 5420/96), showing nondependent, less proteinaceous component to be isointense to cerebrospinal fluid.

A fluid level was seen within the lesion, most likely related to the settling of higher proteinaceous content. The spinal cord was severely compressed due to the size of the cyst. It occupied approximately 90% of the canal diameter.

TREATMENT COURSE

The patient underwent surgery, which involved laminectomy of the C1 and the upper part of the C2 vertebrae, as well as enlargement of the foramen magnum. Once the dura mater was opened, the cystic lesion was identified and emptied and most of the capsule was removed. The extracted fluid was yellowish and odorless, and cultures did not grow any organisms.

Histologic examination of the excised sections revealed portions of a cystic structure, the larger of which had a cavitary space and papillary infoldings, lined by a single layer of cuboidal and columnar epithelium. The columnar epithelium resembled gastric type, with occasional interspersed goblet cells. The cavitary space contained amorphous, lightly basophilic material.

The patient recovered well from surgery, with resolution of his neurologic symptoms. Follow-up MRI at four and 14 months after surgery showed a small residual cyst along the anterior aspect of the proximal cord. To date, the patient remains neurologically intact and has not experienced the shock sensation in his upper and lower extremities.

ABOUT THE CONDITION

The notochord develops during the third week of embryogenesis. The small canal at the level of the primitive pit that transiently connects the yolk sac with the amniotic cavity is termed the neurenteric canal.[2,5] This canal allows temporary contact between the endoderm and the developing neuroectoderm (Figure 3).[17] The presence of this canal may interfere with notochordal development and result in the formation of a neurenteric cyst.[2,5]

Alternatively, persistent endodermal-ectodermal adhesions or adhesions between the notochord and endoderm may produce notochordal dysgenesis and result in a neurenteric cyst.[1–3] These cysts often are connected by a fibrous tract, fistula, or cleft to structures derived from the primitive gut in the thoracic or abdominal cavities and are associated commonly with vertebral anomalies.[3,5,17] These findings support the theory that neurenteric cysts originate from incomplete separation of the noto-

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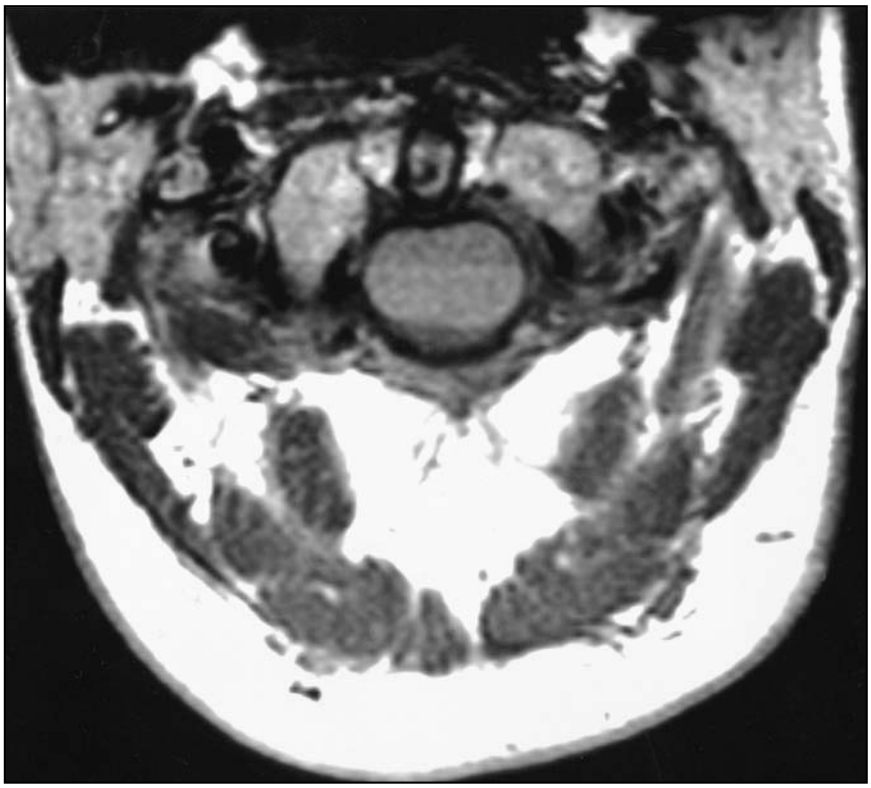

Figure 2. T1-weighted axial magnetic resonance imaging scan, showing the neurenteric cyst.

chord and primitive gut. By contrast, intracranial neurenteric cysts have not been associated with such abnormalities, and this theory of histogenesis when applied to intracranial lesions is based mainly on the histopathology of the cyst.[18]

In a 1984 literature review, Agnoli and colleagues identified 33 reported cases of histologically verified enterogenous intraspinal cysts.[13] Of these, 18 (55%) were located in the cervicodorsal spine. Most (80%) of the 33 cases were intradural extramedullary lesions; 12% were intramedullary.

Among a total of 119 patients with intraspinal cysts reviewed by Wilkins and colleagues,[11,19] 76 were found in men and 43 were found in women, for a male to female ratio of nearly two to one.[20] The diagnosis was established during the first decade of life in 41 patients (34%) and during the second decade in 27 patients (23%). Seven cysts (6%) were found in patients over age 50. The most common location was in the cervical or thoracic spinal canal, anterior or anterolateral to the spinal cord, with occasional intramedullary involvement (Table).[11,19,20]

The case that we describe herein was rare in that the patient's cyst was located both intracranially and intraspinally. Furthermore, despite its intraspinal, intradural location, it was not associated with spinal anomalies.

MRI is the best diagnostic tool and the method of choice for investigation of neurenteric cysts. The

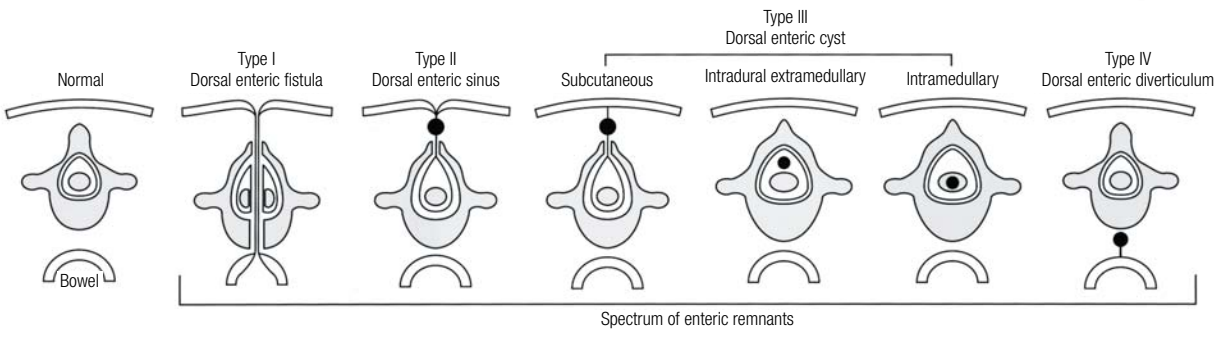

Figure 3. Developmental posterior enteric remnants. Adapted from: Bentley JF, Smith JR. Developmental posterior enteric remnants and spinal malformations: The split notochord syndrome. *Arch Dis Child*. February 1960;35:76–86.[17] © Copyright 1960 by BMJ Publishing Group. Used with permission.

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Table. Common locations of neurenteric cyst, in order of decreasing frequency

Location in the spine
- Cervicothoracic junction
- Thoracic spine
- Cervical spine
- Lumbar spine
- Craniocervical junction
- Cranium

Location relative to the spinal cord
- Anterior
- Anterolateral
- Dorsal (either intradural extramedullary or extradural)
- Intramedullary

signal intensity pattern of cystic lesions on MRI varies with the nature and protein content of the intracavitary fluid. Cerebrospinal fluid (CSF) generates a high signal on T2-weighted images and a low signal on T1-weighted images. Arachnoid cysts, which contain CSF (as do most postoperative cysts), follow a CSF signal in all pulse sequences. Hemorrhagic and colloidal cysts exhibit a higher signal intensity compared to CSF on T1-weighted, T2-weighted, and proton density sequences. Proteinaceous nonhemorrhagic cysts have an intermediate signal intensity that is similar to or slightly higher than that of CSF on T1-weighted sequences and higher than that of CSF on T2-weighted and proton density sequences.[15] In the case we present here, the initial MRI appearance suggested a high protein content.

Histologically, neurenteric cysts resemble gastrointestinal mucosa. For this reason, and due to the variety of locations and characteristics associated with them, neurenteric cysts also go by many other names, including teratoma, teratoid cyst, teratomatous cyst, gastrocytoma, bronchogenic intraspinal cyst, archenteric cyst, enterogenous cyst, and intestinoma.[7,12] The World Health Organization classifies them under the heading of "other malformative tumors and tumorlike lesions" and describes them as cysts "lined by mucin secreting epithelium resembling that of the gastrointestinal tract."[21]

Wilkins and Odum have classified these cysts into three types based upon the histologic features of the cyst wall and its contents.[11] Under this classification system, the walls of Type A cysts mimic gastrointestinal or respiratory epithelium, with a basement membrane supporting single or pseudostratified cuboidal or columnar cells, which may be ciliated. Type B cysts also contain glandular organization, usually producing mucin or serous fluid. Type C cysts are the most complex, containing ependymal or glial tissue within the cyst. ●

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