

An Intramedullary Neurenteric Cyst in the Conus Medullaris with Recurrent Meningitis

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Neurenteric cysts are rare congenital lesions of the spine that are lined with endodermal epithelium. Their most common location is the cervico-dorsal region, and the mass usually lies ventral to the spinal cord. However the conus medullaris area location is an uncommon location. Neurenteric cysts are best treated by decompression and as near total excision of cyst membrane as possible. We report a case of a 7 year-old-girl with a neurenteric cyst in the conus medullaris. The patient had a history of meningitis and a gait disturbance. Magnetic resonance imaging (MRI) showed an intramedullary mass lesion in the conus medullaris with syringomyelia. There was no associated bone or soft-tissue anomaly. The mass was subtotally removed through a posterior approach. However 4 months later, meningeal irritation signs developed and MRI showed recurrence of the cyst. At the second operation, the cystic membrane was totally removed and the patient's neurological symptoms improved postoperatively. We reports a case of recurred neurenteric cyst occurred in unusual location with the review of literature.

KEY WORDS : Intramedullary neurenteric cyst · Conus medullaris · Recurrent meningitis.

Introduction

Neurenteric cyst is a rare congenital malformation, and originates through adhesion and other communication between the neuro-ectoderm and endoderm during the third

gestational week. The most common location is the cervico-odorsal region, and usually a single cyst lies ventral to the spinal cord¹⁾. Intramedullary neurenteric cysts have been reported in fewer than 5% of all neurenteric cysts^{1,3,9,13,14)}. A surgical prognosis appears favorable even with subtotally resection^{1,6,9)}.

We report a case of a neurenteric cyst with recurrence in the conus medullaris.

Case Report

A 6-year-old girl presented with lower back pain and intermittent gait disturbance on 2 months. She had a history of meningitis for one month. Even though cerebrospinal fluid (CSF) culture provided no growth of a causative organism, she was treated with intravenous antibiotics for 4 weeks. On admission, she showed no

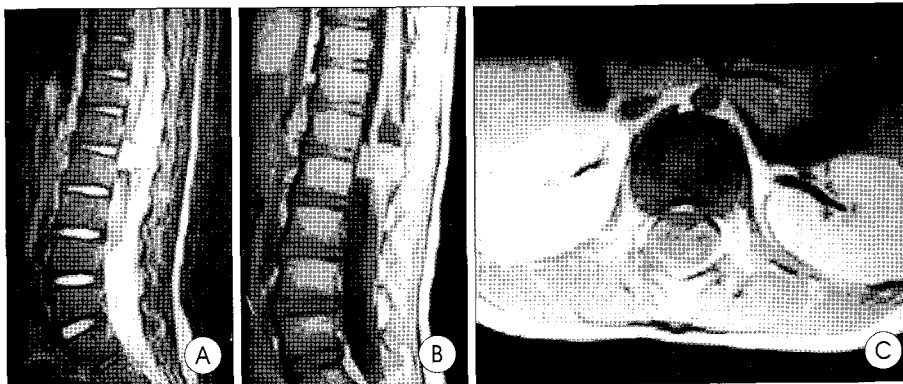


Fig. 1. Preoperative T2 (A) and T1 (B)-weighted contrast-enhanced magnetic resonance images of an intramedullary neurenteric cyst, that has ruptured into the subarachnoid space. In the conus medullaris area, the spinal cord is expanded focally by an intramedullary cyst filled with homogenous fluid. The mass is not enhanced and the cyst slightly hyperintense to cerebrospinal fluid, and isointense to spinal cord on T1-weighted images (C).

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abnormalities except motor-sensory changes with the presence of a mild increased knee jerk and ankle jerk(+ + + / + + +). Babin-ski's sign and ankle clonus were equivocal. An X-ray study of the spine revealed no abnormal bony structure. However, magnetic resonance imaging (MRI) revealed a L2-3 intramedullary lobulating cystic lesion that was slightly hyperintense to CSF, isointense to the spinal cord on T1-weighted images, and hypointense to CSF on T2-weighted sequences (Fig. 1). The lesion was not enhanced on post-contrast T1-weighted images. Vertebra and soft tissue were normal, and laboratory blood and urine finding were within normal limits. The patient underwent surgery by the posterior route. When the dura was opened, partially transparent pia matter and a thinned cord at the dorsal midline were seen. We utilized a posterior midline myelotomy to evacuate the cystic contents which were milky and mucous-like in appearance (Fig. 2). Subtotal excision of the cystic wall was performed because the plane of cleavage between the cyst wall



Fig. 4. Microscopic findings of the intramedullary neurenteric cyst of the spinal cord. This case shows simple tall columnar cells with basally located nuclei. In addition, the wall contains occasional neuromelanocytes (arrows). (H&E, X200).

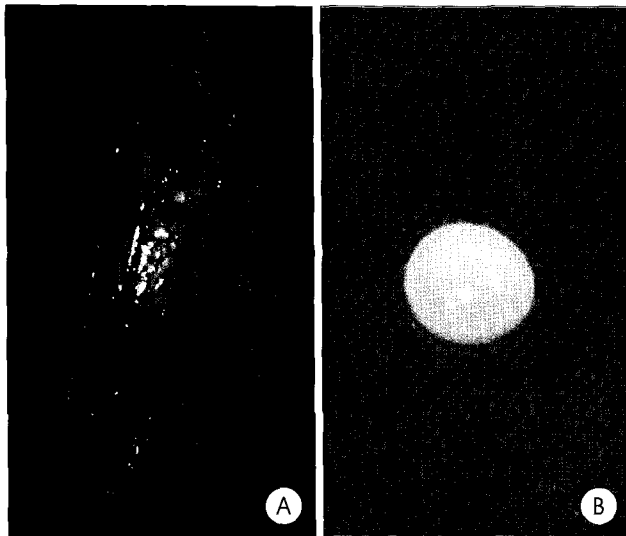


Fig. 2. Intraoperative photograph of the spinal cord and cyst at first operation. The spinal cord has widened focally and shows prominent surface neovascularity (A). The cyst contains a milky white fluid (B).

and the spinal cord was illdefined. Histopathological findings showed no pus, and mucus containing epithelial cells, a thin-walled fibrous cystic lesion lined by simple total columnar cells with basally located nuclei. Based on these histological findings, we made a diagnosis of neurenteric cyst. After operation, the gait disturbance improved. However, 3 months later, she visited the emergency department again due to severe headache and vomiting. She was alert but there was evidence of neck stiffness and a strongly positive Kernig's sign. CSF analysis showed features of bacterial meningitis (white blood cell (WBC)/red blood cell (RBC) 7430/60, total protein (TP)/glucose (Glu) 441/89), but no causative organism was identified in CSF culture. Repeated MRI showed an increased mass with syringomyelia (Fig. 3). The second operation was performed through the previous route. After dura opening, pus like material was found within a thin capsule. This material was sucked out and the cyst wall was totally grossly removed. Histopathological findings showed

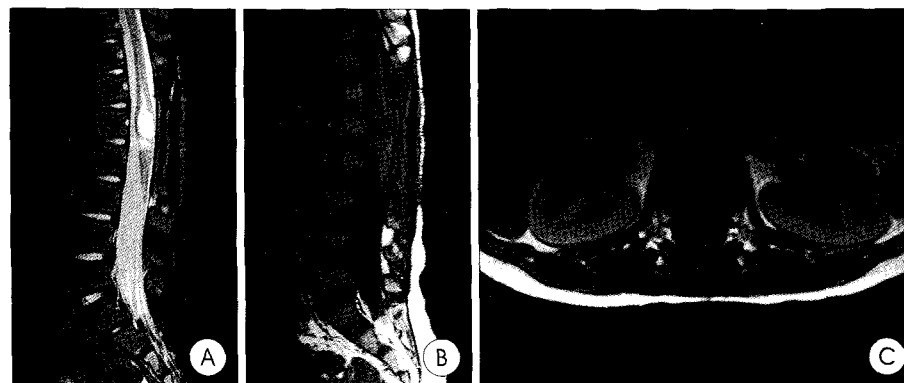


Fig. 3. Second magnetic resonance image finding. A, B, C : Three months after the index operation, the mass signal is not changed, but the sizes of the mass and syringomyelia are greater.

a thin-walled fibrous cystic lesion as same as previous operation finding. Based on these histological findings, we made a diagnosis of neurenteric cyst (Fig. 4). After operation, she was recovered from infection signs.

One year after second operation, follow up MRI was checked. MRI finding showed well visualized syrinx at the same site, but there was no evidence of recurrence and there was no progressing neurologic deficit (Fig. 5).

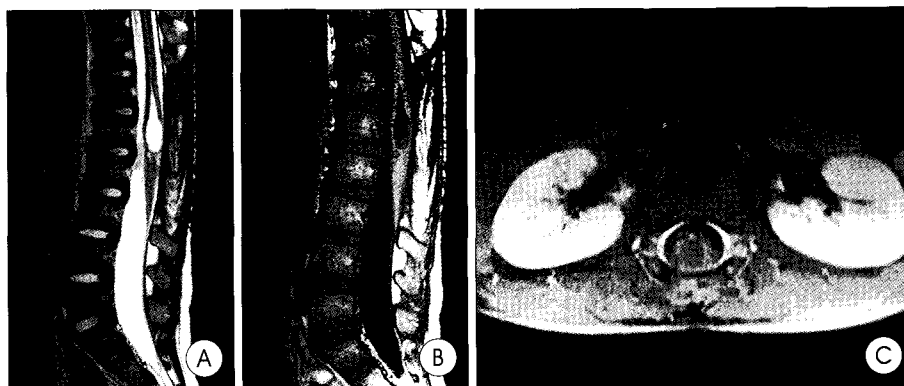


Fig. 5. Magnetic resonance image finding of one year after second operation. A, B, C : There is well visualized syrinx at the same site, but there is no evidence of recurrence.

Table 1. Reported cases of intramedullary neurenteric cysts in literature

Author, Year	Age /Sex	MRI finding	Symptom	Location	Cyst contents	Surgical procedure	Outcome
Rewcastle & Fancoeur, 1964	34/F	not reported	Paraparesis	T7-L2	not reported	Subtotal resection	Worse paresis
Fortuna et al, 1984	47/F	T12 intramedullary cyst, no other dysraphic cyst	Lt. LE paresis	T12-L1	not reported	Subtotal resection	Improved
Rivierez et al, 1997	46/M	L1-2 intramedullary dyst, on other dysraphic cyst	LBP, LE paresis	L1-2	Clear, colorless	Cystosubarachno id shunt	Worse paresis
Lippman et al, 2000	68/F	T10 intramedullary cyst, no other dysraphic anomalies	LBP, paraparesis	T10-11	Milky fluid	Subtotal resection and marsupialization	Worse paresis
Present case, 2006	6/F	L1-2 intramedullary dyst, on other dysraphic cyst	LBP, Gait disturbance	L1-2	Milky fluid	Subtotal resection and marsupialization	Improved

* : Abbreviations : M : male, F : female, LBP : low back pain, Lt : Left, LE : lower extremity

Discussion

True intramedullary neurenteric cysts are highly uncommon and fewer than 5% of all neurenteric cysts are situated in the intramedullary compartment⁹. MRI evaluation was available for 4 cases which are located area of the conus medullaris (Table 1)^{2,7-9,11-14}. Moreover, intramedullary neurenteric cysts are not associated with other congenital abnormalities⁹. Intramedullary neurenteric cysts are usually located at the extremes of the spinal cord¹³, which may be due to the later closure of the neural tube at these levels. To the best of our knowledge, intramedullary neurenteric cyst on the conus medullaris has not been previously reported. These cysts, their clinical courses and radiologist findings are likely to be affected by fluctuations in the rate of mucin production and absorption by the cyst wall¹⁵. Usually in cases of intramedullary lesion, there is no clear cleavage plane between the spinal cord and the cyst wall. Thus operative interventions in cases of intramedullary neurenteric cysts have ranged from cyst aspiration to subtotal resection with cyst marsupialization or cystosubarachnoid shunting^{2,7-9,11-14}.

Simple aspiration is less desirable, because delayed recurrence of the cyst has been reported¹⁰. On the other hand, subtotal resection with cyst marsupialization has yielded good immediate and long-term results⁵, although some cases recurred like the presented case^{4,12}. However, in the presented case, the cause of recurrence was not clear. A re-capsulated cystic wall was observed in the second operation field, and this might be attributable to insufficient cyst wall resection at first operation. Although, it has been recommended that, these lesions be resected completely as possible, however it is difficult to conclude as to whether the initial resection was sufficient, because it is very rarity of this disease⁹. In such cases, spinal cord monitoring can be a useful adjunct to excision⁹. In terms of the suspected recurrent meningitis, treatment was initiated even in the absence of a causative organism. Now we consider that the findings which are supporting the impres-

sion of meningitis were caused by cyst rupture, that aseptic meningitis was developed and no causative organism was present.

We report a case of recurred neurenteric cyst occurred in an unusual location that was complicated by possible recurrent meningitis and included a review of the literature.

Conclusion

An intramedullary neurenteric cyst is a rare disease and is presented on a clear cleavage plane between the spinal cord and the cyst wall, which explains to an extent the variable treatment modalities adopted. Even subtotal resection with cyst marsupialization has been recommended as a treatment modality, although recurrence is possible as it has occurred in our case. In the present case, the authors attribute the recurrence to insufficient cyst wall resection.

In the cases of intramedullary neurenteric cyst, cystic wall must be resected as much as possible using spinal cord monitoring.

The authors describe a case of a recurrent neurenteric cyst that occurred in an unusual location.

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