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Case Report

Spinal intramedullary ependymal cyst: a case report and review of the literature

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Summary

We report a rare case of spinal intramedullary ependymal cyst in a 44-year-old female and reviewed 12 cases reported in the literature. The patient presented with slowly progressive lower limb paresis. She underwent biopsy of the cyst wall and placement of a cysto-subarachnoid shunt with complete recovery at the follow-up examination 18 months after surgery. This is a benign lesion and appropriate management should be performed at an early stage of the disease.

Keywords: Cyst-subarachnoid shunt; ependymal cyst; intramedullary; spine; endoscope.

Introduction

Spinal intramedullary ependymal cyst is extremely rare. Only 12 pathologically proven cases have been reported in the literature [1–4, 7, 8, 11–14]. We report an additional case of a 44-year-old female with intramedullary lumbar ependymal cyst. Clinical presentation, histological characteristics, image findings, surgical management, and outcome of this disease entity are discussed here.

Case presentation

A 44-year-old female complained of slowly progressive right lower limb weakness which had started 1 year before the present admission. There was no history of trauma to the spine. Neurological examination demonstrated a moderate paresis, atrophy and decreased touch, pain and cold sensation in the right leg. Urodynamic study and somatosensory evoked potentials of the lower extremities were normal. Spinal X-ray did not show abnormal findings. Magnetic resonance imaging (MRI) demonstrated a cystic lesion measuring $11 \times 12 \times 45$ mm at the Th11-L1 level. The cyst was iso-intense to cerebrospinal fluid on both T1- and T2-weighted images. There was no capsular or nodular enhancement noted on contrast administration. The cyst was located in the anterior ventral portion of the spinal cord (Fig. 1).

She underwent right hemilaminectomy at Th12 and partial hemilaminectomy at Th11 and L1. After the dura was opened under the microscope, we made a linear myelotomy over the dorso-lateral surface of the cyst and a large cyst cavity filled with clear fluid was encountered. We examined the inside of the cavity using a small flexible endoscope (0.9 mm in diameter, EnodGnost®, POLYDIAGNOST GmbH, Pfaffenhofen, Germany), but there was no apparent mural nodule (Fig. 2A). We removed small pieces of the cyst wall to confirm the diagnosis and to open the cyst wall widely enough to create good communication with the subarachnoid space. Then we placed a cysto-subarachnoid shunt tube measuring 1.4 mm wide and 40 mm long between the cyst and subarachnoid space. Intraoperative monitoring with lower extremity somato-sensory evoked potentials and electromyographic responses of the sphincter and lower extremity muscles did not showed any significant change during the procedure.

The postoperative course was uneventful, and the patient was discharged with improving mobility and sensibility two weeks after surgery. The cystic lesion was markedly reduced in size on postoperative MRI obtained eighteen months after surgery (Fig. 1C). No enhancing nodule suggesting neoplasm was detected on contrast enhancement during this follow-up. She had recovered full lower extremity strength and there were no other clinical symptoms or signs noted 18 months after surgery.

Pathological examination showed ependymal cells lining the cyst, positive for the immunohistochemical staining with CAM5.2, KL-1 and EMA. Cilia like structures were noted along the medial cell surface on highly magnified images (Fig. 2B, C, D).

Discussion

There are frequent reports of extramedullary spinal ependymal cysts. However, an intramedullary spinal ependymal cyst is a very rare lesion. Hyman *et al.* first reported a cervical intradural ependymal cyst in 1938 [6].

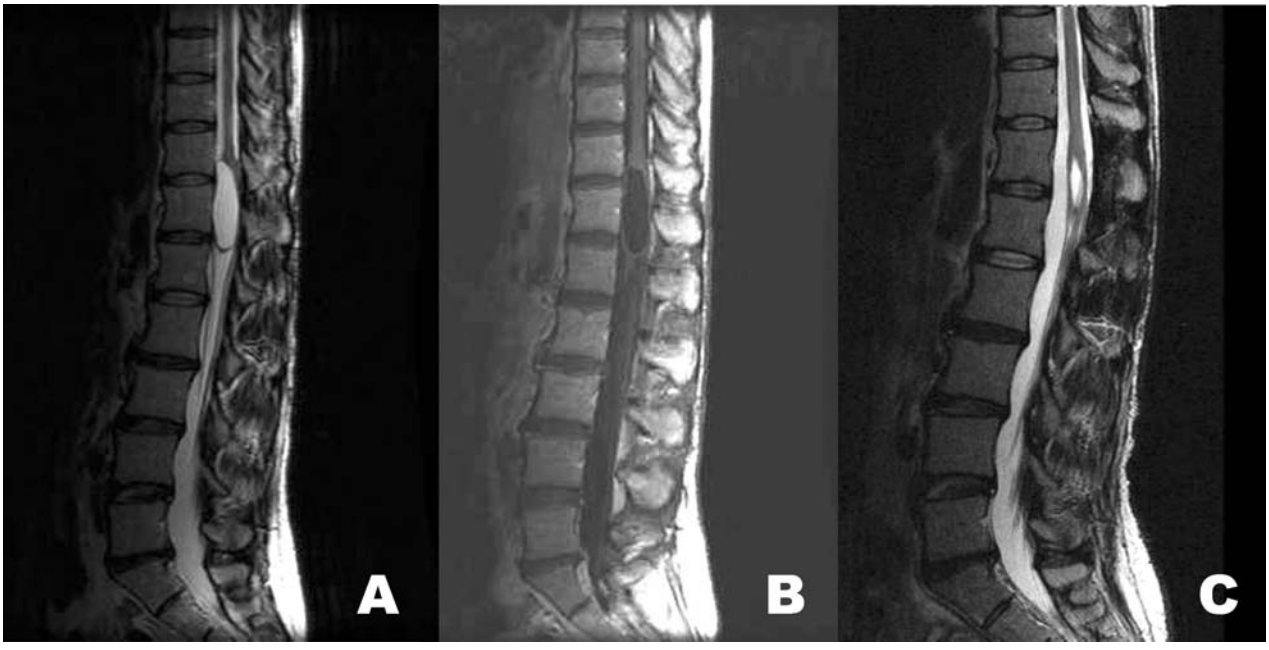

Fig. 1. Pre-operative sagittal magnetic resonance imaging (*MRI*) with T2-weighed image showed a large cystic mass lesion in the conus, at the level of T11 to L1 (A). T1-weighed MRI image with contrast on gadolinium diethylene triamine penta-acetic acid (Gd-DTPA) administration showed no apparent enhancing lesion inside the cyst (B). Post-operative sagittal T2 weighed MRI of the lower spinal cord showed marked shrinkage of the cystic mass lesion inside the conus (C)

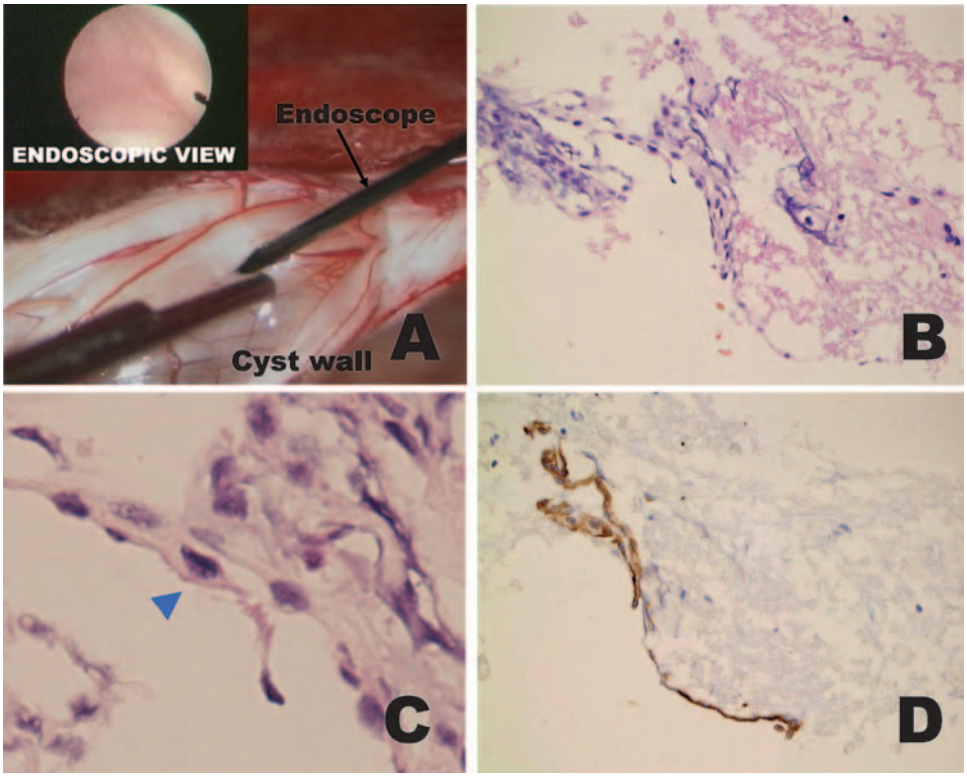

Fig. 2. (A) Intraoperative photograph showing microscopic finding of the cyst and endoscopic view inside the cyst obtained with a 0.9 mm fiberscope. There was no apparent tumour inside the cyst cavity. The pathological photos of surgical specimens obtained from the cyst wall showed thin ependymal cells lining the cavity (B). Very fine cilia like structures were noted on higher magnification (C; Arrow head). The cells were positive for the immunohistochemical staining with CAM5.2 (D), EMA and KL-1

Table 1. *Summary of reported cases of intramedullary spinal ependymal cysts*

No.	Author	Age/sex	Clinical presentation	Duration	Location	Surgery type*	Result/recurrence†
1	Fortuna (case 1) [4]	67/F	intermittent paresis	5 years	T12	Op	self-sufficient with crutches
2	Fortuna (case 2) [4]	57/F	medullary pain	6 years	T12	Op	recovered
3	Dharker [2]	38/F	backache, paraparesis	1 month	T6	Op	no improvement
4	Roussearu [13]	71/F	radicular pain	53 years	T12	Op	little improvement
5	Findler [3]	6/M	cervical pain and weakness	3 weeks	C7-T1	Op	complete recovery
6	Sharma [14]	7/M	urinary hesitancy, paraparesis	1 year	T4	R	complete recovery
7	Pagni [11]	39/M	paresthesia of C6–7 territory	?	C5–7	R	self-sufficient with crutches
8	Robertson (case 1) [12]	48/F	paresthesia	3 years	Bx	Op	little improvement
9	Iwahashi [7]	1/F	weakness	1 month	T6	Op	complete recovery
10	Kumar (case 1) [8]	5/M	excessive falls	?	T4–6	R	recovered
11	Kumar (case 2) [8]	4/M	neck pain and weakness	5 months	C1–5	Op	improved
12	Chhabra [1]	15/M	quadriparesis	?	C2–3	Op	recovered/recurred†
13	Saito	44/F	weakness	1 year	T11-L1	Op + shunt	complete recovery

* Type of surgery: *R* resection of the cyst, *Op* Biopsy or partial removal of the cyst wall and opening, marsupialization of the cyst, shunt; cysto-subarachnoid shunt. † Recurrence: recurrence of the cyst was only found in case #12.

In Table 1, we have summarized the 13 reported cases including our experience to characterize this pathological entity and to direct the most suitable management strategy [1–4, 7, 8, 11–14]. There was no gender preference (6 male and 7 female) and a wide range of ages from one to 71 year of age was shown. Variable clinical presentation was reported including intermittent paresis of the lower limbs, radicular pain, paresthesia, and quadriparesis according to the location and size of the lesion. Location of the cyst also varied from the upper cervical region to the conus, with the conus being the most frequent site (5 cases).

The most widely accepted hypothesis regarding the pathogenesis is that the floor plate of the neural tube is evaginated on the ventral side and becomes isolated, then a cyst forms later [6]. The location of isolated ependymal tissue determines whether the spinal cyst becomes extramedullary or intramedullary. Hence, this lesion is usually separated from the central canal and anteriorly off-center.

Pathologically, the ependymal cyst is characterized by a lining of epithelium, either columnar or cuboidal with or without cilia, that lacks a basement membrane and rests on fibrous tissue [5, 8]. Some pseudostratification may be seen. These cysts are also variously referred to as glio-ependymal cysts, neuroepithelial cysts and choroidal epithelial cysts [10]. Cyst content confirmed as clear fluid during the surgical procedure in all but one case, which demonstrated thick turbid fluid [1].

Magnetic resonance imaging is the best method for evaluating and differentiating spinal cord cysts. The borders of the cyst appear smooth and well defined. The ependymal cyst is iso-intense with CSF on T1- and T2-weighted and proton density images without contrast enhancing lining or nodule on gadolinium diethylene triamine penta-acetic acid (Gd-DTPA) administration. To exclude a neoplastic lesion associated with cyst formation, careful follow-up with contrast-enhanced MRI should be planned even if the initial imaging did not show the enhancing lesion. The cyst is usually located in the anterior cord and off-center from the central canal. This non-central location and confined nature of the cyst should be important radiological findings that are useful for differentiating such cysts from congenital or traumatic syringomyelia in combination with the clinical history. We also should differentiate this lesion from another rare cystic pathology in the conus; the ventriculus terminalis, which is a congenital cystic dilatation in the midline conus usually seen in the pediatric age group [9, 15]. This may be related to spinal dysraphism. But there are few non-pediatric patients reported with this condition [9], and clear differentiation may be sometimes difficult. We assume some of the cases included in the reported group of terminal ventricle might include the ependymal cysts. In differentiating cystic conus lesions, we would like to stress that the off-central location of the cyst, no association with spinal dysraphism, and/or non-pediatric onset of the symptoms predispose the diagnosis of ependymal cyst.

The treatment is essentially surgical. Full recovery from neurological deficit secondary to cord compression can be achieved with early decompression. While total enucleation was possible in three patients, in the other 10 patients, biopsy was carried out and a communication between the cyst cavity and subarachnoid space was established with satisfactory outcome. Recurrence was reported in one patient whose cyst was containing turbid fluid, and the nature of the cyst fluid may have indicated high protein content and might explain the recurrence [1]. In the current case, we placed a small shunt tube to maintain communication between the cyst and subarachnoid space, and our patient recovered completely demonstrating a good neurological condition with no sign of recurrence on imaging studies. Endoscopic confirmation of the absence of any nodular lesions inside the cyst might be useful if the technology is available.

In summary, spinal ependymal cyst is an isolated intra- and extra-parenchymal cyst lined with ependyma, which is usually located in the anterior cord. The cyst does not communicate with the central canal and is located off-center and should be differentiated from syringomyelia or terminal ventricle. Also enhancing nodules should be searched for and followed-up to rule out the possibility of a hidden intramedullary neoplasm. The management strategy should focus on a local procedure providing adequate decompression and establishing communication between the cyst cavity and subarachnoid space. The nature of this lesion is benign and usually shows good recovery of function with a low recurrence rate. Hence, early diagnosis and appropriately less invasive management should be entertained according to the strategy discussed here.

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Comment

The authors report a very rare case of intramedullary ependyma cyst located in the conus region. The literature on this entity is well reviewed and the mechanism of cyst formation is discussed. The authors differentiate intramedullary cyst from other cystic lesions of spinal cord. We agree that the treatment is essentially surgical. A wide opening of the cystic cavity and creating a communication with subarachnoid space is sufficient. The manuscript is well written.

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