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Clinical case

INTRAMEDULLARY EPENDYMAL CYST OF THE CERVICAL SPINAL CORD

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Abstract

Relevance: Intramedullary ependymal cyst is rarely seen pathology. At present there is insufficient information about the intramedullary ependymal cysts (23 cases reported in the literature) in the modern specialized literature. Ependymal cysts are considered to develop due to central nervous system ontogeny defect that occurs in the early stages of fetal development. The neurological symptoms are varied and depend on their localization Surgical treatment of such intramedullary mass lesions is associated with high risk of neurological complications.

Material and methods: The article presents a clinical case of successful surgical treatment of a patient with intramedullary ependymal cyst at the cervical spinal cord with the neurophysiological intraoperative monitoring.

Results: The use of the microscopic magnification, fine microsurgical instruments, neurophysiological intraoperative monitoring allows safe removal of intramedullary mass lesion without postoperative neurologic deficit.

Conclusion: Intramedullary ependymal cyst is a rare and poorly studied entity. The neurological picture of the cysts varies and depends on the location (anterior, posterior or collateral sides of spinal cord). Surgical removal is dangerous due to the high risk of neurological deficit, but is possible in selected cases. Histological examination is usually inconclusive due to the functional significance of spinal cord tissue and the inability to obtain adequate amounts of material for analysis. The results of obtained specimens do not always allow to establish organ affiliation of the tissue.

Keywords: *intramedullary ependymal cyst, cervical spine, surgical treatment.*

Introduction

Intramedullary ependymal cyst is rarely seen pathology. There are 23 clinical observations reported in foreign modern neurosurgical literature. There is no information about patients with the above mentioned nosological form in Russian literature [*Robertson D, Kirkpatrick J, 1991; Morest D, Silver J, 2003; Saito K et al., 2005; Lalitha A, Rout P, 2006; Radouane B et al., 2007; Park C et al., 2012; Byvaltsev V et al., 2013; Byvaltsev V et al., 2014; Franceschini P, Worm P, 2014*].

Ependymal cysts are considered to develop due to CNS ontogeny defect that occurs in the early stages of fetal development. The neurological

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symptoms are varied and depend on their localization [*Robertson D, Kirkpatrick J, 1991; Saito K et al., 2005; Lalitha A, Rout P, 2006; Radouane B et al., 2007; Park C et al., 2012; Franceschini P, Worm P, 2014*]. The surgical treatment of intramedullary mass lesions is associated with the high risk of neurological complications [*Nedzved M, 1990*]. Biopsy diagnosis is difficult to obtain due to functional significance of spinal cord tissue and the inability to obtain adequate amount of material for analysis. The results of obtained specimens do not always allow to establish the organ affiliation of the tissue [*Saito K et al., 2005; Lalitha A, Rout P, 2006; Byvaltsev V et al., 2013*].

This article presents a clinical case of successful surgical treatment of a patient with intramedullary ependymal cyst at the cervical spinal cord with the neurophysiological intraoperative monitoring.

Clinical observation

Patient B of 41 years of age was brought to the "Road Clinical Hospital at Irkutsk-Passenger station" with complaints of pain in the lower extremities, weakness and numbness in hands and feet.

Anamnesis morbi: Patient considers herself sick for about 5 years since she felt pain in the cervical spine, upper limb numbness, weakness, fatigue. The patient periodically received courses of conservative treatment by outpatient neurologist with a positive dynamics. Due to the progression of the weakness in the upper and lower extremities, the patient was directed to the Center of Neurosurgery of "Road Clinical Hospital at Irkutsk-Passenger station" for further evaluation.

Objective status: The general condition is of moderate severity and stable. The condition is active. The consciousness is clear. Skin and visible mucous membranes are clean and have normal color. The thoracic cage is in normal shape. The respiration is clear and symmetrical from both sides. The heart tones are clear, rhythmical, without murmurs. Heart rate is 80 per min. Blood pressure is 115/75 *mm.Hg*. The abdomen is soft and painless. No symptoms of peritoneal irritation. The liver size is not increased. The spleen is not palpable. Stool is normal. Urination is normal without urgency, incontinence or delay.

The neurological status on admission: Patient distinguishes fragrances, the sight fields are normal, ocular motility is in full volume. Pupils D (right) = S (left), with live reaction to the light and convergence, no diplopia. The trigeminal points are painless. Face is symmetrical. There is no nystagmus. Hearing is fine. There are no abnormal bulbar signs. Muscular tonus is normal. The tongue is straight. Swallowing is normal. Movements of the cervical spine are painful while bending. Second degree of increased paravertebral muscles tone. Reflexes: biceps D≥S, triceps D≥S, carporadial D≥S, alive. Percussion of the thoracic spine is painless. Lumbar lordosis is smoothed. Movements in the lumbar spine are in full range and painless. Knee-jerk D≥S, Achilles reflex D≥S is reduced. Lassegue signs are D – 65°, S – 65°. Bekhteriev, Rossolimo and Babinski reflexes are positive on both sides. Muscle tone in the upper extremities is decreased, hypertonicity in the lower extremities. The strength in the hands is 4/5 (D = S), in the legs is 4.5/5 (D = S). There are no sensory or pelvic disturbances.

Results of additional examination methods: MRI of the cervical spine (Fig. 1) showed 16x11x8*mm* ovoid intramedullary lesion at C_5-C_6 levels, more to the left with hyperintense on T2 and hypointense on T1 weighted images, which has homogeneous structure and do not enhances after contrast administration. Subarachnoid spaces are compressed at the C_5-C_7 levels.

The patient did not have traumatic injuries, spine injuries or medical manipulation on the cervical level which eliminates the chances of traumatic origin of the cyst due to the anamnesis

The complexity of the operative therapy was due to the intramedullary location of the lesion at the cervical level. The main objective was not to aggravate the neurological deficit after surgery. ISIS IOM system (Inomed, Germany) was used to reduce the risk of neurological complications of surgery an intraoperative neuromonitoring system.

Detailed study of clinical data and the additional methods' results determined primary diagnosis: Intramedullary cystic lesion at the C_5-C_6 vertebral levels. Upper flaccid paraparesis, lower spastic paraparesis. Left-sided extrapyramidal disturbance. Cervical pain syndrome. Syndrome of increased cervical muscle tone.

Operation: Microsurgical reconstruction of the spinal canal: C_5-C_6 laminectomy. Mielotomy and

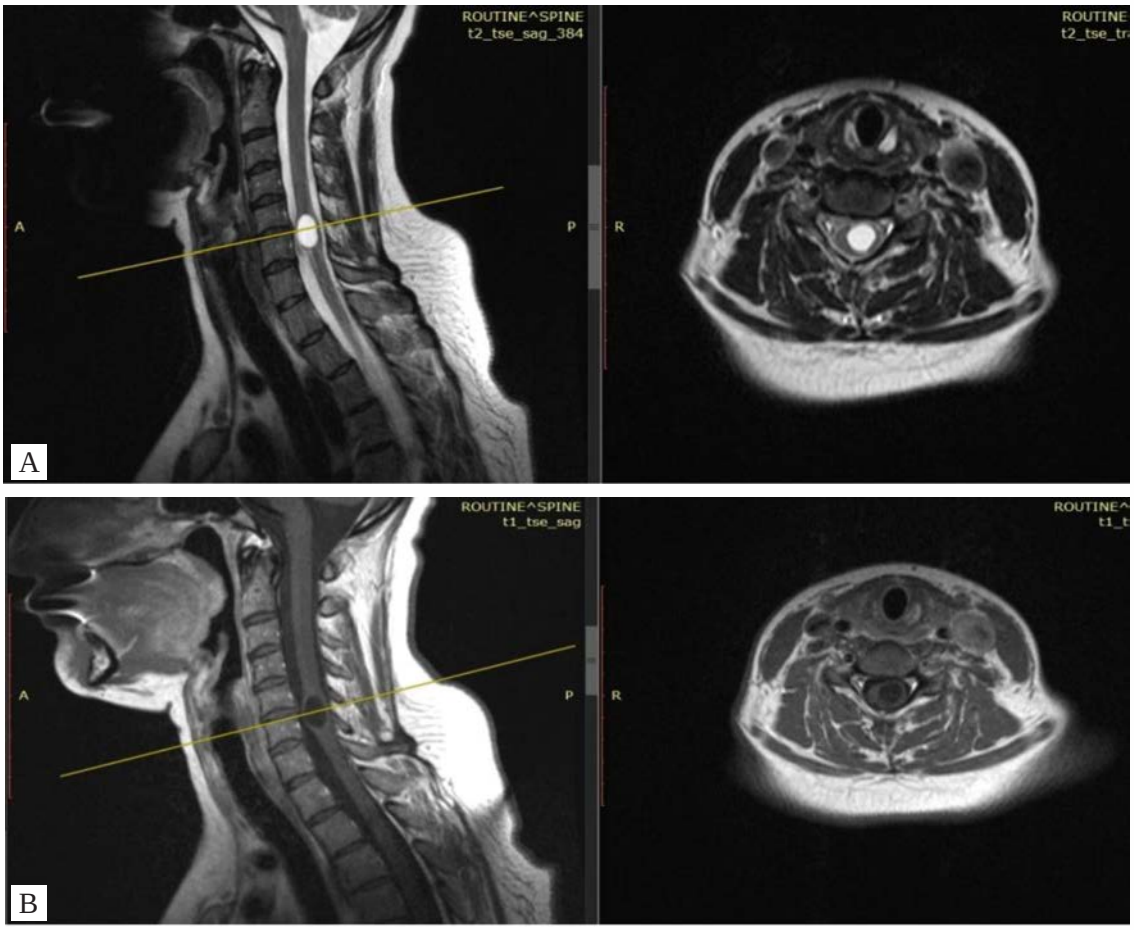

Figure 1. *Preoperative MRI of the cervical spine: A,B – mode T1; B,C – mode T2 with contrast.*

drainage of the ependymal cyst.

Patient was placed in prone position with rigid fixation of the head in the Maifield frame. The level of incision was planned with C-arm (Siemens, Netherlands) after the antiseptic preparation. A standard midline access was performed with resection of spinous processes and laminectomy of C_5-C_6 vertebras. Under the microsurgical magnification (Pentero 900, Karl Zeiss, Germany) a dura mater was visualized and a longitudinal section was done (4 *cm*). After microsurgical dissection of arachnoidal membrane 3 midline myelotomies were performed with a blunt dissector in the posterior fissure, creating a cyst opening of 1,5x2 *mm.* The content of the cyst was evacuated by aspirator (Fig. 2). A piece of cyst wall was taken with biopsy microforceps for histology (Fig. 3). Microsurgical hemostasis was achieved. Suture of wound was in layers. Operation duration - 4 hours 20 minutes. Blood loss - 50 *ml*.

Intraoperative neuromonitoring was conducted by using the following methods: somatosensory evoked potentials (SSEP), transcranial motor evoked potentials (TkMEP). In case of SSEP, needle stimulating electrodes were installed in the projection n. tibialis, n. medianus on both sides, the scalp needle recording electrodes were placed in points Cz, Fz, C3, C4 (according to the international 10-20 system). TkMEP were recorded through the inserted needle electrodes into muscles tenor of hands, m.tibialis anterior from feet, points C1, C2 were stimulated by spiral scalp electrodes. The initial values of the stimulation: SSEP - n. tibialis 20 *mA*, n. medianus 21.86 *mA*; TkMEP - 79 *mA*.

As a result of the stimulation the baseline of SSEP was allocated (Table 1) based on which the comparison with intraoperative fluctuations amplitudes was carried out.

During the surgery no significant changes of evoked potentials (over 50% in amplitude, over 10% of the latency) were identified. There was a temporary decrease in the amplitude of SSEP components by 10-30%, which may have been due to the anesthetic aid.

The patient was allowed to increase activity and

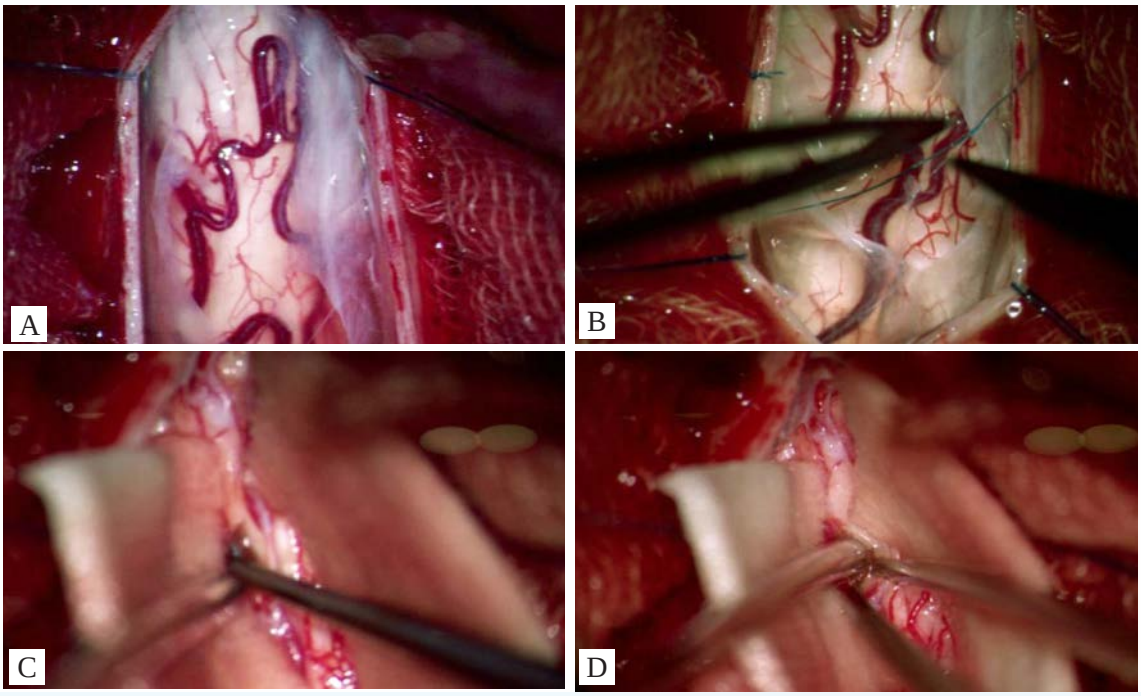

Figure 2. *Intraoperative photos:* ***a)*** *opening the dura mater;* ***b)*** *opening and fixing the arachnoid membrane to dura mater;* ***c)*** *longitudinal mielotomy along the posterior median fissure. Cyst wall is seen at the end of the dissector;* ***d)*** *evacuation of cyst contents by aspirator.*

Table 1.

The significance of somatosensory evoked potentials before and after the intervention

Stimulated nerve	Latency (*mA*)		Amplitude (*mA*)	
	Baseline	Post-operative	Baseline	Post-operative
n. tibialis left	38.20	37.00	2.151	2.347
n. tibialis right	40.30	38.15	1.796	1.802
n. medianus left	18.05	18. 60	5.258	5.709
n. medianus right	19.15	22.70	3.523	3.542

stand the next day after the operation. Sutures were removed on postoperative day 10, the wound healed by the first intention. The patient was discharged under the supervision of neurologist on the 10th day with the positive dynamics in neurological status.

The neurological status at discharge: Movements in the cervical spine are slightly painful while bending. First degree of increased paravertebral muscles tone. Reflexes: biceps D = S, triceps D = S, carporadial D = S, knee D = S, Achilles D = S alive. Percussion of the thoracic spine is painless. Lumbar lordosis is smoothed. Movements in the lumbar spine are painless in full range. Lassegue sign: D – 70°, S – 70° No pathological symptoms. Muscle tone in the upper and lower extremities is improved. The strength in hands and feet was 5 points (D = S). There are no sensory

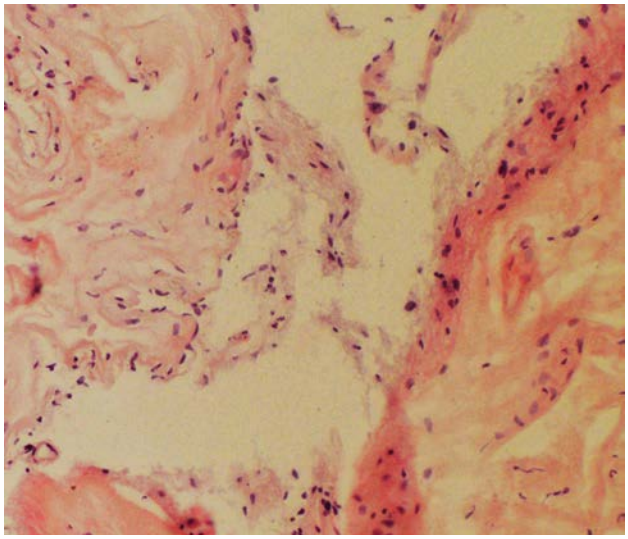

Figure 3. *Morphological image of the cyst wall without epithelial lining. Hematoxylin-eosin, 200 X.*

disturbances. There are no pelvic disturbances.

Patient was recommended to limit physical activity within 1 month postoperatively and was examined by neurosurgeon: full social and physical rehabilitation.

MRI of the cervical spine performed 1 month after surgery (Fig. 4) showed retained intramedullary parasagittal left oval cyst with clear, smooth contours and slightly pronounced perifocal edema at the level of C_5-C_6 vertebras. Intramedullary focus is moderately hyperintense on T2 and isointensive on T1 images of irregularly rounded shape with a diameter of 4.1 *mm* at the C_7 level.

Discussion

Aspects of CNS ontogenesis. Knowledge of sequence and mechanisms of the central nervous system development is important for understanding the ethiology and pathogenesis of ependymal cysts, including those in the spinal cord. The central nervous system undergoes four stages during its development: 1 - the appearance of the neural plate and the formation of the neural tube; 2 - the formation of brain bubbles, separation of front bubbles into pair divisions; 3 - the migration and differentiation of neuronal and glial cells; 4 - sequential pathways of myelination of the brain and spinal cord. Any violation of ontogenesis occurs in a strictly specified time, which is called the teratogenic termination period [*Franceschini P, Worm P, 2014*]. The neurons, neuroglia and ependymal cells are the main three cell types of the central nervous system. In embryogenesis precursors of all those cells are in the epithelium of the neural plate and neural tube [*Nedzved M, 1990*]. These progenitor cells are undifferentiated. Primitive neuroepithelial cells participate in the formation of the walls of cerebral bubbles. The wall of cerebral bubbles consists of three layers: 1) a matrix layer or nucleation consisting of undifferentiated cells; 2) interstitial layer; 3) boundary layer having small cellular elements. While the brain vesicles are enlarging and their walls become thicker, the progenitor cells and primitive neuroepithelial cells become elongating by obtaining radial orientation. The migration of progenitor cells through translocation occurs throughout the central nervous system development [*Morest D, Silver J, 2003*].

Clinical observations from international practice

Publications about intramedullary ependymal cyst are extremely rare. We found 23 cases published in the world literature. There were 14 papers from the 11 authors in PubMed database concerning the localization of intramedullary ependymal cyst. We did not found descriptions of clinical cases with the above pathology in the Russian literature.

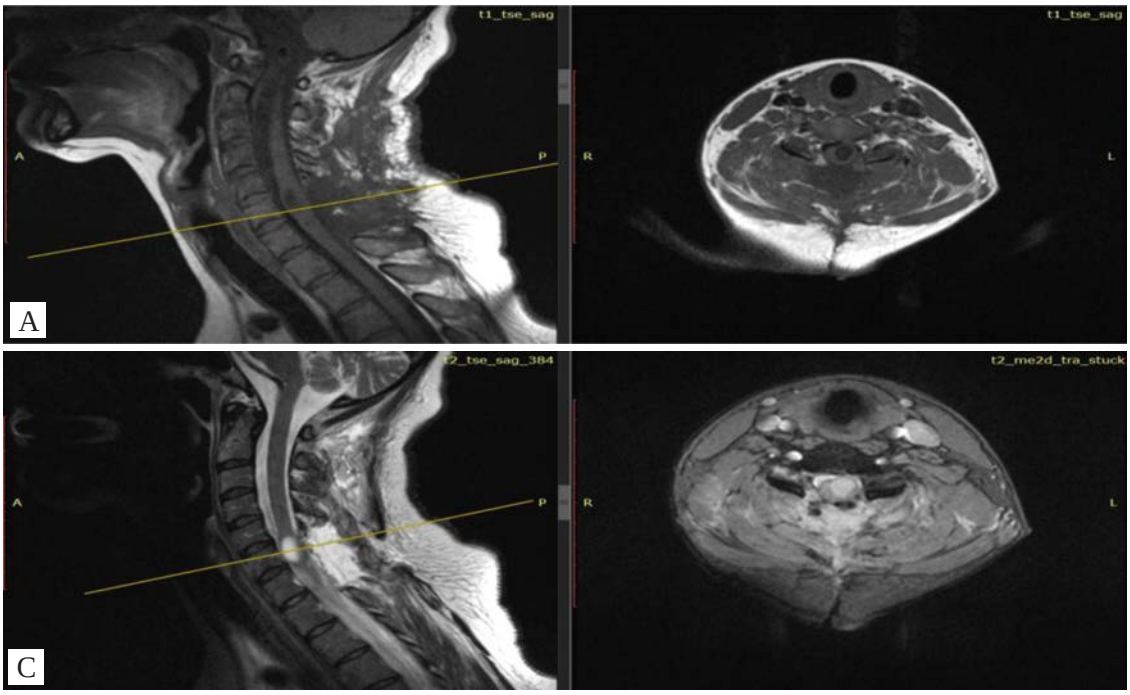

Figure 4. *MRI of the cervical spine after surgery.*

Robertson D.P. and co-authors in 1991 reported 3 cases of intramedullary ependymal cyst: 2 of them were located in the thoracolumbar and 1 in cervical levels. All patients were women aged between 50 and 70, with neurological symptoms of sensory deficit and weakness in limbs. Diagnosis was made with MRI study, during which the contrast did not accumulate in the walls of the cavity or cysts. Surgical treatment was carried out by mielotomy and drainage of cyst cavity. Improvement in neurological status in the postoperative period was reported in all cases. Histological examination in only one case showed monolayer of cubic cells and glial tissue [*Robertson D, Kirkpatrick J, 1991*].

Saito K. and co-authors reported a case about a 44 year old woman with a slowly progressive paresis of the lower extremities with the intramedullary ependymal cyst. Shunt was placed between the cyst and the subarachnoid space as a surgical treatment. Decrease of neurological symptoms in the postoperative period was reported [*Saito K et al., 2005*].

Lalitha A.V. and co-authors published a case of intramedullary ependymal cyst at the level of Th1-Th2 in an 8 year old girl with lower paraparesis who had a good recovery after surgical treatment by drainage [*Lalitha A, Rout P, 2006*].

Radouane B. and co-authors described a case of a 28 years old patient with sciatica and the absence of neurophysiological signs of nerve impulse conduction disorders. MRI of the thoracic spine reviled a cyst at the level of Th11-Th12, which was homogeneous, hypointense on T1 and hyperintense on T2 images, and did not accumulate contrast. The authors excluded disraphism as a potential etiology. A surgery was performed by the imposition of a shunt between the cyst and the subarachnoid space. Clinical improvement was achieved in the postoperative period. Histological examination revealed ependymal cells [*Radouane B et al., 2007*].

Park C.H. and co-authors published data about a 47 year old woman with intramedullary cyst at the level of Th11-Th12 with local pain and paresthesia in the calf region. MRI diagnosed intramedullary cystic lesion in the ventral spinal cord, hypointense on T2 and hypointense on T1-weighted images. The surgical tactics was to remove the cyst wall to form communication between the cyst and the subarachnoid space. Improvement in the neurological status was marked postoperatively. Histological examination revealed cells of simple cubical of epithelia [*Park C et al., 2012*].

A paper by Franceschini and co-authors can be highlighted from the published observations, where they describe a series of 22 registered cases of intramedullary ependymal cyst. Ependymal cysts located in the conus of the spinal cord in 56%, in the cervical region in 22%, in the thoracic spine in 22% [*Franceschini P, Worm P, 2014*]. The authors showed a favorable clinical outcome after cyst decompression. The main surgical treatment was the formation of communication between the cyst and the subarachnoid space.

Calculating the peculiarity of the location and the risks associated with the attempt of cyst wall complete removal in the present clinical observation, intraoperative decision was to perform a microsurgical opening, evacuation of cystic content and subsequent biopsy. The use of intraoperative neurophysiological monitoring hallowed to provide the control and prevention of intra- and postoperative neurological complications, increase the operative measure of radicality in protection of functional state of nervous system's operative segment.

Conclusion

Intramedullary ependymal cyst is a rare and poorly studied pathology. The neurological picture is varied and depends on the location of the cysts (anterior, posterior or lateral parts of the spinal cord). Radical surgical removal is limited due to the high risk of neurological deficit. Histological examination is difficult due to the functional significance of spinal cord tissue, inability to take adequate amounts of material for analysis, and the results of obtained specimens do not always allow to establish organ affiliation of the tissue.

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