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CLINICAL CHARACTERISTICS OF INTRAMEDULLARY SPINAL CORD TUMORS OF VARIOUS HISTOLOGICAL STRUCTURES

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Abstract.
Intramedullary spinal cord tumors (IMSCT) make up from 2 to 4% of all tumors of the central nervous system. These neoplasms "traditionally" have a worse prognosis than extramedullary neoplasms. It is known that 90% of IMSCT are represented by low grade gliomas, which can be successfully removed. Ependymomas (63-65%) and astrocytomas (24.5–30%) constitute the main group of intramedullary spinal cord tumors, glioblastomas (7.5%), oligodendrogliomas (3%) and other tumors (2%) are much less common. The problem of timely detection of IMSCT remains relevant. Unlike brain tumors, these tumors are mostly benign. The clinical course of these tumors is most often acute. By the time obvious neurological signs appear, neurological disorders are already irreversible.
Keywords: spinal cord, intramedullary tumors (IMSCT), histological structure, literature review.

In 1765, Le Cat first reported on the surgical treatment of spinal cord tumors. In 1888, Macewen successfully removed a spinal cord tumor for the first time, and in 1821, Gerutti described intramedullary spinal cord tumors. For a long time, it was believed that the English surgeon V. Gorsley carried out the first surgical intervention for an extramedullary tumor in 1887. However, as S.S. Bryusov (1953) established, priority belongs to the Russian surgeon A.D. Kney, who a few months before Gorsley removed an extramedullary tumor like an hourglass with a favorable outcome. In 1904, for the first time in Russia, Kron diagnosed a spinal cord tumor. In 1905, Cushing performed myelotomy to remove the intramedullary spinal cord tumor in its inoperable course.

Spinal tumor surgery is one of the challenging areas in neurosurgery. Diagnosis of spinal cord tumors in case of their acute manifestation by clinical manifestations can be extremely difficult due to the similarity of the clinical picture with other diseases. Surgical treatment of spinal tumors is always accompanied by a high risk of damage to neural structures or impaired blood supply to the spinal cord, which, in turn, can lead to deep disability of the patient. In this regard, it is obvious that the problem of the pathogenesis of diagnosis and operative treatment of spinal cord tumors is an urgent task at the current stage of the development of neurosurgery and neurology. Despite a huge amount of scientific work, it is not always possible to make an adequate decision on operability in the pathology of the spinal cord and spine [1, 2, 3].

The ratio of spinal and cerebral tumors is approximately 1:6. Among the primary tumors of the spinal cord, extramedullary predominates; of which subdural ones occur about 2.5 times more often than epidural ones. Intramedullary tumors occur in 10-20% of cases of primary tumors of the central nervous system [4, 5, 6]. In childhood and young age, this figure reaches 35% [7].

The bulk of these tumors are up to 95% represented by glial series tumors.

Table 1

Localization and pathomorphological characterization of intradural spinal cord tumors

Localization of tumors	Number of patients (n = 196)
Cervical part	68 (34,7 %)
Thoracicpart	31 (15,8 %)
Lumbarpart	97 (49,5 %)
Pathomorphological characterization of tumors	
Shwannoma	53 (27,0 %)
Meningioma GI GII GIII	7 (3,5 %) 4 (2,0 %) 2 (1,0 %) 1 (0,5 %)
Ependimoma GI GII GIII	46 (23,4 %) 42 (21,4 %) 3 (1,5 %) 1 (0,5 %)
Intradural metastases	6 (3,0 %)
Astrocytic tumors GI GII GIII GIV	36 (18,3 %) 18 (9,1 %) 12 (6,1 %) 5 (2,5 %) 1 (0,5 %)
Hemangioblastomas	19 (9,7 %) 7 (3,57 %)
Dermoid cysts	3 (1,5 %) 8 (4,0 %)
Neurofibromas	5 (2,5 %)
Hemangioma	6 (3,0 %)

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Ependymoma is more often a benign neuroectodermal tumor emanating from the cells of the ependyma lining the ventricles of the brain and the central canal of the spinal cord. Combinations of ependymoma with type I Turko syndrome (multiple endocrine dysplasia syndrome) and type II neurofibromatosis are described. The source of spinal ependyma growth is the cells of the ependymary lining of the central canal of the spinal cord, the accumulation of ependymal cells in the terminal filament of the spinal cord [8, 9, 10]. Ependymomas are encapsulated neoplasms, usually with well-marked edges. They account for 4-6% of primary tumors of the central nervous system, one third of them are intramedullary. According to the WHO pathomorphological classification (1999), the pathohistological markers of these tumors are typical perivascular pseudorosettes. Ependimomas have specific pathohistological features, which are that necrosis and hemorrhage often found in such a tumor are not signs of malignancy, only the presence of frequent mitoses and vascular proliferation speaks in favor of anaplastic (or grade-3) ependimoma.

Astrocytomas are more common in young and middle-aged people. Astrocytoma makes up 24-30% of intramedullary neoplasms. About 75% are benign, 25% are malignant. In adults, it occurs more often in the thoracic part of the SC, followed by the cervical level. Several segments are affected, sometimes the entire length of the SC. About 1/3 of astrocytes contain different cyst sizes [11-13]. At the same time, the malignant nature of astrocytes increases. Unlike ependymoma, intramedullary cysts are often found in spinal cord astrocytomas. If by nature intramedullary tumors are more often benign and slowly growing, then by the nature of growth and location they are the least favorable in terms of the possibility of their surgical removal [14].

Dermoid and epidermoid - are rare in childhood. Slight predominance in ♀. Rarely found in the cervical and upper chest, often in the cone region. Usually intradural extramedullary, but in the cone/ conical tail region there may be an intramedullary component [15].

Atypical teratoid-rhabdoid tumor (ATRT) of the central nervous system is an aggressive malignant tumor that occurs in children mainly under the age of 3 years. It is characterized by the deletion and/or mutation of the SMARCB1/INI1 gene, which is located on the long arm of chromosome 22q11, resulting in the loss of nuclear expression of protein INI1, which can be detected by immunohistochemical examination with anti-INI1 antibody. These tumors are diagnosed mainly in the brain, while localization in the spinal cord is extremely rare, with single publications of clinical observations. Among primary CNS tumors in children, ATRT accounts for 1.6%. In most cases, ATRTs are localized in the cerebellar hemisphere, but are also found in the suprasellar, pineal, temporal, frontal, parietal and occipital regions. At the same time, it should be noted that infratentorial localization is characteristic for young children under 3 years of age, and supratentorial localization for children over 6 years of age [19]. ATRT is extremely rare in the spinal cord (4.6% of all ATRT) [16-17].

Extramedullary-intradural tumors account for 70% of all spinal cord tumors, of which neurinomas and meningiomas occur in 25% of cases. Meningiomas account for 20% of all central nervous system tumors. The average age of patients is 45-61 year; the ratio of women to men is 1.8:1. According to modern histological classifications, meningiomas are divided by the degree of malignancy into three groups: typical (benign), atypical (semi-high-quality), anaplastic (malignant). Anaplastic forms capable of metastasis are found in 1.7% of observations [18].

Multiple meningiomas account for - 8% of their total. Spinal meningiomas account for 1.2% of total meningiomas and 15-30% of all primary spinal tumors. Most spinal meningiomas belong to the group of intradural extramedullary tumors; however, there are tumors with extradural growth [18].

A small number of ectopic intramedullary meningiomas have been described in the literature. There are cases of multiple spinal meningiomas and combinations of spinal meningiomas with cranial. Histologically, among spinal meningiomas, psammomatous forms are often found to be relatively rare for cranial tumors. Spinal meningiomas are often detected in the elderly, in patients over 60 they make up 60% of all primary spinal tumors. Most meningiomas are found in the thoracic spine; in the lumbar spine are extremely rare [19].

Neurinomas of the roots of the spinal nerves are more often observed in the cervical and thoracic parts than in the lumbar. Neurinomas have a rounded, ovoid shape, can be in the form of an hourglass (dumbbell-shaped), reaching an average value of 2-3 cm (in the area of the horsetail and

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when entering the paravertebral region, they can reach large sizes). Developing from sensitive roots, they are located on the posterior lateral surface of the spinal cord. As a rule, neurinomas are encapsulated and well delimited from neighboring structures by neoplasms, they can contain cysts. Spinal cord root neurinomas can be combined with neurofibromatosis. In 15-25% of cases, neurinomas through the intervertebral opening can spread extradurally - the so-called hourglass-type neurinoma, especially for the cervical spine. Peculiarities of extradural tumors in comparison with subdurally located are their histological diversity, large size, prevalence of malignant forms, aggressive growth with pronounced structural change of vertebrae [19].

Metastatic tumors are one of the most common forms of skeletal tumors in general and the spine in particular. Metastases can be single and multiple. For most spinal metastases, a hematogenic dissemination pathway is typical. Especially osteotropic should be considered cancer of the breast, lung, prostate, malignant neoplasms of the kidneys and thyroid glands. At the same time, the localization of the lesion is largely determined by the degree of blood supply to the vertebra. Therefore, the bodies of the vertebrae are more often affected than the arches and spinous processes. In addition, epidural metastasis can occur directly from a metastatic focus or paravertebral tumor, as well as through the cortical bone tissue of vertebral bodies directly into the epidural space. Thoracic is a favorite localization site for metastases [20].

In surgical practice and observations, they use the anatomical classification:

I. Subdural tumors:

- intramedullary
- extramedullary, emanating from the inner leaf of the dura mater, the dental ligament, the pial shell, the intradural part of the cerebrospinal root.

II. Extradural tumors:

- primary of vertebrae, periosteum, ligaments, cartilage, outer leaf of the dura mater;
- secondary metastatic.

Diagnostics.

CT: the density of some IMSCT increases during a contrast study. Contrast CT makes it possible to distinguish IMSCT from intradural-extramedullary tumors; however, it is not able to distinguish between different types of IMSCT.

A new step in the field of improving diagnosis, and therefore treatment of patients with spinal cord diseases, was the use in neurosurgical practice of a non-invasive method of research - magnetic resonance imaging (MRI). Currently, MRI has come first in the diagnosis of most spinal cord and spinal cord diseases, sidelining methods such as myelography and CT myelography. Radiological diagnosis of intramedullary tumors is quite widespread, but most methods that can adequately judge the presence of a tumor lesion require a lot of attention. MRI is one of the most sensitive methods in determining the change in spinal cord size in the presence of an intramedullary tumor, the growth of which is usually accompanied by a thickening of the spinal cord. To determine the localization of the tumor and its relative dimensions, T1 - weighted tomograms are considered the most informative. At the same time, intramedullary tumors have their own features that allow them to be distinguished from tumors of another localization. In T2 weighted images, intramedullary tumors are characterized by an increase in signal (to one degree or another) compared to normal tissues. Moreover, signal amplification can be heterogeneous [21-23].

The true boundaries of the tumor in this mode are also almost impossible to determine, since the peritumoral edema present around has an increased signal and can merge with the signal from the tumor. MRI myelography detects spinal cord expansion and narrowing of subarachnoid space. Without intravenous administration of contrast agent, MRI is not informative, since isodense tumor tissue is difficult to differentiate from spinal cord. In rare cases, centers of increased density, intratumoral hemorrhage or petrificates can be determined. Spondylography is of little importance in the diagnosis of astrocytes, since radiologically visible bone changes are less common than in ependymoma. The use of MRI should be considered the most appropriate. The leading diagnostic method is a CT study. A characteristic symptom for the chordoma is calcification along the periphery of the tumor, due to the presence of necrotized bone and sites. Chordoma is characterized by osteolytic focus of destruction, without clear contours, thinning and destruction of crust layer with presence of paravertebral component, clearly delimited from adjacent organs and soft tissues [24].

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CONCLUSION

Thus, IMSCT s are a relatively rare pathology, but cause great social and economic damage to society, due to the fact that they affect children and adults of working age. With the introduction of modern neuroimaging research methods (MRI, spiral CT, PET) into neurosurgical practice, the detectability of IMTSC has increased significantly. However, the problem of timely identification of IMTSC remains urgent. Unlike brain tumors, these tumors are in most cases benign. The clinical course of these tumors is most often acute. Further histochemical and immunological studies will establish the nature of these neoplasms.

CONFLICT OF INTEREST

The author declares no conflict of interest. Financing. The study was performed without external funding. Compliance with patient rights and principles of bioethics. All patients gave written informed consent to participate in the study.

REFERENCES

1. *Поляков Ю.Ю., Олюшин В.Е., Гуляев Д.А., Тастанбеков М.М. Тактика и алгоритм лечения спинальных опухолей. М.: Материалы 5-й Межрегиональной научно-практической конференции «Актуальные вопросы неврологии», с.99-109, 2008г. [Polyakov Yu.Yu., Olyushin V.E., Gulyaev D.A., Tastanbekov M.M. Tactics and algorithm for the treatment of spinal tumors. M .: Materials of the 5th Interregional Scientific and Practical Conference "Actual Issues of Neurology" P.99-109, 2008. (In Russ.).]*
2. *Бывальцев В.А., Степанов И.А. Особенности и факторы риска рецидивирования интрадуральных опухолей спинного мозга. Сибирский онкологический журнал. 2019; 18(1): 21-29. [Byvaltsev V.A., Stepanov I.A. Features and risk factors for recurrence intradural tumors of the spinal cord. Siberian journal of oncology. 2019; 18(1): 21-29. (In Russ.).]*
3. *Евзиков Г.Ю., Крылов В.В., Яхно Н.Н. Хирургическое лечение внутримозговых спинальных опухолей. М., 2006. [Evzikov G.Yu., Krylov V.V., Yakhno N.N. Surgical treatment of intracerebral spinal tumors. Moscow, 2006. (In Russ.).]*
4. *Харитонова К.И., Окладников Г.И. Патогенез и диагностика опухолей спинного мозга. Новосибирск, 1987. [Kharitonova K.I., Okladnikov G.I. Pathogenesis and diagnosis of spinal cord tumors. Novosibirsk, 1987. (In Russ.).]*
5. *McCormick PC, Stein BM. Spinal Cord Tumors in Adults. In: Youmans J, ed. Neurological Surgery. 4th ed. Philadelphia, PA: WB Saunders; 2003: 3102.*
6. *McCormick PC, Torres R, Post KD, Stein BM. Intramedullary ependymoma of The Spinal Cord. J Neurosurg. 1990; 72:523–532.*
7. *Constantini S, Miller DC, Allen JC, Rorke LB, Freed D, Epstein FJ. Radical Excision of Intramedullary Spinal Cord Tumors: Surgical Morbidity and Long-Term Follow-Up Evaluation in 164 Children and Young Adults. J Neurosurg. 2000; 93(2 Suppl): 183–193.*
8. *Gregorios JB, Green B, Page L, Thomsen S, Monforte H. Spinal Cord Tumors Presenting with Neural Tube Defects. Neurosurgery. 1986; 19:962–966.*
9. *Lynch J, Kelly N, Fitzpatrick B, Regan P. A Sacrococcygeal Extraspinal Ependymoma in a 67-year-old man: a Case Report and Review of the Literature. Br J Plast Surg. 2002; 55:80–82. doi: 10.1054/bjps.2001.3724.*
10. *Morantz RA. Ectopic Ependymoma of the Sacrococcygeal Region. In: Doty JR, Rengachary SS, eds. Surgical Disorders of the Sacrum. New York: Thieme Medical Publishers, 1992: 177–179.*
11. *Желудкова О.Г., Коршунов А.Г., Горбатых С.В., и др. Злокачественные тератоид-рабдоидные опухоли центральной нервной системы у детей // Вопросы гематологии/онкологии и иммунопатологии в педиатрии. - 2003. - Т.2. - №3 - С. 32-39. [Zheludkova O.G., Korshunov A.G., Gorbatykh S.V., et al. Malignant teratoid-rhabdoid tumours of the central nervous system in children. Pediatric haematology/oncology and immunopathology. 2003;2 (3):32–39. (In Russ.).]*
12. *Ольхова Л.В., Желудкова О.Г., Рыжова М.В., Кушель Ю.В., Крянев А.М., Озерова В.И. Атипичная тератоидно-рабдоидная опухоль спинного мозга: клинический случай. Онкопедиатрия / 2018 / том 5 / № 4. Стр.: 268-277 [Olkhova L.V., Zheludkova O.G., Ryzhova M.V., Kushel Yu.V., Kryanev A.M., Ozerov V.I. Atypical Teratoid-Rhabdoid Tumor of the Spinal Cord: A Clinical Case. Oncopediatry / 2018 / Volume 5 / No. 4. Pages: 268-277. (In Russ.).]*
13. *M.Greenberg. Handbook of neurosurgery. 2006, p. 508-515.*
14. *G. Fisher, J. Brotchi. Intramedullary spinal cord tumors. 1996, 115 p.*
15. *McCormick PC, Stein BM. Spinal Cord Tumors in Adults. In: Youmans JR, ed. Neurological Surgery. Philadelphia, PA: WB Saunders; 1996; 3102-3122.*
16. *Ostrom QT, Chen Y, M de Blank P, et al. The Descriptive Epidemiology of Atypical Teratoid/Rhabdoid Tumors in the United States, 2001–2010. Neuro Oncol. 2014;16(10):1392–1399. doi: 10.1093/neuonc/nou090.*
17. *Hilden JM, Meerbaum S, Burger P, et al. Central Nervous System Atypical Teratoid/Rhabdoid Tumor: Results of Therapy in Children Enrolled in a Registry. J Clin Oncol. 2004;22(14):2877–2884. doi: 10.1200/JCO.2004.07.073.*
18. *Aryan HE, Imbesi SG et al. Intramedullary Spinal Cord Astrolipoma: Case Report. // Neurosurgery: October 2003 - Volume 53 - Issue 4 - pp 985-988.*
19. *Пастушин А.И. Хирургическое лечение больных старших возрастных групп при компрессии спинного мозга менингиомами. Нейрохирургия, 1983. Вып. 16. С. 31-34. [Pastushin A.I. Surgical Treatment of Patients of Older Age Groups with Spinal Cord Compression by Meningiomas. Neurosurgery, 1983. Issue. 16. S. 31-34.*
20. *Greenberg M.S. Handbook of Neurosurgery. Florida: Lakeland, 2010.*
21. *Mahmood A, Cacammo DV, Tomechek FJ. Atypical and Malignant Meningiomas: Clinicopathological Review // Neurosurgery. 1993. V. 33. P. 955-963.*
22. *Стародубцев А.И. Ранняя диагностика и хирургическое лечение опухолей спинного мозга и позвоночника. Дис. … д- ра мед наук М., 1985. [Starodubtsev A.I. Early Diagnosis and Surgical Treatment of Tumors of the Spinal Cord and Spine Dis... Doctor of Medical Sciences M., 1985. (In Russ.).]*
23. *Харитонова К.И., Окладников Г.И. Патогенез и диагностика опухолей спинного мозга. Новосибирск, 1987. [Kharitonova K.I., Okladnikov G.I. Pathogenesis and Diagnosis of Tumors of the Spinal Cord, Novosibirsk, 1987. (In Russ.).]*
24. *Karikari I.O, Nimjee S.M. et. al. Impact of Tumor Histology on Resectability and Neurological Outcome in Primary Intramedullary Spinal Cord Tumors: A Single-Center Experience with 102 Patients. // Neurosurgery: January 2011 - Volume 68 - Issue 1- pp 188-197.*

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