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TREATMENT OF INTRAPARENCHYMAL CHORDOID MENINGIOMA: A CLINICAL CASE

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ABSTRACT

Relevance: *Meningioma has many forms, each with a specific histological structure and treatment tactics. Meningiomas are the most common intracranial neoplasms, most of which have an extracerebral location, and their intraparenchymal location is rare.*

The study aimed to *describe a case of treatment of intraparenchymal chordoid meningioma.*

Methods: *The article describes a case of treatment of intraparenchymal chordoid meningioma, which is rare in localization, structure, and possible combination with other systemic diseases.*

Results: *Patient B. was admitted to the clinic for surgical treatment for an intracranial neoplasm; the main complaints were the presence of seizures and their transition to epileptic status. The tumor was removed completely, after which it was sent for further histological and immunohistochemical examination. The examination of micro slides revealed the presence of chordoid meningioma in the patient. In the postoperative period, as well as at control examination after 3 months, the patient showed a decrease in the number of seizures and no transition to epileptic status. MRI control showed no relapse of tumor growth.*

Conclusion: *WHO classifies chordoid meningioma as a rare grade II meningioma. Chordoid meningioma often requires further radiation therapy, especially with partial or subtotal removal, due to a slightly higher risk of relapse compared to grade I meningiomas. This disease can occur independently or in association with Castleman syndrome and can be asymptomatic for a long time; this requires further follow-up of the patient.*

Keywords: *meningioma, chordoid meningioma, intraparenchymal meningioma, Castleman syndrome.*

Introduction: The 2021 WHO Classification of CNS Tumors edition 5 classifies chordoid meningioma as a grade 2 malignancy due to its high recurrence rate [1]. At the same time, this type of meningioma is scarce, and its incidence is <1% of all meningiomas [2, 3]. The term "chordoid meningioma" was first mentioned by Kepes et al. when they described its association with Castleman syndrome in 1988 [4]. However, it is often not associated with systemic diseases [5].

Histologically, this type of meningioma is characterized by spindle-shaped or flattened cells with eosinophilic cytoplasm located in lobules and surrounded by a basophilic myxoid stroma rich in mucin, which makes it possible to stain glass preparations with PAS-iodic acid and alcian blue (AB) [6, 7]. It should be highlighted that the imaging methods do not show significant differences in signal characteristics or the contrast enhancement between chordoid meningioma and other types of meningiomas [8], except for the ADC mode in magnetic resonance imaging (MRI), which has a slightly higher diffusion coefficient [9, 10].

The study aimed to describe a case of treatment of intraparenchymal chordoid meningioma.

Materials and Methods: The article describes the treatment of intraparenchymal chordoid meningioma, which is rare in its localization, structure, and possible combination with other systemic diseases. The patient has provided a signed informed consent for manipulations and the use of his treatment results for scientific studies and other educational, scientific, and advertising purposes.

Clinical case:

Patient information: Male B., 25 years old. According to the patient, in 2017, he developed generalized tonic-clonic seizures, during which he remained conscious. The seizures began with numbness in the left arm and were also observed during sleep. A few days before the onset of these complaints, the patient suffered a cerebral concussion and a soft tissue bruise on the hairy part of the head. The peculiarities of the patient's history are that he repeatedly participated in conflicts and often received blows to the head. However, the role of this factor in developing the neoplasm is questionable. Despite several studies reporting a higher incidence of brain neoplasms in people who have suffered a traumatic brain injury, a systematic review conducted by Darsh S. et al. showed no statistically significant association between traumatic brain injury and brain neoplasms [11].

Clinical findings: The brain CT and MRI revealed "Meningiomatosis. Multiple small neoplasms of the anterior, middle third of the falx. Calcified neoplasm of the right-side parietal lobe."

The patient refused surgical treatment for conservative measures. He was taking Finlepsin 200 mg BID. Since

March 2023, his seizures have become more frequent and often transited to status epilepticus.

Diagnostics: In March 2023, the patient underwent brain computed tomography (CT). The CT scan in axial and frontal anatomical projections (APR) showed a hyperdense neoplasm of the parietal region rightward with a size of 1.36*1.24*1.4 cm. Numerous small hyperdense neoplasms of the anterior middle third of the falx, presumably calcifications (Figure 1), were detected. Over the past month, the patient noted a deterioration of his condition in the form of a seizure syndrome with a transition to epi-status, which was difficult to stop with medications. The neurosurgeons consulted the patient at the National Center of Neurosurgery (Astana, Kazakhstan). Considering the nature of the pathological process and ineffective conservative treatment, he was recommended surgical treatment in the amount of "Craniotomy of the parietal bone on the right side. Microsurgical removal of a neoplasm of the parietal lobe rightward with the application of the neuro-navigation and neuro-monitoring" (Figure 2).

Treatment: Surgical treatment was conducted in a standard mode; the tumor was completely removed. Unique anatomical peculiarities included a marked adhesion of the dura mater to the bone. The CT control was performed the next day (Figure 3).

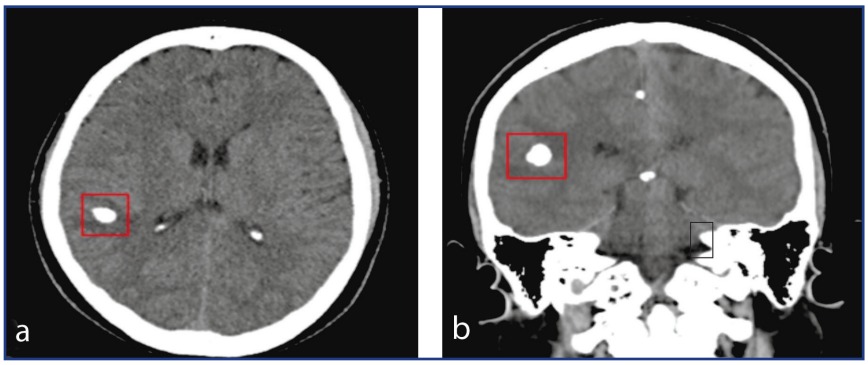

Figure 1 – A preoperative brain CT scan: a – axial projection, b – frontal projection

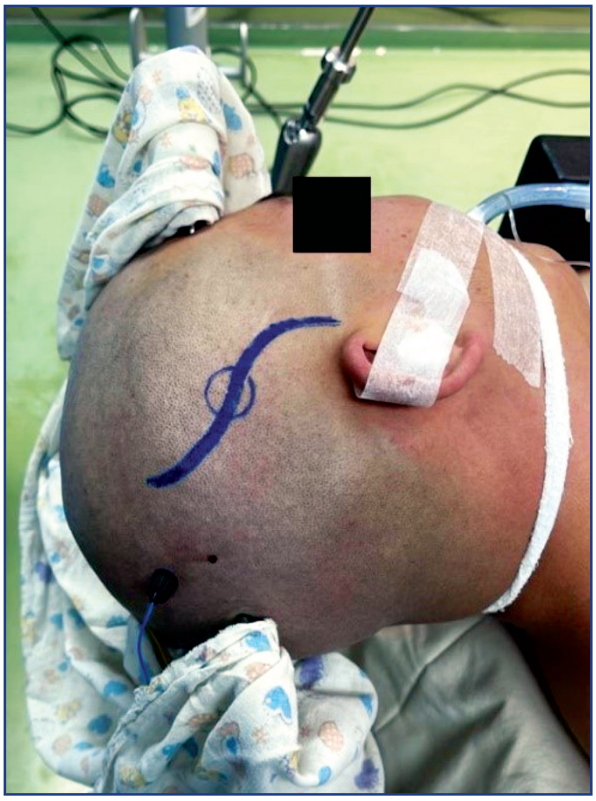

Figure 2 – Planned incision and projective location of the neoplasm

Fragments of the removed neoplasm were sent for examination to the Pathology department, where they were fixed in 10% neutral buffered formalin during the day. After traditional processing, the preparations were stained with hematoxylin and eosin. The immunohistochemical staining was automatically performed us-

ing the "Ventana BenchMark XT" immunostainer with "Ventana Roche" antibodies. The microscopic examination was performed using an OLYMPUS microscope at a total magnification of ×100 and ×200.

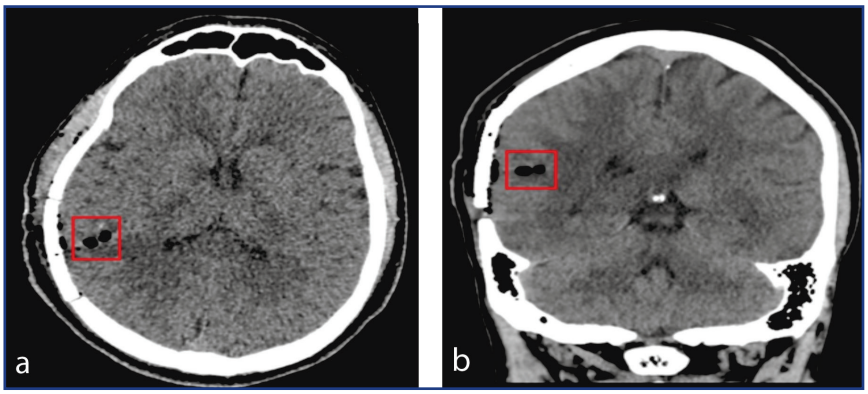

Figure 3 – A postoperative brain CT: a – axial projection, b – frontal projection

In preparations, the tumor tissue was represented by nest-focal growths of cartilage tissue, between which the fibroblast-like cells and thin-walled vessels were visible (Figure 4). Immunohistochemically, the tumor cells were focally expressed in the epithelial membrane antigen (EMA) and vimentin and, in isolated cases, progesterone (Figures 5 & 6).

Based on histopathological and immunohistochemical examinations, the patient was diagnosed with a "Chordoid meningioma, WHO grade 2, ICD-O code 9538/1."

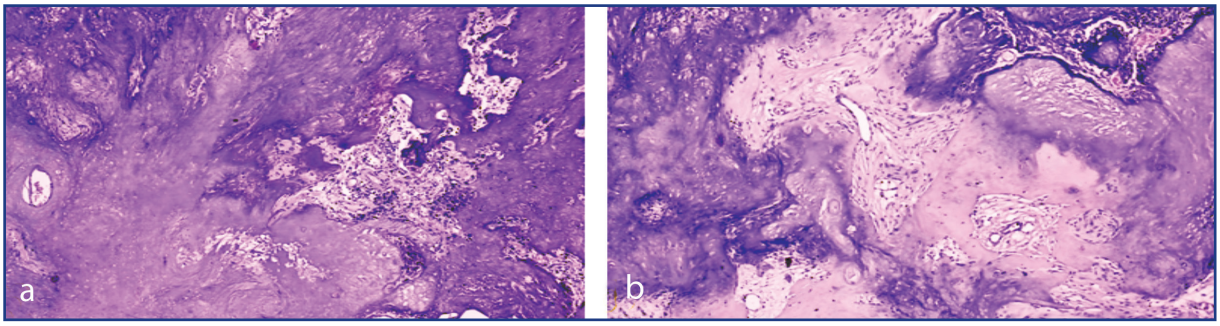

Figure 4 – Chordoid meningioma: a – magnification ×100, b – magnification ×200. Hematoxylin and eosin staining. Used microscope OLYMPUS (Japan)

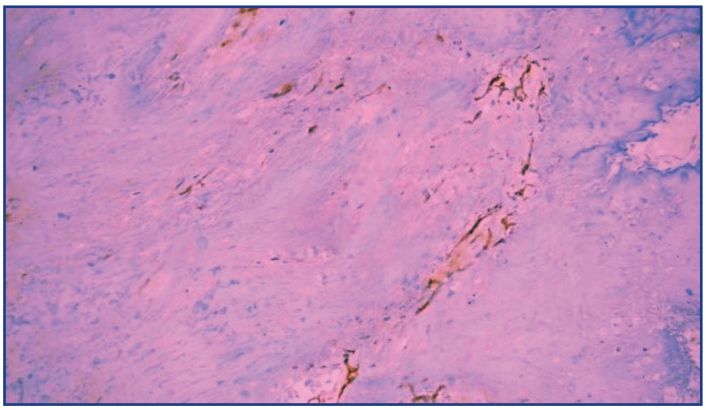

Figure 5 – Immunohistochemical examination, magnification ×100. Focal-positive reaction to the EMA. Used microscope OLYMPUS (Japan)

Post-surgery, the pain syndrome, dizziness, and unsteadiness were decreasing. The wound has healed by primary intention. The patient was discharged from the hospital in satisfactory condition.

A follow-up examination after 3 months showed a reduction in the number of seizures and no transition of seizures to status epilepticus in the observation period. The patient continues the intake of anticonvulsants recommended by the supervising neurologist; their dosage has been reduced. Figure 7 presents a contrasted control brain MRI in the T1-weighted mode.

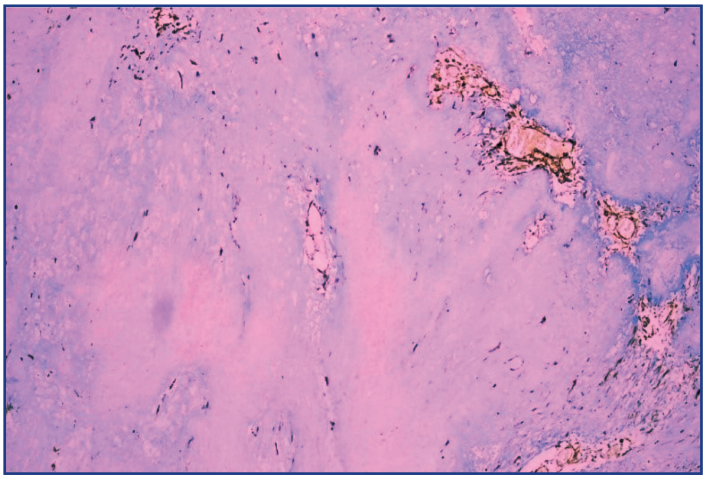

Figure 6 – Immunohistochemical examination, magnification ×100.
Focal positive reaction to vimentin.
Used microscope OLYMPUS (Japan)

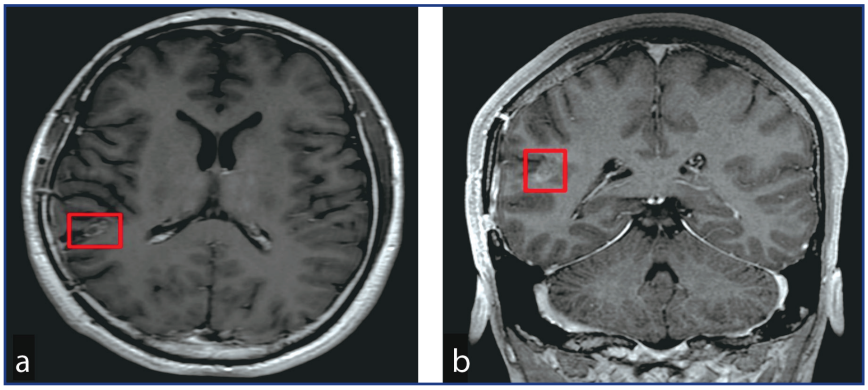

Figure 7 – Follow-up brain MRI, 3 months after surgery: a – axial projection,
b – frontal projection.

Table 1 presents the timeline of the described case of treatment of intraparenchymal chordoid meningioma.

Table 1 – Clinical Case Timeline

Date	Symptoms (seizures)	Method of diagnostics and treatment
2017	Onset of symptoms	Drug therapy (prescription of anti-convulsants)
March 2023	Aggravation (increased number of seizures and frequent transi-tions to status epileptic)	Surgical treatment (June 2023)
Postoperative period (from June 2023)	Improvement (reduction of sei-zure frequency)	Drug therapy (reduction of anti-convulsant dosage)

Discussion: Chordoid meningioma has a wide range of histological differential diagnoses. IHC analysis is a significant factor that plays an important role in differentiating these tumors [12].

Differential diagnostics shall include chordoma, chordoid glioma, myxopapillary ependymoma, and chondrosarcoma. In IHC analysis, this tumor is reactive to EMA and vimentin; in most cases, it is positive for D2-40 and GFAP markers, with an adverse reaction to the S-100 protein and cytokeratin [13, 14].

Patients diagnosed with chordoid meningioma, alone or in combination with other systemic diseases, such as Castleman syndrome, require a detailed examination and additional checkup of other body systems to exclude possible concomitant autoimmune and other cancer diseases. They often need further radiation therapy due to a slightly higher risk of recurrence compared to grade 1 meningiomas, especially in the case of subtotal resection [15].

An intraparenchymal location of the neoplasm is also notable. It is quite rare for primary meningiomas since most meningiomas adjoin the dura mater [16].

Conclusion: Therefore, patients with a confirmed chordoid meningioma require longer monitoring by an oncologist due to a slightly higher risk of recurrence compared to grade I meningiomas. Notably, this disease can occur independently or in association with Castleman syndrome. Also, this disease can be asymptomatic for a long time, which requires further follow-up care due to a risk

of developing concomitant oncological conditions. The patients require a thorough checkup with any symptoms of concomitant diseases. The importance of accurate and timely diagnosis underscores the critical role of diagnostics in guiding patient management and preventing potentially severe complications.

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АҢДАТПА

ИНТРАПАРЕНХИМАЛДЫҚ ХОРДОИДТЫ МЕНИНГОМАДЫ ЕМДЕУ: КЛИНИКАЛЫҚ ЖАҒДАЙ

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Өзектілігі: *Менингиомалардың көптеген формалары бар, олардың әрқайсысының гистологиялық құрылымы жағынан да, емдеу тактикасы жағынан да өзіндік ерекшеліктері бар. Менингиомалар ең жиі кездесетін интракраниальды өсінділер болып табылады, олардың көпшілігі экстрацеребральды орналасуға ие және олардың интрапаренхималық орналасуы сирек кездеседі.*

Зерттеудің мақсаты: *Интрапаренхималық хордоидты менингиоманы емдеу жағдайының сипаттамасы.*

Әдістері: *Мақалада локализациясы, құрылымы және басқа жүйелік аурулармен үйлесуі сирек кездесетін интрапаренхималық хордоидты менингиоманы емдеу жағдайы сипатталған.*

Нәтижелер: *Науқас Б.клиникаға интракраниальды неоплазманы жедел емдеу үшін кірді, негізгі шағымдар ұстамалардың болуы, олардың эпилепсиялық мәртебеге ауысуы болды. Ісік толығымен алынып тасталды, содан кейін ол одан әрі гистологиялық және иммуногистохимиялық зерттеуге жіберілді. Микропрепараттарды зерттеу пациентте хордоидты менингиоманың болуын анықтады. Операциядан кейінгі кезеңде, сондай-ақ 3 айдан кейін бақылау тексеруінің мәліметтері бойынша, науқаста ұстамалар санының төмендеуі байқалды, ұстамалардың эпилепсиялық мәртебеге ауысуы болмады. Жүргізілген МРТ бақылауы неоплазманың өсуінің қайталануының жоқтығын көрсетті.*

Қорытынды: *Хордоидты менингиома ДДҰ классификациясына сәйкес II дәрежелі менингиомалардың сирек кездесетін кіші түрі болып табылады және көбінесе сәулелік терапияны қажет етеді, бұл әсіресе ішінара және субтотальды алып тастауда, I дәрежелі менингиомалармен салыстырғанда қайталану қаупінің сәл жоғары болуына байланысты. Сондай-ақ, аурудың жеке формалары да, Кастельман синдромымен байланысты да бар екенін атап өткен жөн, ал асимптоматикалық курс өте ұзақ уақыт өтуі мүмкін, бұл пациенттің диспансерлік бақылауда болуын талап етеді.*

Түйінді сөздер: *менингиома, хордоидты менингиома, интрапаренхималық менингиома, Кастлман синдромы.*

АННОТАЦИЯ

ЛЕЧЕНИЕ ИНТРАПАРЕНХИМАЛЬНОЙ ХОРДОИДНОЙ МЕНИНГИОГМЫ: СЛУЧАЙ ИЗ ПРАКТИКИ

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Актуальность: *Существует множество форм менингиом, каждая из которых имеет свои особенности, как по гистологической структуре, так и в плане тактики лечения. Менингиомы являются наиболее часто встречаемыми внутричерепными новообразова-*

ниями, большинство из которых имеют экстрацеребральное расположение, а интрапаренхимальное их расположение является редким. Актуальность данной публикации состоит в описании редкого случая лечения интрапаренхимальной хордоидной менингиомы.

***Цель исследования** – описание случая лечения интрапаренхимальной хордоидной менингиомы.*

***Методы:** В статье описан случай лечения интрапаренхимальной хордоидной менингиомы, которая является редкой по локализации, структуре и возможному сочетанию с другими системными заболеваниями.*

***Результаты:** Пациент Б. был госпитализирован в клинику с целью оперативного лечения интракраниального новообразования. Основными симптомами были судороги, переходящие в эпилептический статус. Опухоль была удалена тотально, после чего отправлена на дальнейшее гистологическое и иммуногистохимическое исследование. Проведенное исследование микропрепаратов выявило у пациента наличие хордоидной менингиомы.*

В послеоперационном периоде, а также по данным контрольного осмотра через 3 месяца, у пациента отмечено снижение количества судорог, отсутствовал переход судорог в эпилептический статус. Проведенный МРТ-контроль показал отсутствие рецидива роста новообразования.

***Заключение:** Хордоидная менингиома представляет собой редкий подтип менингиом II степени согласно классификации ВОЗ и часто нуждается в дальнейшей лучевой терапии, что особенно актуально при частичном и субтотальном удалении, ввиду чуть более высокого риска рецидива в сравнении с менингиомами I степени. Также стоит выделить то, что существуют как обособленные формы заболевания, так и связанные с синдромом Кастелмана. При этом заболевание может протекать бессимптомно в течение довольно длительного периода времени, что требует в дальнейшем нахождения пациента на диспансерном наблюдении.*

***Ключевые слова:** менингиома, хордоидная менингиома, интрапаренхимальная менингиома, синдром Кастлемана.*

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