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POLYMORPHOUS LOW-GRADE NEUROEPITHELIAL TUMOR OF THE YOUNG (PLNTY) NEW RADIOLOGICAL FEATURES: A CASE REPORT

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A new form of morphologically and molecularly distinguishable epileptogenic neoplasia with characteristic microscopic findings and a distinct DNA methylation signature as well as frequent genetic anomalies, was revealed in 2017; the tumor was called polymorphous low-grade neuroepithelial tumor of the young (PLNTY). Several specific radiological patterns found in PLNTY when compared with the results of a pathomorphological study being useful in differential diagnosis with other epileptogenic tumors were mentioned in certain papers. Our paper is devoted to some particulars of the radiological picture in two children with pharmacoresistant epilepsy who underwent epileptic surgery with histological verification of PLNTY.

KEYWORDS: Polymorphous low-grade neuroepithelial tumor of the young (PLNTY), structural epilepsy, neuroimaging

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ПОЛИМОРФНАЯ НЕЙРОЭПИТЕЛИАЛЬНАЯ ОПУХОЛЬ НИЗКОЙ СТЕПЕНИ ЗЛОКАЧЕСТВЕННОСТИ МОЛОДОГО ВОЗРАСТА (PLNTY), НОВЫЕ РАДИОЛОГИЧЕСКИЕ ОСОБЕННОСТИ: КЛИНИЧЕСКИЙ СЛУЧАЙ

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В 2017 г. обнаружена новая форма морфологически и молекулярно различимой эпилептогенной неоплазии, имеющая характерные микроскопические находки и отчетливую сигнатуру метилирования ДНК, наряду с частыми генетическими аномалиями, получившая название полиморфная нейроэпителиальная опухоль низкой степени злокачественности молодого возраста (PLNTY). В литературе упоминается о нескольких специфических радиологических паттернах, встречающихся у PLNTY, которые при сопоставлении с результатами патоморфологического исследования могут быть полезны при дифференциальной диагностике с другими эпилептогенными опухолями. Мы сообщаем о некоторых особенностях радиологической картины у двух детей с фармакорезистентной эпилепсией, прошедших процедуру эпилептической хирургии с гистологической верификацией PLNTY.

КЛЮЧЕВЫЕ СЛОВА: полиморфная нейроэпителиальная опухоль низкой степени злокачественности молодого возраста (PLNTY), структурная эпилепсия, нейровизуализация

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Introduction. Low-grade tumors associated with long-term epilepsy in children and young patients are allocated to a separate group of Long-Term Epilepsy Associated Tumors (LEAT), which is constantly updated with new morphological elements [1, p. 7]. PLNTY has been described relatively recently by J. Hesse et al. The infiltrative nature of growth with cell components similar to oligodendroglioma, intense CD34 immunopositivity and genetic abnormalities of the proto-oncogene B-Raf (BRAF), or fibroblast growth factor receptors 2 and 3 (FGFR2, FGFR3) have been noted in their paper [2, p. 422]. These data indicate that PLNTY is a separate biological unit in a wider range of low-grade tumors associated with long-term epilepsy, which made it possible to include it in the new classification of CNS tumors by WHO 2021. Distinctive pathological and molecular characteristics were revealed in comparison with other LEAT tumors in the course of the study of PLNTY [2, p. 418]. It is worth noting that only a few publications are devoted to the peculiarities of the radiological picture found in these tumors. At the same time, in addition to the fact that there is a certain pathomorphological and molecular pattern in the new form of the tumor, the authors claim that it may also have specific radiological patterns [3, p. 576].

Some peculiarities of the PLNTY radiological picture on the example of two patients underwent epileptic surgery for pharmacoresistant structural epilepsy are reported in our paper.

Case 1. Patient Kh., female, 11 years old, diagnosed with symptomatic temporal lobe epilepsy and delayed psycho-speech development. Seizures have been observed since the age of 2.5 years. The phenomenology of seizures: tonic adversive and opercular.

The frequency is up to 2 seizures per month, the duration of the seizures reaches up to 2 minutes. From the age of 3, she periodically underwent examination and treatment in the neuropsychiatric department of the Federal Research and Clinical Center for Children and Adolescents in Moscow.

The patient was prescribed antiepileptic therapy, with repeated changes of regimens and dosage adjustments, which led to a decrease in the frequency of seizures to 1 episode in 2–3 months.

During MRI on a tomograph with a low magnetic field induction, a site of a pathological signal was detected in the right temporal lobe. Having based on the presence of transmantle spread of the focus from the cortex to the wall of the lateral ventricle neurosurgeons initially regarded the tumor as focal cortical dysplasia (FCD) type II.

According to computed tomography (CT) data, small calcifications were revealed in the area of question in the right temporal lobe (Fig. 1).

Pre-surgical MRI was performed on a superconductive tomograph with a modification of the scanning protocol for the individual characteristics of the patient with the inclusion of tractography (DTI) and contrast-free MR perfusion (ASL). When comparing the results of dynamic MRI and CT, against the background of the overall stability of the radiological picture, the minor changes in the relaxation characteristics of the previously identified pathological zone in the right frontal lobe and small calcifications in the area of question were noticed. The «salt and pepper» symptom, the «transmantle» sign, and an increase in perfusion in the structure of the focus were clearly identified on the images made by means of the superconductive MR system; this suggested the presence of neoplasm in the epileptogenic substrate structure (Fig. 2).

During the interdisciplinary consultation, the obtained images were correlated with the results of video EEG monitoring, the clinical picture and the seizures phenomenology. Having studied the brain MRI experts came to the conclusion that the child with a pharmacoresistant manifestation of epilepsy suffers the right temporal lobe tumor from the LEAT group. On the base of the data obtained, surgical treatment was recommended. Performed surgery included microsurgical removal of the right temporal lobe tumor with using neurophysiological monitoring.

During the period of postoperative observation of the patient, no seizures were noticed.

Due to the focal resection of the formation, and the presence of residual fragments on postoperative MRI, it was decided to continue antiepileptic therapy in the same manner, with a possible subsequent gradual reduction in dosages.

On evaluating the biopsy (surgical) material, the following conclusion was obtained: «tiny fragments of benign glioma with a large number of calcifications».

In the course of dynamic observation in 2 years after the operation, a resumption of seizures with the same frequency was noted. After the pre-surgical preparation, a second operation was performed with total resection of residual fragments of tumor tissue, the trans-mantle path and the nearby cortex (Fig. 3, *a*).

A medium-cell tumor with diffuse growth type and numerous microcalcinates, consisting of both oligodendroglial and astrocytic cells was revealed during histopathological examination. The immunohistochemical study revealed diffuse expression of GFAP, CD34 by neoplastic cells.

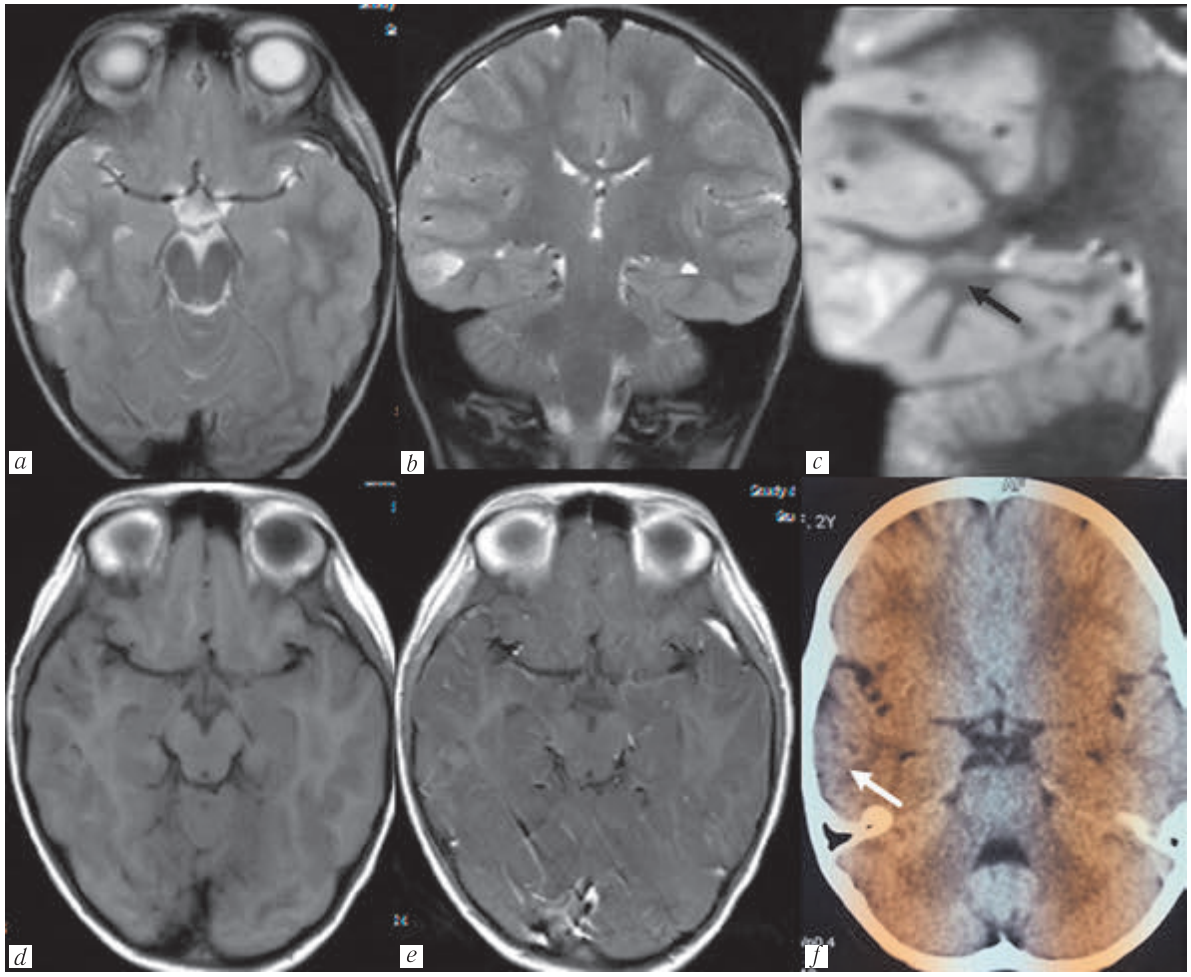

Fig. 1. Patient Kh., female, 7 years old: MRI of 0,4 T. Pulse sequences T2 and STIR show a local signal enhancement site in the posterior parts of the right temporal lobe with the main localization in the cortex, spreading transmantically towards the temporal horn of the right lateral ventricle (black arrow), without signs of perifocal edema and mass effect (*a, b, c*). In the STIR and T1 pulse sequences, the cystic structure of the focus is clearly determined (*d*). The focus does not accumulate a contrast agent (*e*), a few small calcifications are visualized on CT images in the area of question (white arrow) (*f*)

Рис. 1. Пациентка Х., 7 лет: МРТ 0,4 Т и КТ. Импульсные последовательности Т2 и STIR демонстрируют локальный участок повышения сигнала в задних отделах правой височной доли с основной локализацией в коре, распространяющийся трансмантийно в сторону височного рога правого бокового желудочка (черная стрелка), без признаков перифокального отека и масс-эффекта (*a, b, c*). В импульсных последовательностях STIR и Т1 отчетливо определяется кистозная структура очага (*d*). Очаг не накапливает контрастный препарат (*e*), на изображениях КТ в зоне интереса визуализируются немногочисленные мелкие кальцинаты (белая стрелка) (*f*)

Conclusion: The case of the disease corresponds to a polymorphous low-grade neuroepithelial tumor of the young, CNS WHO grade 1 (Fig. 3, *b, c, d*).

Case 2. Patient B., 14 years old, diagnosed with symptomatic, pharmacoresistant temporal lobe epilepsy. The first seizure occurred at the age of 4 years 10 months, a week before there had been a case of trauma, a fall from a tree.

The phenomenology of seizures: episodes of standing motionless («freezing»), accompanied by a «glassy look», unmotivated laughing, loss of contact with other people. Tonic-clonic seizures with the rotation of the eyeballs to the right and tonic withdrawal of the left upper extremity.

The patient was prescribed antiepileptic therapy, with repeated changes of regimens and dosages, without any effect. During MRI at the local clinic, a site of a pathological signal in the posterior parts of the right temporal lobe on the border with the parietal lobe, initially regarded as FCD type II was revealed. During a comprehensive, in-depth pre-surgical examination, with an interdisciplinary consultation, on the basis of a set of radiological signs, a tumor from the LEAT group was suggested (Fig. 4).

The main marker let us suggest the presence of neoplasm was a large calcinate in the central parts of the pathological focus on SWI images, which is considered to be a very rare phenomenon for FCD [4, p. 621].

The dominant focus of epileptiform activity when compared with the results of video EEG monitoring correlated with structural changes being visible on MRI. On the base of the obtained data, surgical treatment was

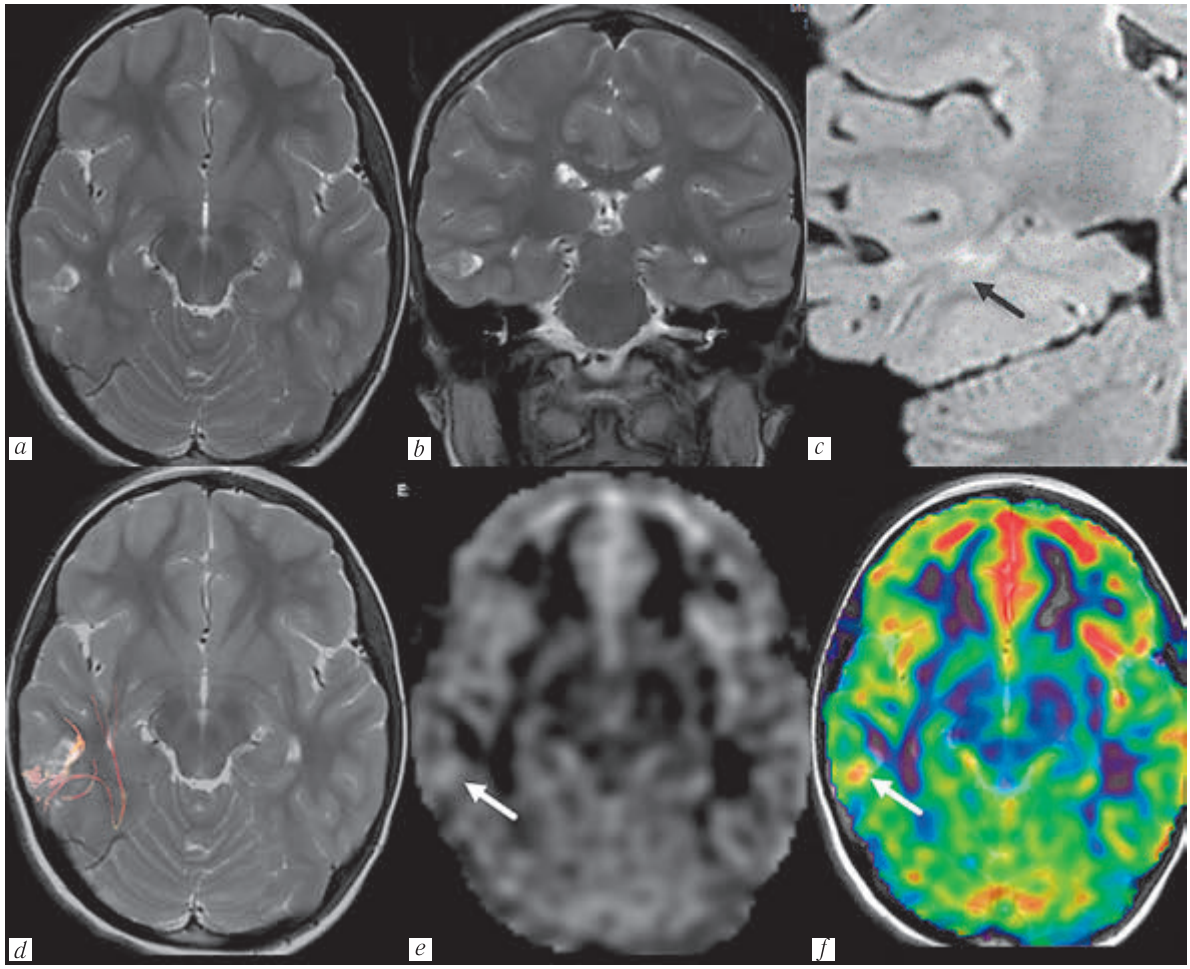

Fig. 2. Dynamic MRI 3,0 T of patient Kh., at the age of 11 years. The protocol was modified for the individual characteristics of the patient. When comparing dynamic MRI scans the «salt and pepper» symptom is noted due to an inhomogeneous increase in the signal in the T2 pulse sequence from the central parts of the focus to the periphery (*a, b*). In the FLAIR 3D pulse sequence with a slice thickness of 1.0 mm, attention is drawn to a more distinct visualization of the transmantial spread of the pathological focus to the wall of the temporal horn of the lateral ventricle (*c*). According to DTI data, there is a deformation of the tracts in the area of question but without signs of their destruction or infiltration (*d*). On the ASL images, a local hyperperfusion site in the structure of the pathological focus is determined (*e, f*)

Рис. 2. Динамические МРТ 3,0 Т пациентки Х., в возрасте 11 лет с модификацией протокола под индивидуальные особенности пациента. При сравнении динамических МРТ отмечается симптом «соль с перцем» за счет неоднородного повышения сигнала в импульсной последовательности Т2 от центральных отделов очага к периферии (*a, b*). В импульсной последовательности FLAIR 3D с толщиной среза 1,0 мм обращает на себя внимание более отчетливая визуализация трансмантийного распространения патологического очага к стенке височного рога бокового желудочка (*c*). По данным DTI отмечается деформация трактов в зоне интереса без признаков их разрушения или инфильтрации (*d*). На изображениях ASL определяется локальный участок гиперперфузии в структуре патологического очага (*e, f*)

indicated. After preoperative preparation, surgical intervention, namely, microsurgical removal of a tumor of the right temporal lobe of the brain using neurophysiological monitoring was performed (Fig. 5, *a*).

A tumor with numerous calcifications was revealed during the pathoanatomic examination of the surgical material. Histologically neoplastic tissue had a diffuse growth pattern and consisted from relatively monomorphic astrocyte-type cells. Mitotic activity, necrosis and pathological proliferation of blood vessels were not revealed. The immunohistochemical study revealed diffuse expression of GFAP by tumor cells as well as subtotal expression of CD34.

Conclusion: The histological picture and immune phenotype correspond to a polymorphous low-grade neuroepithelial tumor of the young, CNS WHO grade 1 (Fig. 5, *b, c, d, e*).

During the period of postoperative observation of the patient no seizures were noticed. Based on the possible incomplete resection of the formation according to the results of postoperative MRI, it was decided to continue antiepileptic therapy in the same manner, with possible subsequent correction of the regimen and dosages.

Discussion. As mentioned above, PLNTY is a relatively new morphological unit, which was included in the classification of the central nervous system tumor

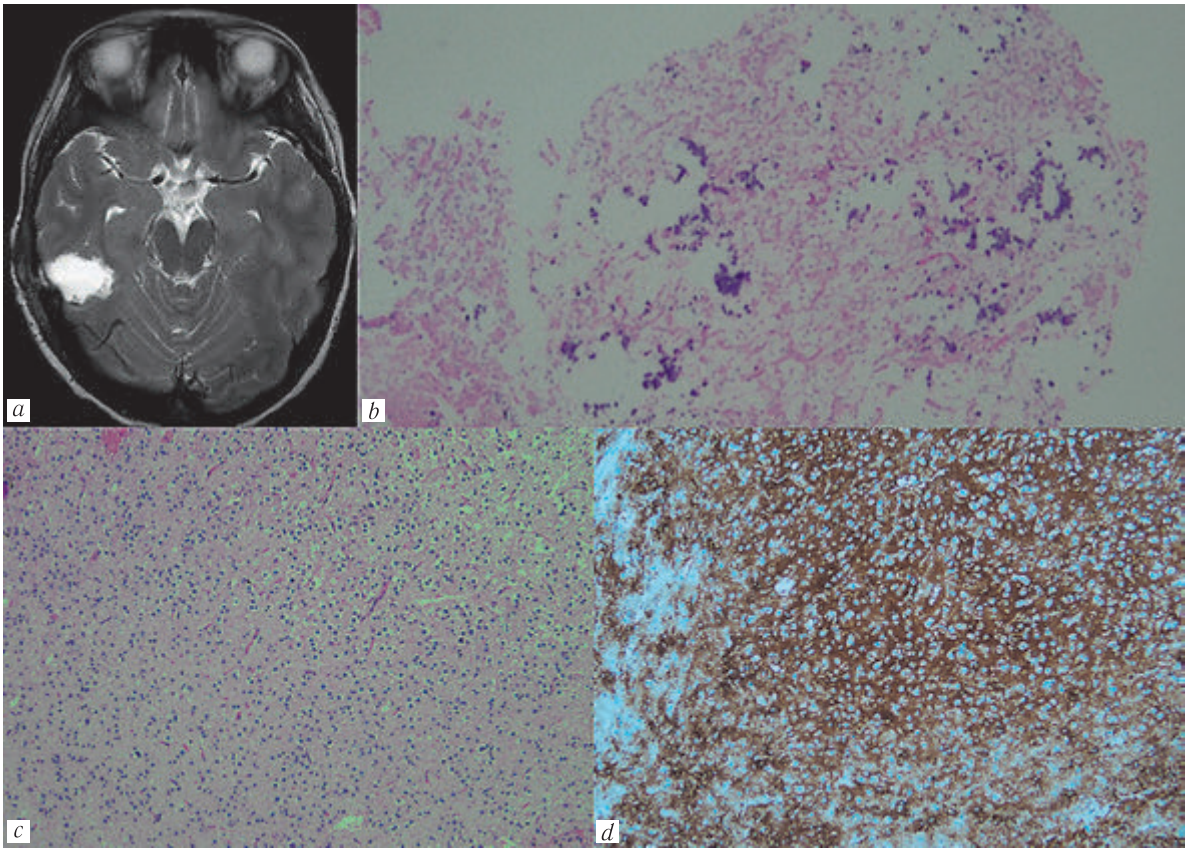

Fig. 3. Postoperative MRI shows a picture of total resection of the tumor and the underlying cortical plate in the right temporal lobe (*a*). A fragment of pathological tissue with numerous microcalcinates. Color of GE (*b*). A tumor with a diffuse growth pattern. Neoplastic cells are represented by elements of both oligodendroglial (top and right) and astrocytic (bottom and left) types. GE, ×100 (*c*). Tumor cells diffusely express CD34. Immune staining, ×100 (*d*)

Рис. 3. МРТ после повторной операции демонстрирует картину тотальной резекции опухоли и подлежащей кортикальной пластинки в правой височной доле (*a*). Фрагмент патологической ткани с многочисленными микрокальцинатами. Окраска ГЭ (*b*). Опухоль с диффузным паттерном роста. Неопластические клетки представлены как элементами олигодендроглиального (сверху и справа), так и астроцитарного (снизу и слева) типов. ГЭ, ×100 (*c*). Клетки опухоли диффузно экспрессируют CD34. Иммунное окрашивание, ×100 (*d*)

by the WHO in 2021 and assigned to the first grade. At the moment, in the literature, taking into account the cases published in the article, no more than one hundred pathomorphologically confirmed cases of PLNTY have been noted. In Russia, until now there have been no publications devoted to this scarce tumor.

As is the case with other neoplasms of the LEAT group, a distinctive sign of PLNTY is its high epileptogenicity [5, p. 5]. Therefore, these tumors are usually revealed by neuroimaging methods after the manifestation of epileptic seizures, or with a long-term history of epilepsy since childhood.

The high prognostic significance of surgical resection of this tumor was pointed out in previously published papers; as a result, a patient did not experience seizures, and possible accompanying cognitive impairment could be minimized. However, not in all patients it is possible to achieve such results [2, p. 419]. The factors associated with seizure control remain unclear at the moment, but perhaps this may be due to the presence of a residual dysplastically altered peritumoral cortex, the so-called FCD IIIb (ILAE 2011) during focal tumor resection [6, p. 6].

Therefore, it seems relevant to use intraoperative neurophysiological methods for recording epileptic activity, in particular electrocorticography (ECoG), for the purpose of total resection of peritumoral MR, namely, negative changes resulting in epileptogenesis [7, p. 41].

We assume that it is for this reason a recurrence of seizures was notices in the patient Kh., and for this reason a second surgical intervention was required two years after the first operation.

Applying our findings to the diagnostic setting, several considerations should be kept in mind.

According to radiological methods, PLNTY may look like a clearly limited lesion, or with a mixed border between the tumor and normal tissue, usually with the presence of a cystic solid structure. Like other tumors of the LEAT group, PLNTY is mostly localized in the temporal lobe.

On MRI, the formation is determined as a cortical/subcortical focus of an iso/hypointensive signal at T1 and an in homogeneously elevated signal at T2, with an ambiguous ratio to contrast enhancement [5, p. 2; 8, p. 180; 9, p. 1327; 10, p. 1255]. In their work Chen Y. et al. reported about a presumptive specific pat-

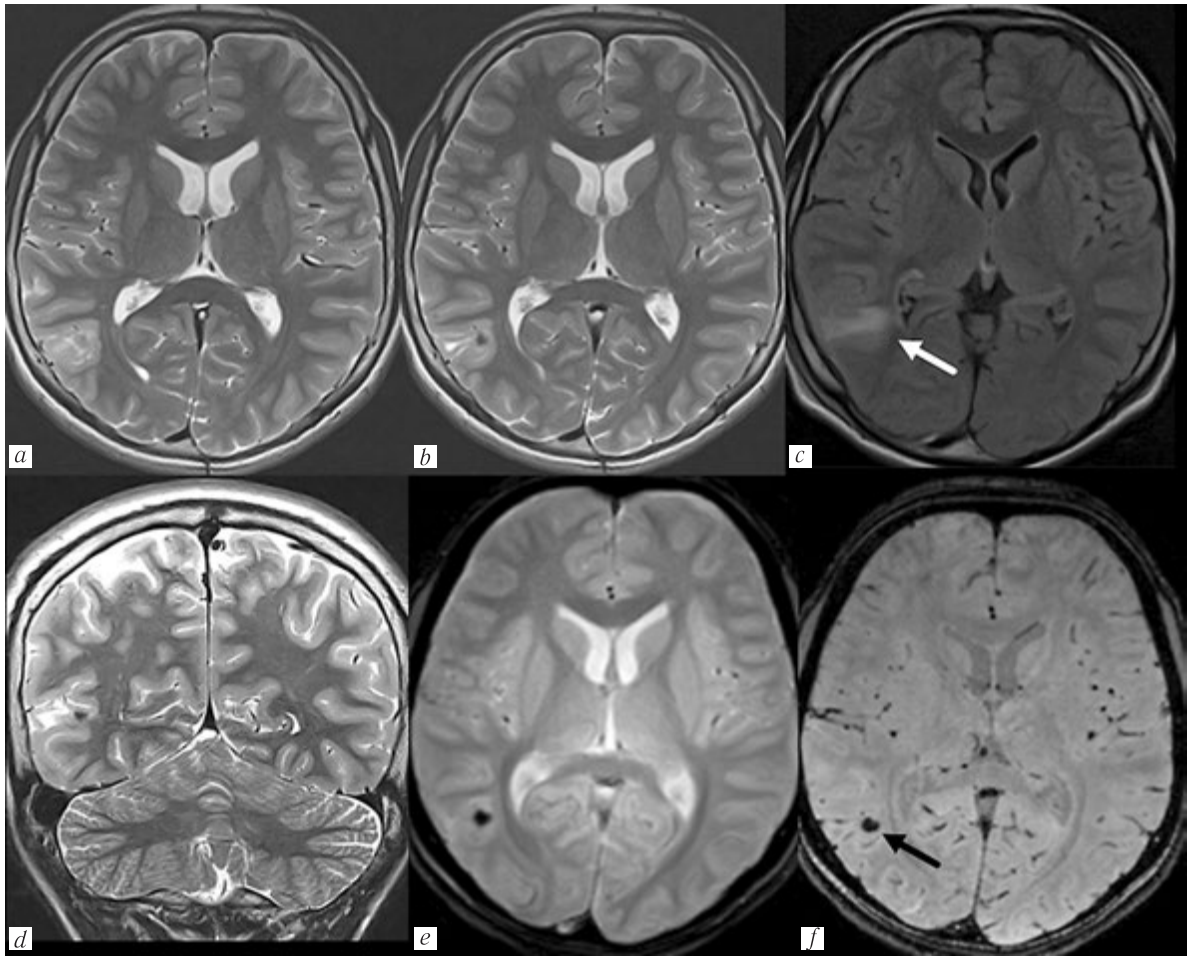

Fig. 4. Patient B., 14 years old. The site of the pathological signal in the posterior parts of the right temporal lobe during the transition to the parietal lobe in the T2 and FLAIR pulse sequences, with the presence of the «salt and pepper» symptom, cystic solid structure, predominant localization in the cortex, without perifocal reaction and mass effect. The FLAIR images clearly visualize the transmantic spread of the focus to the wall of the lateral ventricle (*a, b, c*). In the central parts of the focus, a large calcinate is determined, the ECHO gradient is clearly distinguishable on the pulse sequences T2, and with the magnetization transfer SWI (SWAN) (*d, e, f*)

Рис. 4. Пациент Б., 14 лет. Участок патологического сигнала в задних отделах правой височной доли при переходе в теменную в импульсных последовательностях Т2 и FLAIR, с наличием симптома «соль с перцем», кистозно-солидной структуры, преимущественной локализацией в коре, без перифокальной реакции и масс-эффекта. На изображениях FLAIR отчетливо визуализируется трансмантийное распространение очага до стенки бокового желудочка (*a, b, c*). В центральных отделах очага определяется крупный кальцинат, хорошо различимый на импульсных последовательностях Т2 градиент ЭХО и с переносом намагниченности SWI (SWAN) (*d, e, f*)

tern of PLNTY in the form of a «salt and pepper» sign which was formed due to multiple small or large calcifications in the central parts of the tumor and was clearly visible on T2 and FLAIR images during MRI procedure.

This specific sign was revealed in all three patients described in the study; in addition, a comparison with the radiological picture of other tumors of the LEAT group was made, in particular with the symptom of a «soap bubble» in DNET. At the same time, making a differential diagnosis the authors mentioned the blurring of the boundaries between the area affected and normal tissue and the «transmantle» sign as distinctive sign of FCD [11, p. 5–6].

In our works devoted to the LEAT group tumors, we have repeatedly given examples that both gangliogliomа and DNET, as well as angiocentric glioma, may spread transmantically [12, pp. 14, 15].

In both cases in patients Kh. and B., the tumors were contrast negative; with MRI on T2 and FLAIR, the «salt and pepper» sign was shown due to the presence of heterogeneous calcinates and a cystic-solid structure with local blurring of gray-white differentiation.

In addition to these signs, we would like to emphasize the presence in both cases of a transmantic spread of the formation, forming a similarity of a «trasmantle» sign, localization of the main node of the tumor in the cortex and a «triangular» configuration of the pathological focus with the apex facing towards the lateral ventricle. As evidenced by the foregoing, we could say that PLNTY may mimic the radiological characteristics of type II FCD on routine MRI, which would be interpreted as the main reasons for misdiagnosis in initial (primary) studies [8, p. 181].

Therefore, the images are sure to be interpreted by specialists engaged in visualization of the structural

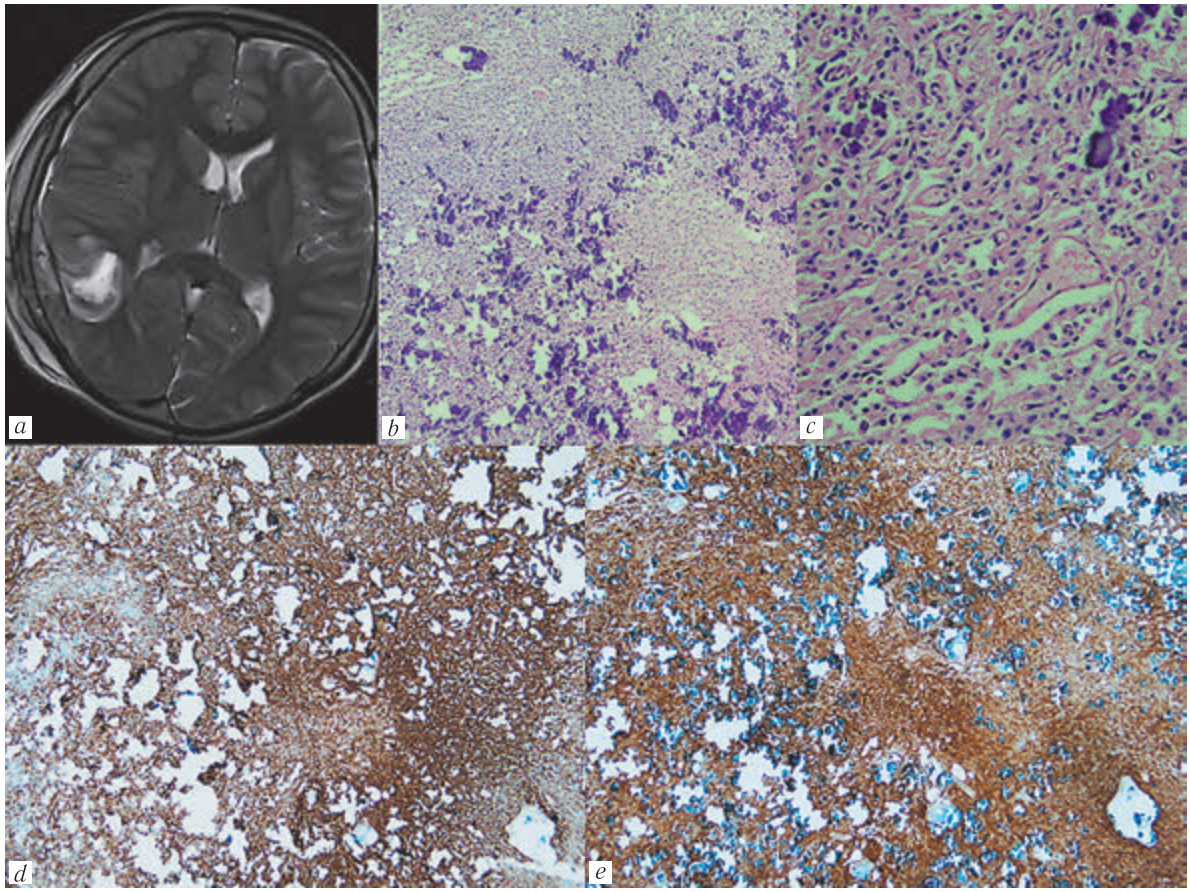

Fig. 5. Postoperative MRI shows a focal resection of the tumor in the right temporal lobe with the presence of blood decay products along the border of the removed tumor bed, subdural hematoma and reactive edema in the area of osteoplastic changes (*a*). Tumor tissue with diffuse growth type and numerous calcinates. Staining with hematoxylin and eosin, ×40 (*b*). The cells morphology is predominantly neoplastic cells of the astrocytic line. Hematoxylin and eosin staining, ×200 (*c*). Diffuse GFAP expression. Immune staining, ×40 (*d*). Widespread expression by CD34 tumor cells. Immune staining, ×40 (*e*)

Рис. 5. Постоперационная МРТ демонстрирует картину очаговой резекции опухоли в правой височной доле с наличием продуктов распада крови по границе ложа удаленной опухоли, субдуральной гематомы и реактивного отека в зоне костно-пластических изменений (*a*). Опухолевая ткань с диффузным типом роста и многочисленными кальцинатами. Окраска гематоксилином и эозином, ×40 (*b*). Клетки преимущественно имеют морфологию неопластических клеток астроцитарной линии. Окраска гематоксилином и эозином, ×200 (*c*). Диффузная экспрессия GFAP. Иммунное окрашивание, ×40 (*d*). Распространенная экспрессия опухолевыми клетками CD34. Иммунное окрашивание, ×40 (*e*)

basis of epilepsy as well as an additional modification of the protocol for the individual characteristics of a patient with the mandatory inclusion of SWI (SWAN), DTI and contrast–free/contrast MR perfusion (ASL/DSC) sequences, or combination with CT results.

J. S. Benson и соавт. mentioned about a curious observation during DSC devoted to hyperperfusion sites in the PLNTY structure, although no traces of neovascularization were found during pathoanatomic examination [3, p. 577].

In patient Kh., a hyperperfusion site was also found in the structure of an epileptogenic substrate of unclear etiology, but according to ASL data it became possible to suspect the presence of neoplasm at the preoperative stage. These results worth further studying, especially in the light of a single mention of the atypical characteristics of PLNTY when using PET CT with ^{11}C-methionine and ^{18}F-fluorodeoxyglucose [13, p. 5–6].

Thus, if there is a revealing of dynamically stable, contrastnegative pathological epileptogenic substrate in the temporal lobe with a cystic/multicystic structure, multiple or single large calcinates, a «salt and pepper» sign according to T2/FLAIR data and in some cases the lesion manifests transmantic spreading to the wall of the lateral ventricle, it is advisable to consider and include the presence of PLNTY. In order to narrow the differential row, modification of the scanning protocol for the individual characteristics of a patient in real time as well as the combination of the MRI, CT and PET CT results seem to be quite appropriate.

Conclusion. Polymorphous low-grade neuroepithelial tumor of the young (PLNTY) is a new, not yet widely recognized epileptogenic neoplasia associated with severe epilepsy in childhood.

Despite the similarity of radiological signs with other tumors that induce epilepsy, several signs can be distinguished, such as the «salt and pepper» sign, multiple calcifications in the structure and predominantly cortical localization of the tumor.

The newly revealed visualization patterns, namely, a resemblance of a «transmantle» sign, a «triangular» configuration of the pathological focus (when the base is located in the basement membrane of the cortex, whereas the tip is facing towards the lateral ventricle), possible hyperperfusion according to ASL/DSC data, require further observation in statistically significant groups of patients with PLNTY.

In the cases when an epileptogenic substrate of unclear etiology is revealed, it is recommended to modify the routine protocol of MR scanning with the mandatory addition of contrast enhancement and pulse sequences SWI (SWAN), DTI, ASL/DSC.

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