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Case Report

A "polymorphous low-grade neuroepithelial tumor of the young (PLNTY)" diagnosed in an adult. Report of a case and review of the literature

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ABSTRACT

Background: Polymorphous low-grade neuroepithelial tumor of the young (PLNTY) is a rare neuropathological entity, recently introduced in neuro-oncology. These tumors, histologically similar to oligodendrogliomas, cause epilepsy, occurring in children and young adults. Only few cases of PLNTY have been described in literature and all reported cases invariably focused on the onset of these tumors in children and young adults.

Case Description: We report the case of a 50-year-old woman suffering from epilepsy since the 1st year of her life. Computed tomography scan and magnetic resonance imaging of the brain documented the presence of a calcified mass involving left temporal lobe. The tumor was surgically excised and the histological examination showed a hypocellular and massively calcified neoplasm with morphological and immunohistochemical features consistent with the diagnosis of "PLNTY."

Conclusion: A review of the literature revealed that there are 31 cases of PLNTY reported in literature, most of which are children or young adults. The present case represents the second PLNTY diagnosed in a middle-aged adult to the best of our knowledge, suggesting that PLNTY should always be included in the differential diagnosis of low-grade brain tumors, also in adult patients.

Keywords: Adult, Case report, Epilepsy, Neuroepithelial tumor, Polymorphous low-grade neuroepithelial tumor of the young

INTRODUCTION

Low-grade neuroepithelial tumors represent a wide spectrum of brain lesions characterized by heterogeneous histological and molecular features, indolent biological behavior (WHO Grade I and II) and frequent association with early onset of epilepsy in children and young adults.[9,22] Many of them belong to the wide category of the so-called "long-term epilepsy associated brain tumors (LEATs)" or epileptomas: [9] pilocytic astrocytoma (PA), ganglioglioma (GG), pleomorphic xanthoastrocytoma (PXA), dysembryoplastic neuroepithelial tumor (DNT), oligodendroglioma, and angiocentric glioma are the most common tumor entities included in this group.[22] The term "polymorphous low-grade

neuroepithelial tumor of the young (PLNTY)" indicates a recently introduced epileptogenic neoplasm, first described in 2017 by Huse *et al*.,[18] affecting children and young adults and almost invariably characterized by oligodendroglioma-like histological appearance, diffuse cluster of differentiation-34 (CD34) immunohistochemical expression, and MAP kinase pathway-related molecular aberrations.[18] Since then, few other cases of PLNTY have been reported in the literature,[6,8,12,14,15,19,21,26,28,30] all in the form of single case reports or short case series, and most of them included children or young adults.[6,8,12,14,15,19,21,28,30] The temporal lobe is the most frequently affected site and epilepsy is the most common presenting symptom.[18] Histologically, despite being a morphologically heterogeneous tumor, PLNTY constantly shows an oligodendroglioma-like cell component, infiltrative growth pattern, and aberrant immunohistochemical staining for CD34.[8,18] Calcifications are also common histological findings.[18] On the molecular point of view, PLNTYs display a distinct DNA methylation profile and frequently harbor B-Raf proto-oncogene (BRAF)-V600E mutations or fusions involving fibroblast growth factor receptors 2 and 3 (FGFR2 and FGFR3).[8,18] In most cases, PLNTY exhibits indolent clinical behavior, simulating a WHO Grade I neoplasm, although a well-defined histological grade has not yet been assigned.[18] Gross total surgical resection represents the standard of care, aiming to reduce the mass effect of tumor on surrounding normal brain and to obtain control of seizures; the onset of local recurrences seems to be related to incomplete surgical excisions.[18]

We herein report a rare case of PLNTY presenting as an extensively calcified mass located to the left temporal lobe, in a 50-year-old woman with a long time history of drug-resistant epilepsy. A critical review of the scientific literature on PLNTY, highlighting that this tumor may be potentially diagnosed at any age and should not be considered only as a pediatric tumor, is also included.

CASE REPORT

History and clinical findings

A 50-year-old lady, left handed, was referred to the department of neurological surgery, because of a drug-resistant epilepsy. She had a medical history, featured by the onset of seizures since early childhood. The patient was treated over several years with a combination of different anti-epileptic drugs, but she never obtained a complete control of seizures. During the past 2 years before admission, the patient experienced the worsening of crises. These were featured by mental confusion, reduced consciousness, dyskinesia in the upper right limb, and gaze deviation towards the right side. The crises occurred with a frequency of 3–4 episodes per day and occasionally had secondary generalization. She had also hypothyroidism. When the patient was hospitalized, neurological examination revealed moderate mental impairment with slowed speech and gait. Neuroradiological assessment was performed. Brain computed tomography (CT) revealed the presence of a densely calcified lesion involving left temporal lobe and insula [Figure 1a]. Brain magnetic resonance imaging (MRI) documented the presence of a hypointense (both in T1 and T2 sequences), nonenhancing lesion, surrounded by a hyperintense area detected by long-TR sequences [Figure 1b]. The patient was scheduled for surgery.

Surgical procedure

The patient was positioned supine with the left shoulder elevated and head 60° rotated to the right side and fixed by radiolucent three-point Mayfield clamp. Preoperative CT scan with and without iodinate contrast was performed after positioning. Images of postpositioning CT scan were merged with preoperative MRI and used for neuronavigation. Motor evoked potentials, somatosensory evoked potentials and direct monopolar and bipolar cortical and subcortical stimulation were used for neuromonitoring. The intraoperative imaging protocol used in the present case has been previously detailed.[4,5,11] Under general anesthesia, a left frontotemporal craniotomy was performed. Using intraoperative ultrasound (i-US) and neuronavigation, lesion was approached through a trans-sulcal approach transtemporal (T1-T2). i-US revealed a hyperechoic tumor and calcifications were clearly visible. Intraoperatively, lesion was distinguishable from normal white matter, albeit, in some areas, it appeared infiltrating. Calcifications were detected and resected. According to the institutional intraoperative protocol for intra-axial brain tumor, we also used 5-aminolevulinic acid (5-ALA) fluorescence, but the tumor was not fluorescent. When resection was deemed complete, intraoperative CT (i-CT) was performed and a small calcification was detected. The images of i-CT scan were used for updated neuronavigation, which was useful to localize and resect the small calcified tumor remnant. Complete resection of the lesion was achieved as confirmed by last i-US acquisition and final i-CT. Neuromonitoring did not document any variation of during all surgical procedure.

Postoperative course and outcome

Postoperative MRI confirmed the radical resection of the lesion [Figure 1c]. Postoperative course was uncomplicated and the patient was discharged on the 6th postoperative day, without any neurological deficit. The frequency and intensity of crises eventually decreased, but the pharmacological therapy was not modified, as electroencephalographic alterations still persisted also during follow-up.

Pathological findings

Grossly, the tumor sample consisted of multiple fragments, markedly hard in consistency, measuring 2 cm in aggregate

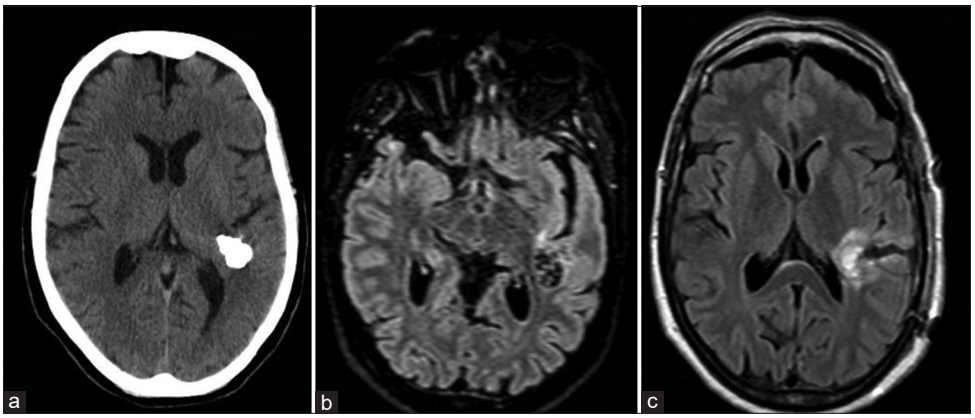

Figure 1: Preoperative neuroradiological assessment. Computed tomography scan (a) demonstrating the presence of a densely calcified left temporal and insular lesion. FLAIR sequences of magnetic resonance (MR) scan confirmed the presence of the calcified tumor (b). Postoperative MR confirmed the complete resection of the lesion (c).

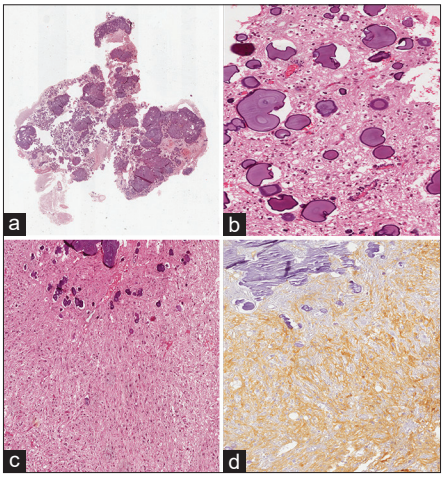

Figure 2: Histological examination. (a) Low magnification showing the densely calcified nature of the tumor mass (hematoxylin and eosin; original magnification 25×); (b) tumor was mainly composed of small and roundish cells with oligodendroglioma-like morphology intermingled with calcifications ranging from small calcospherules to larger and confluent psammomatous bodies (hematoxylin and eosin; original magnification 150×); (c) a less represented neoplastic component with astrocytic-like morphology was also found (hematoxylin and eosin; original magnification 50×); (d) neoplastic cells were diffusely and strongly stained with CD34 (immunoperoxidase; original magnification 50×).

diameter. Histologic examination showed an extensively calcified and hypocellular lesion [Figure 2a], mainly composed of abundant calcifications, ranging from small calcospherules to larger and confluent psammomatous bodies, intermingled with a much smaller component of bland looking, small and roundish oligodendroglioma-like neoplastic cells [Figure 2b], showing small rounded nuclei, perinuclear halos and arranged in an infiltrative manner; in addition, tumor exhibited a less represented, spindled, astrocytic-like component [Figure 2c]. Some neoplastic cells were vaguely distributed around capillary blood vessels, resulting in a focal formation of perivascular pseudorosette-like structures. No Rosenthal fibers or eosinophilic granular bodies were found. Neither necrosis, nor mitoses, nor microvascular proliferation was identified. Immunohistochemically, neoplastic cells were strongly and diffusely stained with CD34 [Figure 2d], glial fibrillary acidic protein (GFAP) and oligodendrocyte transcription factor 2 (OLIG2), and retained the α-thalassemia X-linked mental retardation syndrome (ATRX) nuclear expression. No immunohistochemical staining was obtained with isocitrate dehydrogenase 1 (IDH1)-R132H, synaptophysin, chromogranin-A, epithelial membrane antigen, neuronal nuclear antigen (Neu-N), and BRAF-V600E. Ki-67 proliferation index was unremarkable (<1%). Molecular analyses showed absence of IDH1/IDH2 and BRAF mutations. Fluorescence *in situ* hybridization for 1p/19q codeletion was negative. Based on both morphological and immunohistochemical features, a diagnosis of "PLNTY" was rendered.

DISCUSSION

Apart from the most common glial tumors,[2,3,10] the central nervous system (CNS) can be the primary site of onset of different types of neoplasms, including neuroepithelial, neuronal, embryonal, and mesenchymal tumors.[1,27] Primary CNS tumors exhibit a large series of signs and symptoms, largely dependent on the specific brain location, including seizures or other neurological deficits.[9,22] LEATs or epileptomas form a separate category of low-grade gliomas and glioneuronal neoplasms affecting children and young adults and characterized by indolent biological behavior, spontaneous onset of epilepsy at a young age, cortical localization (primarily to the temporal lobe, followed by frontal lobe and other cortical sites), frequent association with type IIIb focal cortical dysplasia, and, despite having different morphologies, similar radiological and molecular features.[9,26] PLNTY is a recently described, uncommon low-

grade neuroepithelial neoplasm, not yet included in the WHO classification of CNS tumors, but part of the LEAT category.[18,26] Histologically, despite its variable appearance, PLNTY almost invariably exhibits an oligodendroglioma-like morphology, infiltrative growth in the surrounding brain parenchyma, and diffuse and strong CD34 immunopositivity and MAP-kinase pathway-related molecular aberrations.[18,23-25] Notably, no CD34 immunoexpression is usually found in adult CNS and, although the exact nature of CD34-positive cells in CNS is not been fully established, it has been hypothesized that CD34 immunoexpression indicated an origin from dysplastic and/or differentiating in a divergent way neural precursor.[9,26] Classically, immunohistochemical expression of CD34 is restricted to leukemic blasts, myofibroblasts, and, with less specificity, endothelial cells.[26] While among non-CNS neoplasms, myeloid leukemia and multiple soft-tissue neoplasms, such as solitary fibrous tumors, gastrointestinal stromal tumors, and malignant peripheral nerve sheath tumors among the others, show diffuse or heterogeneous CD34 immunostaining,[26] in CNS, an extravascular CD34 positivity may be commonly found in the large neural or ramifying neuroepithelial cells usually associated to glioneuronal tumors but, notably, not in the neoplastic cells.[13,26]

Based on both the facts that PLNTY has a distinct DNA methylation profile and almost invariably presents genetic features involving the MAP kinase pathway components, such as BRAF-V600E mutations or FGFR3- transforming acidic coiled-coil-containing protein 3 (TACC3), FGFR2-KIAA1598, and FGFR2-CTNNA3 fusions, it has now been widely accepted that PLNTY is a distinct tumor entity, belonging to the wide spectrum of low-grade neuroepithelial neoplasms.[18,26] PLNTY has been first described as separate entity by Huse *et al*.[18] who reported a series of 10 cases arising in young patients aged from 4 to 32 years with associated epilepsy in 8/10 cases. These authors[18] found that all reported cases shared similar morphological and immunohistochemical features: (i) presence of a round cell component with rounded nuclei and perinuclear clear halos, reminiscent of oligodendroglioma, (ii) infiltration of the surrounding brain tissue, (iii) at least focal perivascular pseudorosette formation, (iv) presence of diffuse calcifications, (v) immunoexpression of GFAP, OLIG-2 combined to diffuse CD34 immunostaining, and (vi) absence of staining with synaptophysin, Neu-N, and IDH1-R132H. Although the presence of an oligodendroglioma-like morphology was a constant finding, some of the PLNTY cases reported by Huse *et al*.[18] showed a more polymorphous histological appearance, ranging from round cell with perinuclear halo, to spindled, fibrillary and pleomorphic cell component, more similar to the morphology of astrocytic tumors. No high-grade features, including pleomorphism, necrosis, and microvascular proliferation, were described in this series.[18] Among the cases that were molecularly screened by Huse *et al*.,[18] BRAF-V600E mutations were found in three cases, while the presence of FGFR3-TACC3, FGFR2-CTNNA3, and FGFR2-KIAA1598 fusion transcripts in one case, one case, and two cases, respectively; notably, it must be emphasized that BRAF mutations and fusions involving FGFR2/3 genes were mutually exclusive.[18] Finally, on DNA methylation profile analysis, the PLNTY cohort reported by Huse *et al*.[18] exhibited a peculiar gene signature, closely related to other low-grade tumors, including GGs in particular, PAs and DNTs, confirming the existence of PLNTY as separate and distinct tumor entity.

The unusual age of onset of the present case led us to critically perform a literature review and we found that, since its first description,[18] few cases of PLNTY have been reported in the English literature, mostly in the form of single case or small case series,[6,8,12,14,15,19,21,26,28,30] and almost all cases have been described in children and young adults;[6,8,12,14,15,19,21,28,30] the main findings of the literature review are summarized in [Table 1]. Apart from the present case, to the best of our knowledge, only one case has been documented in a middle-aged adult patient by Riva *et al*. in 2018;[26] in particular, apart from the patient's age (57 years), the other two peculiarities of the above-mentioned case[26] were the absence of associated seizures and the histopathological detection of microvascular proliferation areas admixed with neoplastic oligodendroglioma-like cells. Moreover, we think that the two young adult cases of "massively calcified low-grade gliomas" described by Hewer *et al*. in 2016[17] are reasonably PLNTYs based on both clinicoradiological data and morphology, despite the absence of CD34 immunostaining and molecular data in the original report; accordingly, we believe that a histopathological revision of the "doubtful" cases in light of the recent acquisitions about PLNTY would be useful to expanding the age spectrum in which this entity can occur.

The two most peculiar features of the present case were the age at the diagnosis and the onset as a lowly cellular and diffusely calcified nodule. Even in this regard, we retrospectively reviewed the scientific literature to investigate the presence of papers focusing on the calcified aspect of PLNTY nodules; Johnson *et al*. in 2019[19] highlighted the predominantly calcified nature of PLNTY masses, reporting a series of nine pathologically proven PLNTY cases in which the presence of diffuse calcifications was a constant finding; notably, authors emphasized the concept that the peculiar radiological pattern of prominent central calcification was quite suggestive of PLNTY and allowed to radiologically distinguish it from other highly calcified CNS lesions.[19] Furthermore, among the cases of "diffusely calcified pediatric low-grade gliomas" published before PLNTY first description, at least four of them[16] should be retrospectively diagnosed as PLNTYs in light of the current knowledge about this entity; moreover, in our opinion, the true incidence of PLNTY is largely underestimated due to the fact that many of the published cases of diffusely calcified

Table 1: Summary of the clinicopathological features of the PLNTY cases reported in literature to date.

Case	Age (years)	Gender	Presentation	Location	Treatment	Outcome	Molecular findings
Huse *et al.* (n=10)[18]	Range 4–32	4 M; 6 F	Epilepsy (i=8) Dizziness (*n*=1) Visual disturbances (*n*=1)	R temporal (*n*=7) L temporal (*n*=1) R occipital (*n*=1) R frontal (*n*=1)	GTE (*n*=8) PTE (*n*=1) N/A (*n*=1)	NED (n=7) Postoperative psychosis (n=1) New seizures (n=1) N/A (n=1)	BRAFV600E (*n*=3) FGFR3-TACC3 (*n*=1) FGFR2-KIAA1598 (*n*=2) FGFR2-CTNNA3 (*n*=1) - N/A (*n*=3)
Bitar *et al.*[8]	31	M	Epilepsy	Temporal	N/A	N/A	BRAFV600E
Riva *et al.*[26]	57	M	Headaches	R frontal	GTE	NED	FGFR3-TACC3
Sumdani *et al.* [28]	19	M	Epilepsy	R parietal	GTE	Postoperative seizures	BRAFV600E
Johnson *et al.* (n=9)[19]	Range 5–34	2M; 7F	N/A	R temporal (*n*=3) L temporal (*n*=3) R parietal (*n*=1) L parietal (*n*=1) Third ventricle (*n*=1)	N/A	N/A	BRAFV600E (*n*=4) KIAA1598-FGFR2 (*n*=2) FGFR2 rearrangement (*n*=1) BRAF fusion (*n*=1) NTRK2 disruption (*n*=1)
Gupta *et al.*[15]	30	M	Epilepsy	R temporal	GTE	NED	BRAFV600E
Chen *et al.* (n=3)[12]	14; 15; 16	2 M; 1 F	Epilepsy	R temporal (*n*=1) L temporal (*n*=1) R frontal (*n*=1)	GTE	NED	BRAFV600E (*n*=1) FGFR3-TACC3 (*n*=1) N/A (*n*=1)
Benson *et al.*[6]	44	F	Behavioral disturbances	L temporal	GTE	NED	BRAFV600E
Lelotte *et al.*[21]	33	F	Epilepsy	R temporal	GTE	N/A	BRAFV600E
Tateishi *et al.*[30]	14	M	Epilepsy	L temporal	GTE	NED	BRAFV600E
Ge *et al.* (n=2)[14]	14; 25	2F	Epilepsy	Temporal (*n*=1) Occipital (*n*=1)	GTE	NED	BRAFV600E (*n*=1) N/A (*n*=1)

M: Male, F: Female, R: Right, L: Left, GTE: Gross total excision, PTE: Partial tumor excision, NED: No evidence of disease, N/A: Not available

lesions previously diagnosed as PXA, PA, or pediatric oligodendroglioma may, instead, be PLNTYs and, therefore, should be histopathologically reevaluated.[7,20,24,29]

On the histopathological point of view, as PLNTY is a round cell with clear perinuclear halo tumor; the differential diagnosis usually includes CNS round/clear cell neoplasms (oligodendroglioma, clear cell ependymoma, and PA with oligodendroglioma-like features) that, however, can be excluded due to the combination of morphological, immunohistochemical (CD34 strong and diffuse expression), and molecular features of PLNTY.[18,26,28]

Moreover, the diagnostic pre- and intra-operative diagnostic assessment used in this case and commonly applied at our institution for all intrinsic brain lesion allowed a thorough study of imaging features of PLNTY. These tumors have common radiological features with calcified cavernous malformations and oligodendrogliomas. i-US revealed a hyperechoic lesion with characteristic posterior shadowing due to the presence of calcium. We also documented that these lesions do not show any 5-ALA induced fluorescence, under ultraviolet light. The present case represents the second confirmed report of PLNTY arising in a middle-aged patient and it led us to reflect on the real need to no longer consider PLNTY as a tumor of the pediatric/young age; although it is likely that, being a slow-growing neoplasm, PLNTY arises at an early age and then is diagnosed late, it is also true that, as PLNTY is associated in almost all cases with the onset of seizures, the hypothesis that a patient can neglect the symptoms and not undergo further diagnostic tests is quite remote.

CONCLUSION

The present case emphasizes the concept that, in the presence of an appropriate clinicoradiological context, neuroradiologists, neurosurgeons, and neuropathologists must include PLNTY in the differential diagnosis even in presence of an adult patient.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Altieri R, Morrone A, Certo F, Parisi G, Buscema G, Broggi G, *et al*. Tentorial angioleiomyoma: A rare neurosurgical entity. Case report and review of the literature. World Neurosurg 2019;130:506-11.
2. Altieri R, Barbagallo D, Certo F, Broggi G, Ragusa M, di Pietro C, *et al*. Peritumoral microenvironment in high-grade gliomas: From flairectomy to microglia-glioma cross-talk. Brain Sci 2021;11:200.
3. Barbagallo D, Caponnetto A, Barbagallo C, Battaglia R, Mirabella F, Brex D, *et al*. The GAUGAA motif is responsible for the binding between circSMARCA5 and SRSF1 and related downstream effects on glioblastoma multiforme cell migration and angiogenic potential. Int J Mol Sci 2021;22:1678.
4. Barbagallo G, Maione M, Peschillo S, Signorelli F, Visocchi M, Sortino G, *et al*. Intraoperative computed tomography, navigated ultrasound, 5-amino-levulinic acid fluorescence and neuromonitoring in brain tumor surgery: Overtreatment or useful tool combination? J Neurosurg Sci 2019;Online ahead of print.
5. Barbagallo GM, Palmucci S, Visocchi M, Paratore S, Attinà G, Sortino G, *et al*. Portable intraoperative computed tomography scan in image-guided surgery for brain high-grade gliomas: Analysis of technical feasibility and impact on extent of tumor resection. Oper Neurosurg (Hagerstown) 2016;12:19-30.
6. Benson JC, Summerfield D, Carr C, Cogswell P, Messina S, Gompel JV, *et al*. Polymorphous low-grade neuroepithelial tumor of the young as a partially calcified intra-axial mass in an adult. AJNR Am J Neuroradiol 2020;41:573-8.
7. Berhouma M, Jemel H, Kchir N. Calcified pilocytic astrocytoma of the medulla mimicking a brainstem "stone". Pathologica 2008;100:408-10.
8. Bitar M, Danish SF, Rosenblum MK. A newly diagnosed case of polymorphous low-grade neuroepithelial tumor of the young. Clin Neuropathol 2018;37:178-81.
9. Blumcke I, Aronica E, Urbach H, Alexopoulos A, Gonzalez-Martinez JA. A neuropathology-based approach to epilepsy surgery in brain tumors and proposal for a new terminology use for long-term epilepsy-associated brain tumors. Acta Neuropathol 2014;128:39-54.
10. Broggi G, Salvatorelli L, Barbagallo D, Certo F, Altieri R, Tirrò E, *et al*. Diagnostic utility of the immunohistochemical expression of serine and arginine rich splicing factor 1 (SRSF1) in the differential diagnosis of adult gliomas. Cancers (Basel) 2021;13:2086.
11. Certo F, Altieri R, Maione M, Schonauer C, Sortino G, Fiumanò G, *et al*. FLAIRectomy in supramarginal resection of glioblastoma correlates with clinical outcome and survival analysis: A prospective, single institution, case series. Oper Neurosurg (Hagerstown) 2021;20:151-63.
12. Chen Y, Tian T, Guo X, Zhang F, Fan M, Jin H, *et al*. Polymorphous low-grade neuroepithelial tumor of the young: Case report and review focus on the radiological features and genetic alterations. BMC Neurol 2020;20:123.
13. Deb P, Sharma MC, Tripathi M, Chandra PS, Gupta A, Sarkar C. Expression of CD34 as a novel marker for glioneuronal lesions associated with chronic intractable epilepsy. Neuropathol Appl Neurobiol 2006;32:461-8.
14. Ge R, Fang HF, Chang YQ, Li Z, Liu CF. Clinicopathological features of polymorphous low-grade neuroepithelial tumor of the young. Zhonghua Bing Li Xue Za Zhi 2020;49:1131-5.
15. Gupta VR, Giller C, Kolhe R, Forseen SE, Sharma S. Polymorphous low-grade neuroepithelial tumor of the young: A case report with genomic findings. World Neurosurg 2019;132:347-55.
16. Gupta K, Harreld JH, Sabin ND, Qaddoumi I, Kurian K, Ellison DW. Massively calcified low-grade glioma-a rare and distinctive entity. Neuropathol Appl Neurobiol 2014;40:221-4.
17. Hewer E, Knecht U, Ulrich CT. Two adult cases of massively calcified low-grade glioma: Expanding clinical spectrum of an emerging entity. Neuropathology 2016;36:508-9.
18. Huse JT, Snuderl M, Jones DT, Brathwaite CD, Altman N, Lavi E, *et al*. Polymorphous low-grade neuroepithelial tumor of the young (PLNTY): An epileptogenic neoplasm with oligodendroglioma-like components, aberrant CD34 expression, and genetic alterations involving the MAP kinase pathway. Acta Neuropathol 2017;133:417-29.
19. Johnson DR, Giannini C, Jenkins RB, Kim DK, Kaufmann TJ. Plenty of calcification: Imaging characterization of polymorphous low-grade neuroepithelial tumor of the young. Neuroradiology 2019;61:1327-32.
20. Kim YE, Shin HJ, Suh YL. Pilocytic astrocytoma with extensive psammomatous calcification in the lateral ventricle: A case report. Childs Nerv Syst 2012;28:649-52.
21. Lelotte J, Duprez T, Raftopoulos C, Michotte A. Polymorphous low-grade neuroepithelial tumor of the young: Case report of a newly described histopathological entity. Acta Neurol Belg 2020;120:729-32.
22. Louis DN, Ohgaki H, Weistler OD, Cavenee WK. WHO Classification of Tumours of the Central Nervous System. Lyon: International Agency for Research on Cancer (IARC); 2016.
23. Nagaishi M, Arai M, Osawa T, Yokoo H, Hirato J, Yoshimoto Y, *et al*. An immunohistochemical finding in glioneuronal lesions

associated with epilepsy: The appearance of nestin-positive, CD34-positive and tau-accumulating cells. Neuropathology 2011;31:468-75.
24. Niimi M, Yoshida K, Mayanagi K, Kawase T. Extensive and dense calcification in the core of a ventrally exophytic brainstem glioma. Brain Tumor Pathol 2002;19:101-3.
25. Prabowo AS, Iyer AM, Veersema TJ, Anink JJ, Schouten-van Meeteren AY, Spliet WG, *et al*. BRAF V600E mutation is associated with mTOR signaling activation in glioneuronal tumors. Brain Pathol 2014;24:52-66.
26. Riva G, Cima L, Villanova M, Ghimenton C, Sina S, Riccioni L, *et al*. Low-grade neuroepithelial tumor: Unusual presentation in an adult without history of seizures. Neuropathology 2018;38:557-60.
27. Sloan EA, Chiang J, Villanueva-Meyer JE, Alexandrescu S, Eschbacher JM, Wang W, *et al*. Intracranial mesenchymal tumor with FET-CREB fusion-a unifying diagnosis for the spectrum of intracranial myxoid mesenchymal tumors and angiomatoid fibrous histiocytoma-like neoplasms. Brain Pathol 2020;31:e12918.
28. Sumdani H, Shahbuddin Z, Harper G, Hamilton L. Case report of rarely described polymorphous low-grade neuroepithelial tumor of the young and comparison with oligodendroglioma. World Neurosurg 2019;127:47-51.
29. Tamura M, Kohga H, Ono N, Zama A, Shibasaki T, Horikoshi S, *et al*. Calcified astrocytoma of the amygdalo-hippocampal region in children. Childs Nerv Syst 1995;11:141-4.
30. Tateishi K, Ikegaya N, Udaka N, Sasame J, Hayashi T, Miyake Y, *et al*. BRAF V600E mutation mediates FDG-methionine uptake mismatch in polymorphous low-grade neuroepithelial tumor of the young. Acta Neuropathol Commun 2020;8:139.

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