POLYMORPHOUS LOW-GRADE NEUROEPITHELIAL TUMOUR OF THE YOUNG (PLNTY) IN A 50-YEAR-OLD MALE: A RARE CASE REPORT WITH HISTOPATHOLOGICAL AND IMMUNOHISTOCHEMICAL CORRELATION

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ABSTRACT

Background: Polymorphous low-grade neuroepithelial tumour of the young (PLNTY) is a recently recognized low-grade epilepsy-associated neuroepithelial tumour characterized by oligodendroglioma-like morphology, prominent calcification, CD34 positivity, and alterations involving the MAP kinase pathway. PLNTY predominantly affects children and young adults and is currently classified as a CNS WHO grade 1 tumour. Occurrence in older adults is exceedingly rare.

Case Presentation: We report a rare case of PLNTY in a 50-year-old male who presented with recurrent generalized tonic–clonic seizures. Computed tomography of the brain demonstrated bilateral frontal white matter hypodensities involving the genu of the corpus callosum with focal calcifications. Histopathological examination revealed a diffusely infiltrating glial neoplasm composed of uniform round cells with oligodendroglioma-like morphology, branching capillary vasculature, and extensive calcifications without necrosis or microvascular proliferation. Immunohistochemistry showed Olig2 positivity, retained ATRX expression, wild-type p53, negative IDH1 (R132H), and low Ki-67 proliferative index (<1%). Based on morphologic and immunohistochemical findings, a diagnosis of PLNTY, CNS WHO grade 1, was established.

Conclusion: This case expands the recognized age spectrum of PLNTY and emphasizes the importance of considering this entity in the differential diagnosis of calcified oligodendroglioma-like tumours in adults. Accurate diagnosis requires careful integration of histopathology, immunohistochemistry, and molecular characteristics to avoid misclassification and overtreatment.

Keywords: PLNTY, Polymorphous low-grade neuroepithelial tumour of the young, epilepsy-associated tumour.

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INTRODUCTION

Polymorphous low-grade neuroepithelial tumour of the young (PLNTY) is a recently recognized rare low-grade neuroepithelial tumour of the central nervous system, first described as a distinct entity by Huse et al. in 2017.[1] It is now included in the 2021 World Health Organization Classification of Tumours of the Central Nervous System under the group of paediatric-type diffuse low-grade gliomas and is designated as CNS WHO grade 1.[2] PLNTY is considered an important member of the spectrum of low-grade epilepsy-associated tumours because of its strong association with seizures, indolent clinical course, and favourable prognosis following surgical excision.

PLNTY most commonly affects children, adolescents, and young adults, with the majority of reported cases occurring in the temporal lobe and presenting clinically with long-standing epilepsy or recurrent seizures. [1,3] However, with increasing recognition of this entity, cases have also been reported in older adults, suggesting that the age spectrum of PLNTY may be wider than initially described. The occurrence of PLNTY in a 50-year-old adult, as seen in the present case, is distinctly uncommon and clinically relevant because such tumours in adults may be mistaken for oligodendroglioma or other diffuse low-grade gliomas.

Radiologically, PLNTY typically presents as a well-circumscribed or infiltrative cortical/subcortical lesion, often associated with calcification and minimal or absent contrast enhancement. [3] the presence of calcification is a characteristic imaging feature and may be identified on computed tomography as nodular or coarse calcific foci. Magnetic resonance imaging usually shows a

T2/FLAIR hyperintense lesion with variable cystic change and limited mass effect. Although the temporal lobe is the most common site, extratemporal locations including frontal, parietal, and occipital lobes have also been described. The bilateral frontal involvement with extension into the genu of the corpus callosum in the present case represents an unusual radiological pattern.

Histopathologically, PLNTY is characterized by polymorphous architecture and oligodendroglioma-like cytology. Tumour cells are usually round to oval with perinuclear clearing, giving an appearance closely resembling oligodendroglioma. [1] Prominent calcification, including psammomatous and coarse dystrophic calcifications, is a frequent and diagnostically useful feature. The tumour may show an infiltrative growth pattern but lacks high-grade features such as necrosis, microvascular proliferation, brisk mitotic activity, and marked nuclear atypia. These features support its classification as a low-grade neoplasm.

Immunohistochemically, PLNTY commonly shows expression of glial markers such as Olig2 and frequently demonstrates aberrant CD34 expression. [1,4] Retained ATRX expression, wild-type p53 pattern, low Ki-67 proliferation index, and absence of IDH1 R132H mutation are important supportive findings. These features help distinguish PLNTY from adult-type diffuse gliomas, particularly oligodendroglioma and astrocytoma. Oligodendroglioma typically shows IDH mutation and 1p/19q co-deletion, whereas PLNTY is generally IDH-wild type and lacks 1p/19q co-deletion. [2]

At the molecular level, PLNTY is strongly associated with alterations involving the mitogen-activated protein kinase pathway, including BRAF V600E mutation and FGFR2 or FGFR3 fusions. [3,5] These molecular alterations have helped establish PLNTY as a biologically distinct tumour entity. Identification of these alterations is useful not only for diagnosis but also for understanding the pathogenesis and potential targeted therapeutic implications of this rare tumour. However, in many routine diagnostic settings, especially where molecular testing is not readily available, diagnosis depends on careful correlation of clinical presentation, radiological findings, histomorphology, and immunohistochemistry.

The main differential diagnoses of PLNTY include oligodendroglioma, diffuse astrocytoma, dysembryoplastic neuroepithelial tumour, ganglioglioma, and other low-grade epilepsy-associated tumours.[6] Differentiating PLNTY from these entities is important because PLNTY generally follows an indolent course and may be adequately treated with surgical excision alone. Misclassification as an adult-type diffuse glioma may lead to unnecessary aggressive treatment, including radiotherapy or chemotherapy.

The present case is being reported because of its unusual occurrence in a 50-year-old male and its atypical bilateral frontal white matter involvement with extension into the genu of the corpus callosum. This case highlights the need to consider PLNTY in the differential diagnosis of calcified oligodendroglioma-like glial tumours even in older adults. Recognition of this rare entity is essential for accurate diagnosis, appropriate treatment planning, and prognostication.

CASE REPORT

A 50-year-old male with no significant past medical or neurological history presented to the Neurology outpatient department with recurrent episodes of generalized tonic–clonic seizures for the past two months. The seizures were sudden in onset, associated with transient loss of consciousness, tonic stiffening of all four limbs followed by clonic movements, tongue bite, and postictal confusion lasting approximately 15–20 minutes. There was no prior history of epilepsy, febrile seizures, head injury, central nervous system infection, stroke, or any chronic systemic illness such as hypertension or diabetes mellitus. There was also no family history of seizure disorder or intracranial neoplasms.

On clinical examination, the patient was conscious, oriented, and hemodynamically stable. Detailed neurological examination performed in the interictal period revealed no focal neurological deficits. Cranial nerve examination was normal, motor and sensory systems were intact, deep tendon reflexes were physiological, and cerebellar signs were absent. Fundoscopic examination did not reveal papilledema or evidence of raised intracranial pressure.

Routine laboratory investigations including complete blood count, liver function tests, renal function tests, serum electrolytes, blood glucose levels, coagulation profile, and viral serology were within normal limits. Electroencephalography demonstrated intermittent epileptiform discharges arising predominantly from the frontal region.

RADIOLOGICAL FINDINGS

Computed tomography (CT) scan of the brain revealed ill-defined non-enhancing hypodense lesions involving the bilateral frontal lobes, predominantly affecting the deep and subcortical white matter. The lesion extended across the midline into the genu of the corpus callosum. Multiple nodular as well as amorphous calcific foci were noted within the lesion, more prominently involving the right frontal lobe. No evidence of acute hemorrhage, significant surrounding edema, hydrocephalus, or substantial mass effect was identified.

The radiological differential diagnoses considered included low-grade glial neoplasm, oligodendroglioma, diffuse astrocytoma, and other calcified epilepsy-associated tumors. In view of the recurrent seizures and imaging findings suggestive

of a low-grade infiltrative neoplasm, the patient underwent surgical excision of the lesion for definitive diagnosis and management.

PATHOLOGICAL FINDINGS

Gross Examination

The specimen received in the Department of Pathology consisted of multiple irregular gray-white to gray-brown soft tissue fragments collectively measuring 3.5 × 2 × 1 cm. The cut surface appeared heterogeneous with focal gritty areas corresponding to calcification. No areas of hemorrhage or necrosis were grossly identified.

Microscopic Examination

Histopathological examination of hematoxylin and eosin-stained sections revealed a diffusely infiltrating low-grade glial neoplasm composed predominantly of relatively uniform small round monomorphic cells arranged in sheets and focally infiltrative patterns within the cerebral parenchyma. Tumor cells exhibited round to oval nuclei with finely granular chromatin and inconspicuous nucleoli. A characteristic perinuclear halo or clearing was observed around many tumor cells, imparting an oligodendroglioma-like appearance.

The tumor demonstrated low overall cellularity with a delicate arborizing network of thin-walled branching capillaries distributed throughout the lesion. Extensive calcification was one of the striking histologic features. Numerous psammomatous calcifications along with coarse dystrophic calcific deposits were diffusely scattered within the tumor. Focal microcystic change was also identified.

Importantly, there was no evidence of significant nuclear pleomorphism, brisk mitotic activity, necrosis, endothelial proliferation, or high-grade transformation. No Rosenthal fibers, eosinophilic granular bodies, or ganglion cells were identified. The adjacent brain parenchyma showed mild reactive gliosis.

Based on the morphologic features, a diagnosis of low-grade infiltrating glial neoplasm with oligodendroglioma-like morphology and prominent calcification was considered. However, in view of the unusual histologic pattern and differential diagnostic considerations, immunohistochemical analysis was performed for further characterization.

IMMUNOHISTOCHEMISTRY

Immunohistochemical examination demonstrated diffuse nuclear positivity for Olig2 within the tumor cells, confirming glial lineage. ATRX immunostaining showed retained nuclear expression, thereby arguing against astrocytic lineage-associated ATRX loss. Immunostaining for p53 revealed a wild-type expression pattern without significant overexpression.

IDH1 (R132H) immunostaining was negative in tumor cells, effectively excluding conventional IDH-mutant oligodendroglioma and diffuse astrocytoma. The Ki-67 proliferation index was very low, measuring less than 1%, supporting the indolent biological behavior of the lesion.

The immunohistochemical profile was summarized as follows:

Marker	Result
Olig2	Diffuse nuclear positivity
ATRX	Retained nuclear expression
p53	Wild-type expression
IDH1 (R132H)	Negative
Ki-67 labeling index	<1%

The absence of IDH1 mutation, retained ATRX expression, low proliferative activity, and characteristic oligodendroglioma-like morphology with extensive calcification strongly favored the diagnosis of Polymorphous Low-Grade Neuroepithelial Tumor of the Young (PLNTY), CNS WHO grade 1.

The patient had an uneventful postoperative recovery and remained seizure-free during short-term follow-up on antiepileptic medication. No evidence of tumor recurrence or progression was identified on follow-up neuroimaging.

DISCUSSION

Polymorphous low-grade neuroepithelial tumor of the young (PLNTY) is a recently recognized entity within the spectrum of low-grade epilepsy-associated neuroepithelial tumors (LEATs). Since its initial description by Huse et al., PLNTY has gained increasing recognition because of its unique histopathological and molecular characteristics. [9]

The tumor predominantly affects children and young adults and commonly presents with chronic epilepsy or recurrent seizures. However, the present case is unusual because the patient was a 50-year-old male, highlighting that PLNTY may also occur outside the typical pediatric and young adult age group.

Clinically, seizures are the most common presenting symptom in PLNTY because of the predominantly cortical location of these tumors.[9,10] Our patient presented with recurrent generalized tonic–clonic seizures without associated focal neurological deficits, which is consistent with previous reports. Broggi et al. described a similar rare adult case of PLNTY presenting with seizures, emphasizing that adult-onset PLNTY should be considered in the differential diagnosis of epilepsy-associated low-grade glial tumours. [11]

Radiologically, PLNTY usually appears as a cortically based lesion with prominent calcification and minimal or absent contrast enhancement. [12] in the present case, CT imaging revealed bilateral frontal hypodense lesions with nodular and amorphous calcifications extending into the genu of the corpus callosum. Although temporal lobe involvement is more frequently reported in literature, frontal lobe localization has also been documented. The bilateral frontal involvement with callosal extension observed in this case is relatively

uncommon and may mimic infiltrating diffuse glioma radiologically.

Histopathologically, PLNTY characteristically demonstrates oligodendroglioma-like morphology with diffuse infiltrative growth pattern, round monomorphic cells with perinuclear halos, branching capillary vasculature, and extensive calcification. [9,13] These features were well demonstrated in the present case. The absence of necrosis, endothelial proliferation, brisk mitotic activity, and marked atypia further supported the low-grade nature of the lesion. Similar histological findings were reported by Gupta et al., who emphasized the importance of recognizing extensive calcification and polymorphous architecture as distinguishing features of PLNTY. [10]

The major histopathological differential diagnosis in the present case was oligodendroglioma. Both entities share oligodendroglioma-like cytology and calcification; however, oligodendroglioma typically demonstrates IDH mutation and 1p/19q co-deletion. [14] In our case, immunohistochemistry showed negative IDH1 (R132H) staining and retained ATRX expression, effectively excluding oligodendroglioma and diffuse astrocytoma. The low Ki-67 proliferative index (<1%) also supported the indolent biological behavior of the tumor.

Immunohistochemically, PLNTY commonly expresses Olig2 and CD34 and usually lacks IDH mutation. [9,15] the present tumor showed diffuse Olig2 positivity with retained ATRX and wild-type p53 expression. Similar findings were documented by Pekmezci et al., who reported that PLNTY belongs to the expanding group of MAP kinase pathway–altered low-grade glioneuronal tumors.[15] Molecularly, PLNTY is characterized by alterations involving the MAP kinase pathway, particularly BRAF V600E mutation and FGFR2/FGFR3 fusions.[12,16] Although molecular analysis could not be performed in the present case, the characteristic histologic and immunophenotypic findings strongly supported the diagnosis.

The biological behavior of PLNTY is generally indolent, and most patients achieve favorable outcomes following complete surgical excision. [10,11] Surgical resection remains the treatment of choice, and recurrence is uncommon in completely excised lesions. The present patient also showed good postoperative recovery without radiological evidence of recurrence during follow-up. Recognition of PLNTY is clinically important because misdiagnosis as diffuse glioma may lead to overtreatment with unnecessary adjuvant radiotherapy or chemotherapy.

The present case expands the recognized clinicopathological spectrum of PLNTY and highlights the importance of considering this rare entity in the differential diagnosis of calcified oligodendroglioma-like tumours occurring in adults. Careful integration of radiological findings, histomorphology, immunohistochemistry, and molecular features is essential for accurate diagnosis and appropriate patient management.

CONCLUSION

PLNTY is a rare and recently recognized low-grade neuroepithelial tumour that predominantly affects younger individuals but may rarely occur in older adults. This case highlights an unusual presentation of PLNTY in a 50-year-old male presenting with new-onset seizures and bilateral frontal lobe involvement.

Awareness of this entity is essential because of its morphologic overlap with oligodendroglioma and other low-grade gliomas. Accurate diagnosis requires careful integration of radiologic, histopathologic, immunohistochemical, and molecular findings. Recognition of PLNTY prevents diagnostic pitfalls, avoids overtreatment, and facilitates appropriate prognostication and management.

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