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Polymorphous Low-Grade Neuroepithelial Tumor of the Young (PLNTY): A Newly Described Entity of Special Radiological Significance

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Abstract

Intracranial granulomas are a major cause of seizures in India, the most common etiologies being neurocysticercosis and tuberculosis. However, other pathologies including rare low-grade tumors may mimic these granulomas on imaging. In this article, we presented the case of a young woman patient with drug-resistant epilepsy. On imaging, there was a small calcified lesion in the brain parenchyma. In view of concordant electroclinical and imaging data on presurgical evaluation, the lesion was excised and the patient was seizure free. On histopathological evaluation, it was found to be a polymorphous low-grade neuroepithelial tumor of the young (PLNTY) – a rare, recently reported entity that can mimic an intracranial granuloma on imaging.

Keywords

- intracranial granuloma
- polymorphous low-grade neuroepithelial tumor of the young
- seizures

Introduction

Epilepsy is a common neurological disorder. Despite the availability of an increasing number of antiseizure medications, drug-resistant epilepsy is a challenging problem in a sizable number of patients. Calcified granulomas either secondary to neurocysticercosis (NCC) or tuberculosis are common causes of epilepsy, especially in the Indian subcontinent.[1,2] Recently a distinct entity, known as a polymorphous low-grade neuroepithelial tumor of the young (PLNTY) has been described, which has distinct morphological and molecular patterns.[3] However, the distinctive radiological features are yet to be defined. These tumors have a high rate of calcification[4] and can hence easily mimic granulomas, posing a diagnostic challenge to radiologists.

In this article, we discussed the case of a young woman patient with refractory seizures who was presumed to have an intracranial granuloma as an epileptogenic focus, based on neuroimaging, but on histopathology it was diagnosed as PLNTY.

Case Report

A 13-year-old adolescent girl child patient presented with drug refractory focal seizures that started at the age of 8 years

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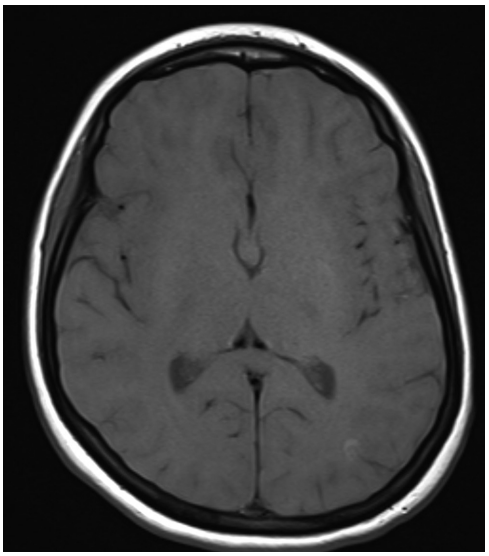

Fig. 1 T1 axial image at the level of the atrium of the lateral ventricles shows a slightly hyperintense T1 focus in the right parietal lobe cortex and subcortical white matter.

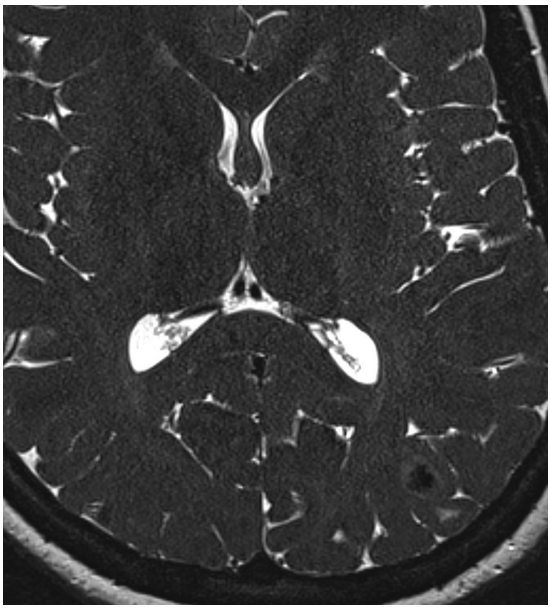

Fig. 2 T2 axial image shows mild T2 hyperintensity in the corresponding area.

as episodes of right-sided head and eye deviation. The seizures had increased in frequency and changed in semiology to weekly drop attacks with loss of awareness, over the last 2 to 3 years despite five antiseizure medications. Her early childhood history was remarkable for being the second of twins, born with low birth weight, and requiring neonatal intensive care unit (NICU) stay for neonatal hypoglycemia and neonatal seizures. Clinically, her growth and developmental milestones as well as scholastic performance were normal.

In view of the seizures being drug resistant, the patient underwent a presurgical evaluation. Long-term video electroencephalogram (EEG) monitoring recorded seven habitual seizures that were electro-clinically consistent with a left posterior temporoparietal focus. Magnetic resonance imaging (MRI) showed a rounded 1-cm area of signal abnormality in the left parieto-occipital region lateral to the parieto-occipital sulcus. There was intrinsic T1 shortening along the margins of this lesion with areas of susceptibility on susceptibility weighted images. With contrast administration, a faint peripheral enhancement was noted. No definite cystic component was identified. Given these features, a partly mineralized intracranial granuloma is a primary differential consideration (►**Figs. 1**–**4**).

As the localization based on electro-clinical and imaging data were concordant, she was offered surgery for the lesion with intraoperative electrocorticography (ECoG). Intraoperative ECoG showed frequent polyspikes and focal fast activity around the lesion. During resection, the surgeon reported a gritty feel of the resected lesion that could be consistent with calcification.

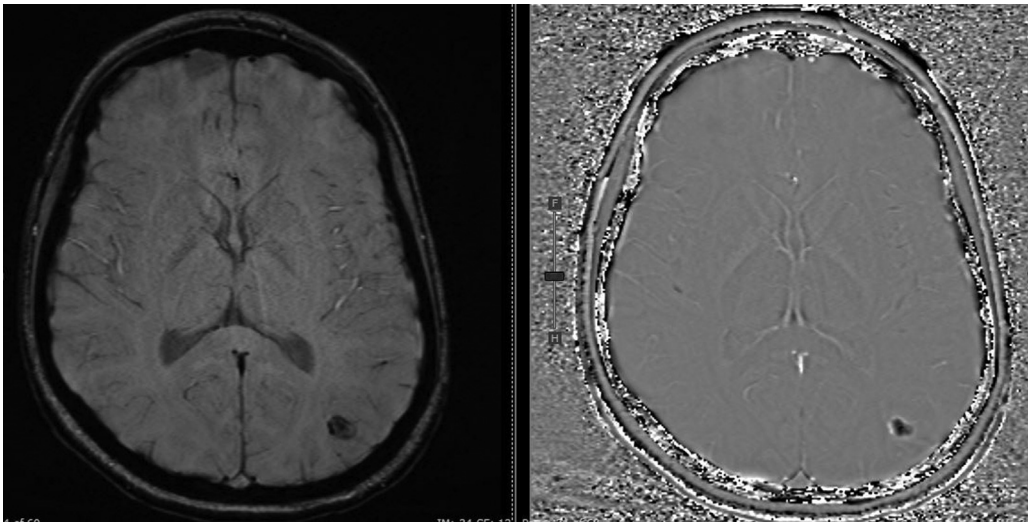

Fig. 3 Axial susceptibility weighted imaging (SWI) and phase images through the same level show a low signal area in the left parietal lobe on both images suggesting calcification.

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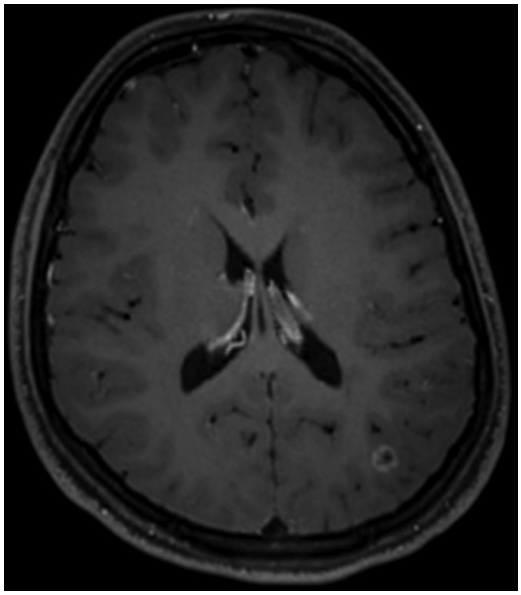

Fig. 4 Postcontrast image shows peripheral ring enhancement of this lesion.

However, histopathological analysis showed the presence of a low-grade diffuse glial neoplasm composed of oligodendrocytelike cells mixed with spindled astrocytes arranged in sheets, exhibiting moderate nuclear pleomorphism, with multiple foci of calcification. There were no Rosenthal fibers or granulomas. On immunohistochemical analysis, the tumor cells were positive for GFAP, CD34, and OLIG2, and negative for BRAF p.V600E, which was consistent with the diagnosis of PLNTY, NOS (not otherwise specified), CNS WHO grade 1 (►**Fig. 5**).

Discussion

Tumors presenting with drug-resistant seizures are termed as long-term epilepsy-associated tumors (LEATs). The commonly encountered tumors in this group are glioneuronal tumors (GNTs) including gangliogliomas (GGs) and dysembryoplastic neuroepithelial tumors, pleomorphic xanthoastrocytomas, and pilocytic astrocytomas. PLNTY, first described by Huse et al,[3] may share clinical features of other LEATs such as propensity for epileptogenicity. In 2016, Huse et al first reported a new entity named PLNTY and defined its distinct pathological features including the presence of oligodendrogliomalike cellular components, an infiltrative growth pattern, and intense CD34 immunopositivity.[3]

Histologically, the tumor needs to be differentiated from other subtypes of low-grade neuroepithelial tumors (LGNT). PLNTY shows abundant calcifications and CD34 expression,

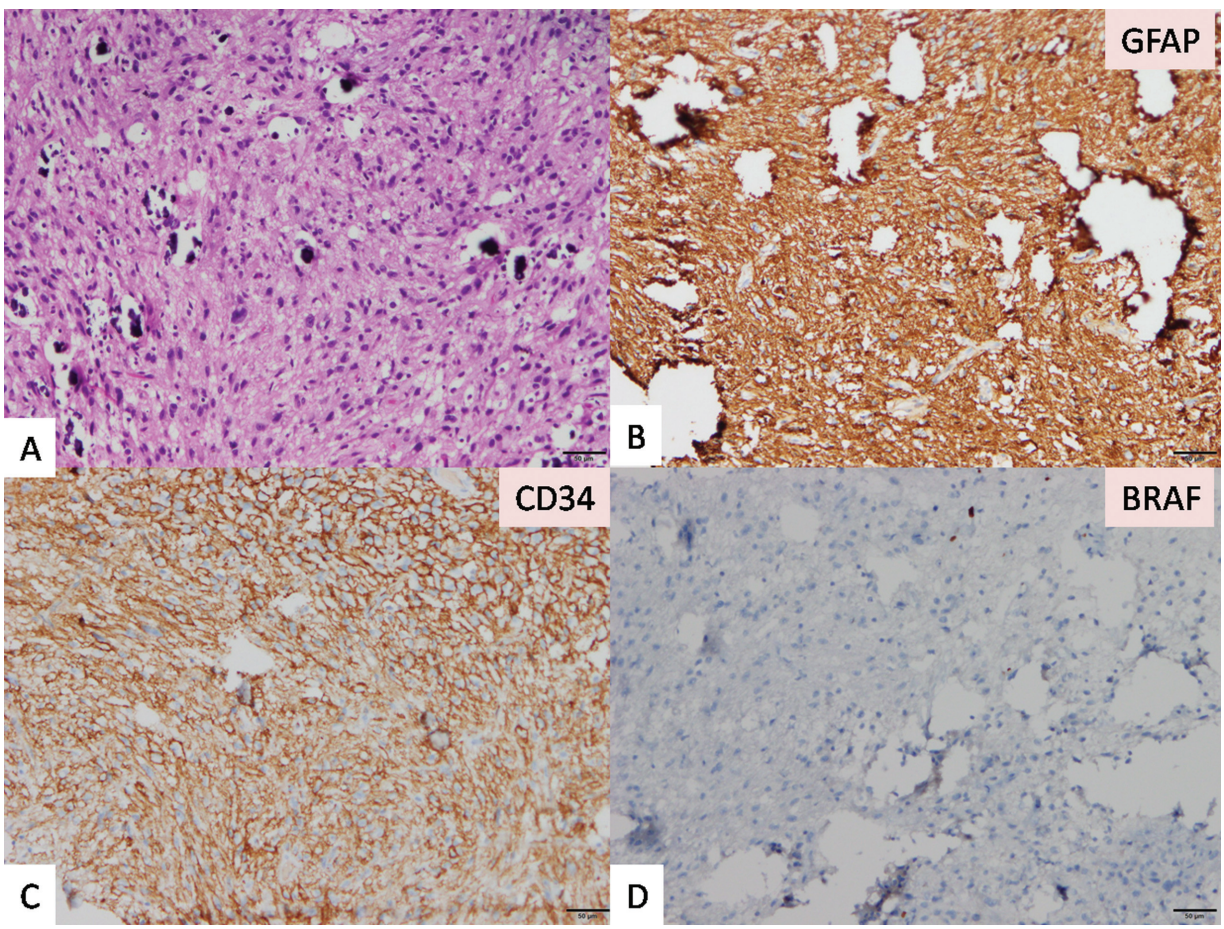

Fig. 5 (**A**) Photomicrographs show tumor composed of clear and polymorphic cells with calcification. The tumor cells are positive (**B**) GFAP and (**C**) CD34 and (**C**) negative for BRAFp.V600E.

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diffuse positivity of oligodendrocyte transcription factor 2 (OLIG2), and BRAF proto-oncogene, serine/threonine kinase (BRAF V600E), which are additional features of PLNTY with the absence of other features of LGNTs.[5] Multiple case reports have described this tumor to occur in patients between the ages of 4 and 32 years, with a median age of 17.5 years.[5]

Palejwala et al, in their study, found 29 reported cases of PLNTY. They stated that molecular profiles did not make differences in epilepsy-associated outcomes.[6]

Chen et al described the radiological features in a series of three patients of PLNTY as a solid or solid cystic mass mostly located in the temporal lobe followed by the occipital lobes with an unclear boundary with normal brain tissue.[7]

Benson et al described a temporal lobe PLNTY lesion in an adult with peripheral cystic change and calcifications.[5]

Macroscopic calcification is a common feature of PLNTY with a prominent pattern of central calcification.[4] They are often well circumscribed on T1- and T2-weighted MRI, with central hypointensity due to dense calcification and hyperintensity in the periphery of the calcification.

The different signal intensities on imaging due to calcifications leads to the distinctive "salt and pepper sign" which is used to describe PLNTY lesions is often attributed to the granulate mixed signals on T2-weighted images. The mixed signals are due to gritty calcification.[7] They also show postcontrast gadolinium enhancement and sometimes have cystic components.[4]

In our case, the oft-described "salt and pepper appearance" was not well appreciated probably due to the small size of the lesion. Also, the lesion was in the parietal lobe rather than in the more commonly reported temporal lobe. Clinically, the high frequency of drug-resistant seizures and the presence of intense electrical spiking around the lesion on the intraoperative ECoG were atypical of NCC, and more consistent with a dysplastic lesion.

Other differentials for calcified intracranial lesions other than the above-mentioned include that need to be considered are oligodendrogliomas, coccidiomycosis, toxoplasmosis, abscesses, and metastasis.[8]

The nodular calcified stage of NCC can have a similar appearance to PLNTY in the form of mild contrast enhancement surrounding a calcific focus, with minimal perilesional edema.[9]

Overall, this entity is sparsely described in the radiology literature. (It has not been reported in India in our search.). Awareness of this entity on imaging is especially important in India given the high prevalence of calcified granulomas that it can mimic.

Conclusion

In the Indian subcontinent with a high prevalence of tuberculosis and NCC, a granuloma is the most common cause of a calcified intracranial epileptogenic lesion. However, if there are atypical clinical and EEG features in the presence of a single calcified lesion in a patient with refractory epilepsy, an entity such as PLNTY needs to be considered.

Funding
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Conflict of Interest
None declared.

References

1 Shetty G, Avabratha KS, Rai BS. Ring-enhancing lesions in the brain: a diagnostic dilemma. Iran J Child Neurol 2014;8(03): 61–64
2 Reddy DS, Volkmer R II. Neurocysticercosis as an infectious acquired epilepsy worldwide. Seizure 2017;52:176–181
3 Huse JT, Snuderl M, Jones DT, 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(03):417–429
4 Johnson DR, Giannini C, Jenkins RB, Kim DK, Kaufmann TJ. Plenty of calcification: imaging characterization of polymorphous low-grade neuroepithelial tumor of the young. Neuroradiol 2019;61 (11):1327–1332
5 Benson JC, Summerfield D, Carr C, et al. Polymorphous low-grade neuroepithelial tumor of the young as a partially calcified intra-axial mass in an adult. Am J Neuroradiol 2020;41(04): 573–578
6 Palejwala AH, O'Neal CM, Quinton MR, Battiste JD, Peterson JEG, Dunn IF. Polymorphous low-grade neuroepithelial tumor of the young: rare tumor and review of the literature. Rare Tumors 2022; 14:20363613221083360
7 Chen Y, Tian T, Guo X, 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 (01):123
8 Mukherjee S, Das R, Begum S. Tuberculoma of the brain: a diagnostic dilemma: Magnetic resonance spectroscopy a new ray of hope. J Assoc Chest Phys 2015;3:3–8
9 Zhao J-L, Lerner A, Shu Z, Gao XJ, Zee CS. Imaging spectrum of neurocysticercosis. Radiol Infect Dis 2015;1(02):94–102

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