ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

www.surgicalneurologyint.com

Surgical Neurology International

Editor-in-Chief: Nancy E. Epstein, MD, Clinical Professor of Neurological Surgery, School of Medicine, State U. of NY at Stony Brook.

SNI: Neuro-Oncology

Editor
Mitsutoshi Nakada, MD
Kanazawa University, Ishikawa, Japan

SNI
Open Access

Case Report

Primary peripheral T-cell central nervous system lymphoma

Cylaina E. Bird[1], Jeffrey I. Traylor[1], Jenna Thomas[1], James P. Caruso[1], Benjamin Kafka[1], Flavia Rosado[2], Kyle M. Blackburn[3], Kimmo J. Hatanpaa[2], Kalil G. Abdullah[1]

Departments of [1]Neurological Surgery, [2]Pathology and [3]Neurology, The University of Texas Southwestern Medical Center, Dallas, Texas, United States.

E-mail: Cylaina E. Bird - cylaina.bird@utsouthwestern.edu; Jeffrey I. Traylor - jeffrey.traylor@utsouthwestern.edu; Jenna Thomas - jenna.thomas@utsouthwestern.edu; James P. Caruso - james.caruso@phhs.org; Benjamin Kafka - benjamin.kafka@phhs.org; Flavia Rosado - flavia.rosado@utsouthwestern.edu; Kyle M. Blackburn - kyle.blackburn@utsouthwestern.edu; Kimmo J. Hatanpaa - kimmo.hatanpaa@utsouthwestern.edu; *Kalil G. Abdullah - kalil.abdullah@utsouthwestern.edu

***Corresponding author:**
Kalil G. Abdullah,
Department of Neurological Surgery, University of Texas Southwestern Medical Center, Dallas, Texas, United States.

Kalil.abdullah@utsouthwestern.edu

Received : 06 March 2021
Accepted : 07 August 2021
Published : 13 September 2021

DOI
10.25259/SNI_224_2021

Quick Response Code:

ABSTRACT

Background: Primary peripheral T-cell central nervous system lymphoma (PCNSL) is a rare, aggressive tumor that arises in the craniospinal axis and has an increased risk in individuals who are immunocompromised. This lesion often mimics other benign and malignant processes on radiographic imaging, leading to misdiagnosis and delays in treatment. We present a case of a patient with a history of Sjögren's syndrome and progressive neurologic symptoms who underwent craniotomy for diagnosis.

Case Description: A 61-year-old woman with a history of Sjögren's syndrome, progressive aphasia, left facial droop, and right-sided paresthesias for 4 months presented for evaluation and management. An enhancing, infiltrative lesion in the left frontal lobe with underlying vasogenic edema was appreciated and suggestive of a primary or metastatic neoplasm. The patient underwent an open biopsy for further evaluation of the lesion. Extensive histopathologic evaluation revealed a diagnosis of T-cell PCNSL. The patient was started on induction methotrexate and temozolomide followed by consolidative radiotherapy.

Conclusion: Autoimmune conditions are a risk factor for T-cell PCNSL development. T-cell PCNSL has radiographic and gross histologic features that are consistent with a broad differential, including gliomas and inflammatory processes. Prompt diagnosis and extensive histopathological evaluation is essential to ensure appropriate treatment.

Keywords: Autoimmune disease, Glioma, Primary central nervous system lymphoma, Sjögren's syndrome

INTRODUCTION

Primary central nervous system lymphoma (PCNSL) is a rare lymphoma that accounts for 2–6% of all primary brain tumors.[1,3,4,9,11,14,23,24,31,33-35] Only 2% of these cases are known to be of T-cell origin.[1,2,4,6,11,13,15,17,21-23,25-29,31-35] T-cell PCNSL has less than a 20–30% 5-year survival rate.[14,35] Therefore, early and accurate diagnosis and treatment is imperative for best clinical outcomes. The presentation of T-cell PCNSL is challenging as it mimics other neoplastic and inflammatory diseases, including primary glial neoplasms. We present a case with a complex presentation of T-cell PCNSL and review literature relevant to the evaluation, diagnosis, and treatment of this uncommon pathology.

CASE REPORT

A 61-year-old woman with a medical history of Sjögren's syndrome and a long history of treatment with immunomodulatory therapy presented to clinic after recurrent and fluctuating stroke-like symptoms including aphasia, right-sided paresthesia, and left facial droop over a 4-month period. Her symptoms were initially attributed to an acute thromboembolic left middle cerebral artery stroke. However, examination revealed progressive expressive and receptive aphasia, right-sided House-Brackmann II facial droop, and decreased strength in the right upper extremity. Magnetic resonance (MR) imaging revealed an enhancing, infiltrative lesion in the left frontal lobe with progressive vasogenic edema in the left pre- and postcentral gyri [Figure 1]. MR spectroscopy displayed an elevation of choline, decrease in N-acetyl aspartate, and evidence of large lipid lactate peak suggestive of an intrinsic or lymphomatous neoplasm. This imaging was not consistent with a subacute infarct. The differential diagnosis for the lesion was lymphoma, infection, high-grade glioma, or progressive inflammation resulting in necrosis. Work-up for metastatic disease was negative.

Given the broad differential diagnosis and the progression of symptoms, the patient underwent a craniotomy with asleep motor mapping for open biopsy of the lesion. Intraoperatively, the lesion appeared ashen and gray with some areas of mineralization under the arachnoid. Histopathologic evaluation of the biopsy specimen demonstrated a small focus of perivascular lymphocytic infiltrate composed of predominately medium-sized atypical lymphoid cells in a background of histiocytes and small lymphocytes [Figure 2]. Areas of incomplete coagulation necrosis and extensive parenchymal and perivascular inflammatory changes were also noted. The atypical cells displayed irregular nuclear contours, inconspicuous nucleoli, and scant amount of cytoplasm. Immunohistochemical stains demonstrated that the atypical cells were CD3-positive, CD8-positive phenotypically aberrant T-cells with variably diminished expression of CD2 and CD5 and with complete loss of CD7. An Epstein–Barr virus (EBV) encoded RNA *in situ* hybridization study was negative for EBV. T-cell clonality polymerase chain reaction was positive for rearrangements in the Vg1-8 regions of the T-cell receptor gene. Infectious workup with special stains and immunostains revealed no evidence of *Toxoplasma* spp., varicella zoster virus, spirochetes, fungal, or bacterial infections. The findings indicated focal brain involvement by CD8-positive peripheral T-cell lymphoma (PTCL), clinically consistent with PCNSL. The postoperative period was uneventful. The patient was evaluated for systemic disease using FDG PET/CT scan. F18-FDG PET/CT scan did not demonstrate any FDG avid disease a month after biopsy and before treatment initiation. The patient was started on high-dose methotrexate (HD-MTX) with alternating temozolomide for 8 cycles. She had improvement in her aphasia and stability of her T2 fluid-attenuated inversion recovery signal with treatment [Figure 3]. She recently started consolidation therapy with radiation.

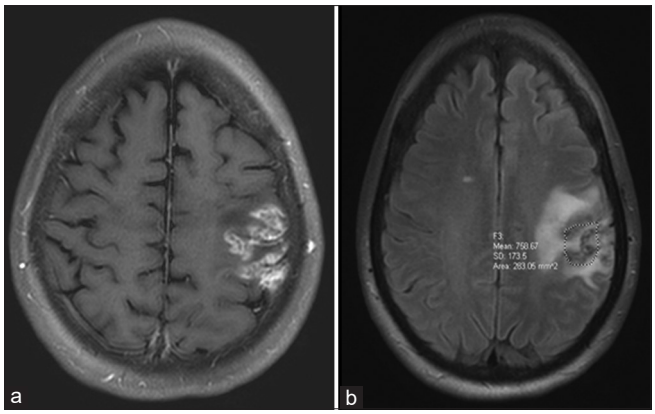

Figure 1: Neuroradiologic images on preoperatively. Representative axial T1-weighted postgadolinium contrast image (a) and axial T2-weighted FLAIR image (b).

DISCUSSION

T-cell PCNSL is an uncommon extranodal non-Hodgkin lymphoma in the craniospinal axis. It has an incidence of about 2% in Western countries, but is more commonly diagnosed in the east.[4-6,13,29,32] T-cell PCNSL often presents as a single supratentorial lesion.[4-6,10,11,18,23,33,35,36,38-41,43,44,46,53]

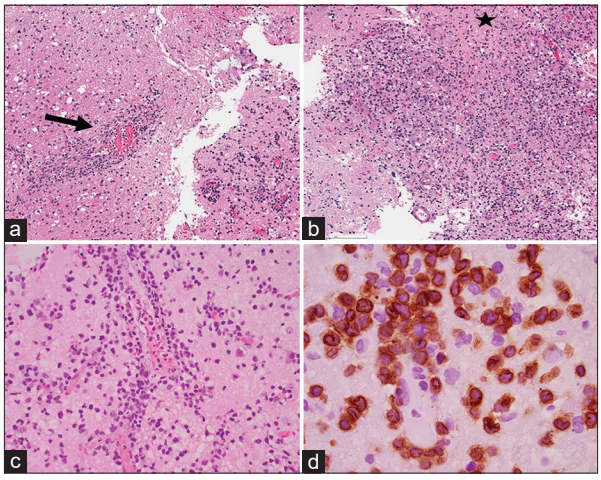

Figure 2: Histopathologic examination of the left frontal lobe lesion brain biopsy (a) and (b) hematoxylin-eosin (H&E, ×2) staining shows perivascular inflammatory infiltrates (arrow) and incomplete coagulation necrosis (star), (c) (H&E, ×10): perivascular inflammatory infiltrates of atypical medium-sized lymphoid cells and (d) (CD3 immunohistochemical stain, ×100 oil): the positive CD3 highlights the nuclear irregularities of the atypical cells and indicates the T-cell lineage of this process.

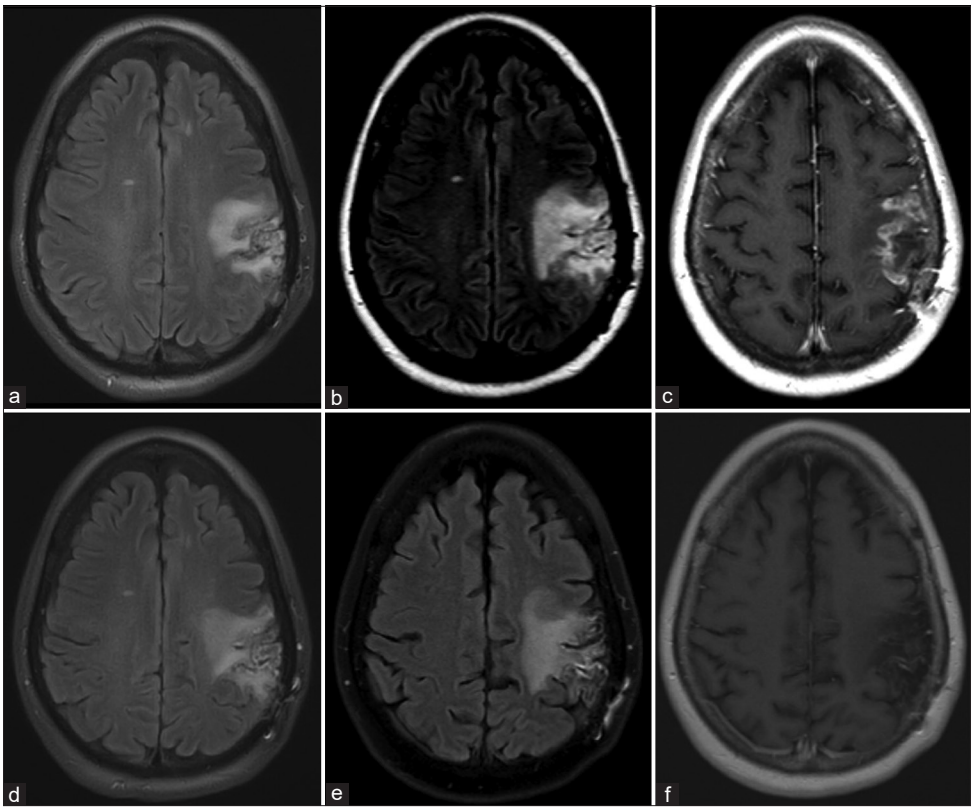

Figure 3: Follow-up neuroradiologic images. One-month postoperative representative axial T2-weighted FLAIR image (a), 2-month postoperative representative axial T2-weighted FLAIR image (b), 2-month postoperative representative axial T1-weighted postgadolinium contrast image (c), 4-month postoperative representative axial T2-weighted FLAIR image (d), 1-year postoperative representative axial T2-weighted FLAIR image with appreciated stable FLAIR signal (e), 1-year postoperative representative axial T1-weighted postgadolinium contrast image with no appreciated contrast-enhanced disease (f).

[Table 1] describes the available T-cell PCNSL cases in the current literature. The pathophysiologic origin of PCNSL is not well established. However, immune compromise is a known risk factor for PCNSL overall.[7,8,16] The patient presented in this report had a history of Sjögren's syndrome treated with immunomodulatory therapies, which may have predisposed her to develop T-cell PCNSL.

Gadolinium-enhanced MR imaging is the preferred radiographic study for PCNSL lesions.[8,21] PCNSL can be suspected when a lesion is isointense to hypointense as compared to gray matter on T2-weighted MR images.[8,11,14,41,55] T-cell PCNSL has heterogeneous enhancement after contrast administration, often interpreted as necrosis and hemorrhage, leading to a radiographic suggestion of glioma.[13,55] MR spectroscopy is helpful to radiographically distinguishing between an inflammatory process and a neoplastic process, such as glioma.[35] On MR spectroscopy, PCNSL lesions commonly display high lipid resonance, high lactate, and low N-acetyl aspartate.[41] It is also important to evaluate patients with suspected PCNSL for systemic involvement. Systemic lymphoma has been detected in 12.5% of patients diagnosed with PCNSL.[52] Thus, in accordance with current guidelines, systemic staging with PET/CT scans is important in suspected PCNSL cases to rule out systemic disease.[20,48] However, false-positive findings have been appreciated in about 4% of patients diagnosed with PCNSL who undergo this diagnostic imaging procedure.[52] Thus, the lower diagnostic yield of this modality should be considered carefully when evaluating patients with suspected PCNSL.[20,52]

Given the radiographic findings and clinical presentation, the differential diagnosis of our patient included gliomas, metastasis, subacute infarction, demyelinating diseases, and space-occupying infectious or parasitic lesions. The presentation of the patient may have been complicated by her history of intermittent high-dose corticosteroid use to help manage her autoimmune symptoms. Corticosteroids are a known lymphocytotoxic drug and can interfere with diagnosis when administered before biopsy.[8,14]

Diagnosis of PCNSL neoplasms is challenging, often requiring integration of clinical, radiologic, and histopathologic findings.[8,14] Histologic features of PTCL in the setting of PCNSL are illustrated by this case report

Table 1: Summary of published T-cell PCNSL cases.

Age (years)	Sex	CNS Site	Reference
56	Female	Bilateral cerebral hemispheres, brainstem, and wall of left lateral ventricle	Dulai *et al.* (2007)[13]
67	Male	Cerebellar lesion	Kleopa *et al.* (1996)[26]
43	Male	Cerebellar lesion	Villegas *et al.* (1997)[54]
37	Female	Cerebellar vermis lesion	Corns *et al.* (2010)[11]
42	Male	Cerebellar vermis lesion	Demetriades *et al.* (2003)[12]
53	Female	Left cerebellar lesion	Villegas *et al.* (1997)[54]
32	Male	Right and left cerebellar lesions	Bednar *et al.* (1991)[3]
63	Female	Right cerebellar lesion	Inoue *et al.* (1990)[22]
48	Female	Right cerebellar lesion	Knorr *et al.* (1992)[27]
60	Male	Right cerebellar peduncle lesion	McCue *et al.* (1993)[36]
16	Male	Right cerebral lesion	Ng *et al.* (1988)[42]
32	Female	Enhancement in third ventricle and ependyma of temporal horn	Nigo *et al.* (2016)[43]
26	Male	Intraventricular lesion	Splavski *et al.* (2016)[51]
40	Male	Left basal ganglia lesion	Kato *et al.* (2014)[25]
52	Female	Left basal ganglia lesion	Mineura *et al.* (1993)[38]
54	Male	Left caudate nucleus lesion	Provinciali *et al.* (1988)[46]
12	Female	Left frontal lesion	Shalabi *et al.* (2015)[49]
29	Female	Left frontal lesion	Zhao *et al.* (2017)[55]
79	Male	Left parietal lesion	Morisako *et al.* (2020)[40]
81	Male	Left temporo-occipital and paraventricular lesions	Gupta *et al.* (2017)[18]
32	Male	Right frontal lesion	Harder *et al.* (2003)[19]
63	Male	Right frontoparietal lesion	Goldbrunner *et al.* (1996)[16]
66	Female	Right frontoparietal lesion	Novak and Katzin (1995)[44]
23	Male	Right frontotemporal and right cerebellar lesions	Manenti *et al.* (2013)[35]
46	Male	Right parietal lesion	Takeshita *et al.* (1999)[53]
89	Female	Right periventricular lesion	Liu *et al.* (2003)
54	Male	Right superior temporal gyrus	Dulai *et al.* (2007)[13]
42	Female	Right temporal lesion	Dulai *et al.* (2007)[13]
13	Male	Right temporal lesion	Momota *et al.* (2015)[39]
67	Female	Multifocal lesions	Behbahani and Lyons (2011)[4]
56	Male	Multifocal lesions	Clark *et al.* (2010)[9]
56	Female	Multifocal lesions	Dulai *et al.* (2007)[13]
31	Female	Multifocal lesions	Liu *et al.* (2019)[30]
29	Male	Multifocal lesions	Lotan *et al.* (2012)[32]
36	Female	Multifocal lesions	Pulsoni *et al.* (1999)[47]
17	Male	No radiographic abnormality	Lai *et al.* (1991)[28]
64	Male	No radiographic abnormality	Lai *et al.* (1991)[28]
76	Female	Parasagittal lesion	Comes *et al.* (2019)[10]
10	Male	Parasagittal lesion	Gualco *et al.* (2010)[17]

PTCNSL: Peripheral T-cell central nervous system lymphoma

and include a prominent perivascular infiltrate of small- to medium-sized cells with irregular hyperchromatic nuclei, occasional distinct nucleoli, and scant cytoplasm.[4,13,18] The loosely scattered distribution of the mildly atypical perivascular cells accompanied by prominent reactive changes mimic inflammatory processes, making the differentiation between T-cell PCNSL and benign conditions difficult.[4,13,18,25,30] The utilization of immunohistochemical stains is useful to highlight the atypia and demonstrates phenotypic aberrancies that support the diagnosis of lymphoma. Initial immunohistochemical stains for CD3 and CD20 are a common approach to determine whether the neoplasm is of T-cell or B-cell origin, respectively.[13] Complete or partial loss of CD5 and CD7 is common phenotypic aberrancies; however, loss of CD3 is rare.[37] In most cases of T-cell PCNSLs, the neoplastic cells are CD8 positive and CD4 negative.[13,19,37] Molecular assays for T-cell receptor γ chain gene rearrangement can aid in the diagnosis of T-cell PCNSL in cases with equivocal morphologic and phenotypic abnormalities.[12,13,23,30,37]

T-cell PCNSL has a poor prognosis.[12] In a review of 45 patients with T-cell PCNSL, Shenkier *et al.* (2005) found that median progression-free survival was 22 months and

overall survival was 25 months.[50] Historically, whole-brain radiation therapy (WBRT) was the treatment of choice, as these lesions are very sensitive to radiotherapy.[10,14] However, this method causes a risk of severe neurotoxicity and is no longer the standard of care.[14] Recently, hippocampal sparing WBRT has been proposed and evaluated as a radiation method that limits the neurotoxicity.[45]

HD-MTX followed by radiotherapy is first-line treatment for PCNSL.[14,21] T-cell PCNSL patients with regimens that include HD-MTX have a longer survival compared to individuals who have nonmethotrexate-based chemotherapy regimens.[1,50] Multiple consolidation approaches include WBRT, additional chemotherapy, or autologous stem cell transplant.[8,18] Close follow-up is required due to most patients having a recurrence within 5 years of treatment.[8]

CONCLUSION

T-cell PCNSL is a rare, aggressive tumor in the craniospinal axis. The presentation of this lesion is variable and often includes progressive neurologic symptoms. As the radiographic and histologic features are often nonspecific and mimic other lesions, including inflammatory processes and high-grade gliomas, it is a challenging tumor to diagnosis. For patients with preexisting inflammatory conditions who present with progressive neurologic symptoms and inconclusive radiographic findings, T-cell PCNSL should be considered and a biopsy performed with subsequent immunohistochemical and molecular analysis of the specimen.

Acknowledgments

We wish to acknowledge Dr. Seema Jabbar, MD for her valuable contributions in the histopathologic evaluation of diagnosis in the case presented.

Declaration of patient consent

Institutional Review Board (IRB) permission obtained for the study.

Financial support and sponsorship

This research was funded by the Eugene P. Frenkel, M.D. Scholar in Clinical Medicine Endowment (KGA). The University of Texas Southwestern Medical Center – Dallas holds a Physician-Scientist Institutional Award from the Burroughs Wellcome Fund (CEB).

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Aaroe AE, Nevel KS. Central nervous system involvement of natural killer and T cell neoplasms. Curr Oncol Rep 2019;21:40.
2. Ahn SY, Kwon SY, Jung SH, Ahn JS, Yoo SW, Min JJ, *et al*. Prognostic significance of interim 11C-methionine PET/CT in primary central nervous system lymphoma. Clin Nucl Med 2018;43:e259-64.
3. Bednar MM, Salerni A, Flanagan ME, Pendlebury WW. Primary central nervous system T-cell lymphoma. Case report. J Neurosurg 1991;74:668-72.
4. Behbahani M, Lyons M. Primary central nervous system T-cell lymphoma of the brain. Open Neurosurg J 2011;4:62-5.
5. Bellinzona M, Roser F, Ostertag H, Gaab RM, Saini M. Surgical removal of primary central nervous system lymphomas (PCNSL) presenting as space occupying lesions: A series of 33 cases. Eur J Surg Oncol 2005;31:100-5.
6. Bhagavathi S, Wilson JD. Primary central nervous system lymphoma. Arch Pathol Lab Med 2008;132:1830-4.
7. Cheng G, Zhang J. Imaging features (CT, MRI, MRS, and PET/CT) of primary central nervous system lymphoma in immunocompetent patients. Neurol Sci 2019;40:535-42.
8. Chukwueke UN, Nayak L. Central nervous system lymphoma. Hematol Oncol Clin North Am 2019;33:597-611.
9. Clark AJ, Lee K, Broaddus WC, Martin MJ, Ghatak NR, Grossman CE, *et al*. Primary brain T-cell lymphoma of the lymphoblastic type presenting as altered mental status. Acta Neurochir (Wien) 2010;152:163-8.
10. Comes PC, André A, Nguyen-Khac F, Carpentier A, Bielle F, Amelot A. Intracranial cell lymphomas that mimic meningiomas: Case report to understand complex genetic, radiologic, and histopathologic entities. World Neurosurg 2019;125:339-42.
11. Corns R, Crocker M, Kumar A, Salisbury J, Tolias C, Sadler G, *et al*. Low grade cerebellar T-cell lymphoma: A novel response to treatment; a case report. Acta Neurochir (Wien) 2010;152:1075-7.
12. Demetriades A, McEvoy A, Galloway M, Kitchen N. Peripheral T-cell lymphoma presenting as a solitary vermian mass. Br J Neurosurg 2003;17:473-5.
13. Dulai MS, Park CY, Howell WD, Smyth LT, Desai M, Carter DM, *et al*. CNS T-cell lymphoma: An under-recognized entity? Acta Neuropathol 2008;115:345-56.
14. Ferreri AJ, Marturano E. Primary CNS lymphoma. Best Pract Res Clin Haematol 2012;25:119-30.
15. Fuentes-Raspall R, Solans M, Auñón-Sanz C, Sáez M, Marcos-Gragera R. Incidence and survival of primary central nervous system lymphoma (PCNSL): Results from the Girona cancer registry (199T 2013). Clin Transl Oncol 2018;20:1628-30.
16. Goldbrunner R, Warmuth-Metz M, Tonn JC, Vince GH, Roosen K. Primary Ki-1-positive T-cell lymphoma of the brain-an aggressive subtype of lymphoma: Case report and review of the literature. Surg Neurol 1996;46:37-41.
17. Gualco G, Wludarski S, Hayashi-Silva L, Filho PM, Veras G, Bacchi CE. Primary central nervous system peripheral T-cell lymphoma in a child. Fetal Pediatr Pathol 2010;29:224-30.
18. Gupta N, Nasim M, Spitzer SG, Zhang X. Primary central

nervous system T-cell lymphoma with aberrant expression of CD20 and CD79a: A diagnostic pitfall. Int J Surg Pathol 2017;25:599-603.
19. Harder A, Dudel C, Anagnostopoulos I, Hummel M, Brück W. Molecular genetic diagnosis of a primary central nervous system T cell lymphoma. Acta Neuropathol 2003;105:65-8.
20. Hoang-Xuan K, Bessell E, Bromberg J, Hottinger AF, Preusser M, Rudà R, *et al*. Diagnosis and treatment of primary CNS lymphoma in immunocompetent patients: Guidelines from the European association for neuro-oncology. Lancet Oncol 2015;16:e322-32.
21. Illerhaus G. III. Current concepts in primary central nervous lymphoma. Hematol Oncol 2015;33 Suppl 1:25-8.
22. Inoue M, Kawaguchi T, Yokoyama H, Shimada M. Primary T-cell lymphoma with myelopathy associated with HTLV-1. Neurosurgery 1990;27:148-51.
23. Jellinger KA, Paulus W. Primary central nervous system lymphomas-new pathological developments. J Neurooncol 1995;24:33-6.
24. Karschnia P, Batchelor TT, Jordan JT, Shaw B, Winter SF, Barbiero FJ, *et al*. Primary dural lymphomas: Clinical presentation, management, and outcome. Cancer 2020;126:2811-20.
25. Kato Y, Hayashi T, Kawai-Masaoka A, Ichimura T, Sasaki A, Uchino A, *et al*. Primary central nervous system cytotoxic T-cell lymphoma mimicking demyelinating disease. Intern Med 2014;53:1197-200.
26. Kleopa K, Becker G, Roggendorf W, Reichmann H. Primary T-cell lymphoma of the cerebellum. J Neurooncol 1996;27:225-30.
27. Knorr JR, Ragland RL, Stone BB, Woda BA, Gelber ND. Cerebellar T-cell lymphoma: An unusual primary intracranial neoplasm. Neuroradiology 1992;35:79-81.
28. Lai A, Wierzbicki A, Norman P. Immunocytological diagnosis of primary cerebral non-Hodgkin's lymphoma. J Clin Pathol 1991;44:251-3.
29. Lin TK, Yeh TH, Hsu PW, Chuang CC, Tu PH, Chen PY, *et al*. Primary central nervous system lymphomas of the brain: A retrospective analysis in a single institution. World Neurosurg 2017;103:550-6.
30. Liu XH, Jin F, Zhang M, Liu MX, Wang T, Pan BJ, *et al*. Peripheral T cell lymphoma after chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS): A case report. BMC Neurol 2019;19:266.
31. Long H, Li S, Zhang Y, Li R, Fong T, Yang C, *et al*. Primary central nervous system T-cell lymphoma: An analysis from the surveillance, epidemiology, and end results program. J Clin Neurosci 2020;79:74-9.
32. Lotan I, Khlebtovsky A, Inbar E, Strenov J, Djaldetti R, Steiner I. Primary brain T-cell lymphoma in an HTLV-1 serologically positive male. J Neurol Sci 2012;314:163-5.
33. Löw S, Han CH, Batchelor TT. Primary central nervous system lymphoma. Ther Adv Neurol Disord 2018;11:1-16.
34. Malikova H. Primary central nervous system lymphoma: Is whole-body CT and FDG PET/CT for initial imaging reasonable? Quant Imaging Med Surg 2019;9:1615-8.
35. Manenti G, di Giuliano F, Bindi A, Liberto V, Funel V, Garaci FG, *et al*. A case of primary T-cell central nervous system lymphoma: MR imaging and MR spectroscopy assessment. Case Rep Radiol 2013;2013:916348.
36. McCue MP, Sandrock AW, Lee JM, Harris NL, Hedley-Whyte ET. Primary T-cell lymphoma of the brainstem. Neurology 1993;43:377-81.
37. Menon MP, Nicolae A, Meeker H, Raffeld M, Xi L, Jegalian AG, *et al*. Primary CNS T-cell Lymphomas: A clinical, morphologic, immunophenotypic, and molecular analysis. Am J Surg Pathol 2015;39:1719-29.
38. Mineura K, Sawataishi J, Sasajima T, Kowada M, Sugawara A, Ebina K. Primary central nervous system involvement of the so called 'peripheral T-cell lymphoma'. Report of a case and review of the literature. J Neurooncol 1993;16:235-42.
39. Momota H, Kato S, Fujii M, Tsujiuchi T, Takahashi Y, Kojima S, *et al*. Primary peripheral T-cell lymphoma, not otherwise specified, of the central nervous system in a child. Brain Tumor Pathol 2015;32:281-5.
40. Morisako T, Shishido-Hara Y, Inaba T, Takeuchi H, Miyagawa-Hayashino A, Kodama Y, *et al*. Primary CNS CD45-depleted T-cell lymphoma: The first pathologically confirmed case. J Neuropathol Exp Neurol 2020;79:817-20.
41. Nabavizadeh SA, Vossough A, Hajmomenian M, Assadsangabi R, Mohan S. Neuroimaging in central nervous system lymphoma. Hematol Oncol Clin North Am 2016;30:799-821.
42. Ng HK, Lo ST, Poon CY, Poon WS. Primary cerebral T-cell lymphoma: A case report. Br J Neurosurg 1988;2:523-8.
43. Nigo M, Richardson S, Azizi E, Cohen JM, Shapira I. Ventriculitis caused by primary T-cell cns lymphoma in an immunocompetent patient. J Clin Oncol 2016;34:e145-7.
44. Novak JA, Katzin WE. Primary central nervous system T-cell lymphoma with a predominant CD8 immunophenotype. Cancer 1995;75:2180-5.
45. Oskan F, Ganswindt U, Schwarz SB, Manapov F, Belka C, Niyazi M. Hippocampus sparing in whole-brain radiotherapy. A review. Strahlenther Onkol 2014;190:337-41.
46. Provinciali L, Signorino M, Ceravolo G, Pasquini U. Onset of primary brain T-lymphoma simulating a progressive leukoencephalopathy. Ital J Neurol Sci 1988;9:377-81.
47. Pulsoni A, Gubitosi G, Rocchi L, Iaiani G, Martino B, Carapella C, *et al*. Primary T-cell lymphoma of central nervous system (PTCLCNS): A case with unusual presentation and review of the literature. Ann Oncol 1999;10:1519-23.
48. Scott BJ, Douglas VC, Tihan T, Rubenstein JL, Josephson SA. A systematic approach to the diagnosis of suspected central nervous system lymphoma. JAMA Neurol 2013;70:311-9.
49. Shalabi H, Angiolillo A, Vezina G, Rubenstein JL, Pittaluga S, Raffeld M, *et al*. Prolonged complete response in a pediatric patient with primary peripheral T-cell lymphoma of the central nervous system. Pediatr Hematol Oncol 2015;32:529-34.
50. Shenkier TN, Blay JY, O'Neill BP, Poortmans P, Thiel E, Jahnke K, *et al*. Primary CNS lymphoma of T-cell origin: A descriptive analysis from the international primary CNS lymphoma collaborative group. J Clin Oncol 2005;23:2233-9.
51. Splavski B, Muzevic D, Ladenhauser-Palijan T, Splavski B Jr. Primary central nervous system anaplastic large T-cell lymphoma. Med Arch 2016;70:311-3.
52. Suh CH, Kim HS, Park JE, Jung SC, Choi CG, Kim SJ. Primary

central nervous system lymphoma: Diagnostic yield of whole-body CT and FDG PET/CT for initial systemic imaging. Radiology 2019;292:440-6.
53. Takeshita M, Kubo O, Tajika Y, Kawamoto T, Hori T, Takakura K. Primary central nervous system T-cell lymphoma. Case report. Neurol Med Chir (Tokyo)1999;39:452-8.
54. Villegas E, Villà S, López-Guillermo A, Petit J, Ribalta T, Graus F. Primary central nervous system lymphoma of T-cell origin: Description of two cases and review of the literature. J Neurooncol 1997;34:157-61.
55. Zhao Q, Zeng LS, Feng XL, Zhang HM. Magnetic resonance imaging characteristics of primary central nervous system T-cell lymphoma. Chin Med J (Engl) 2017;130:374-6.

How to cite this article: Bird CE, Traylor JI, Thomas J, Caruso JP, Kafka B, Rosado F, *et al.* Primary peripheral T-cell central nervous system lymphoma. Surg Neurol Int 2021;12:465.