Primary peripheral T-cell central nervous system lymphoma

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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 histopathologic 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. Only 2% of these cases are known to be of T-cell origin. T-cell PCNSL has less than a 20–30% 5-year survival rate. 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.

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. T-cell PCNSL often presents as a single supratentorial lesion.
[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. 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. PCNSL can be suspected when a lesion is isointense to hypointense as compared to gray matter on T2-weighted MR images. T-cell PCNSL has heterogeneous enhancement after contrast administration, often interpreted as necrosis and hemorrhage, leading to a radiographic suggestion of glioma. MR spectroscopy is helpful to radiographically distinguishing between an inflammatory process and a neoplastic process, such as glioma. On MR spectroscopy, PCNSL lesions commonly display high lipid resonance, high lactate, and low N-acetyl aspartate. 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. Thus, in accordance with current guidelines, systemic staging with PET/CT scans is important in suspected PCNSL cases to rule out systemic disease. However, false-positive findings have been appreciated in about 4% of patients diagnosed with PCNSL who undergo this diagnostic imaging procedure. Thus, the lower diagnostic yield of this modality should be considered carefully when evaluating patients with suspected PCNSL.

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.

Diagnosis of PCNSL neoplasms is challenging, often requiring integration of clinical, radiologic, and histopathologic findings. 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)[15]                        |
| 67          | Male  | Cerebellar lesion                                                         | Kleopa et al. (1996)[23]                      |
| 43          | Male  | Cerebellar lesion                                                         | Villegas et al. (1997)[24]                    |
| 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                                                   | Liu et al. (2000)[25]                         |
| 32          | Male  | Right and left cerebellar lesions                                        | Bednar et al. (1991)[26]                      |
| 63          | Female| Right cerebellar lesion                                                  | Inoue et al. (1990)[27]                       |
| 48          | Female| Right cerebellar lesion                                                  | Knorr et al. (1992)[28]                       |
| 60          | Male  | Right cerebellar peduncle lesion                                         | McCue et al. (1993)[29]                       |
| 16          | Male  | Right cerebral lesion                                                    | Ling et al. (1988)[30]                        |
| 32          | Female| Enhancement in third ventricle and ependyma of temporal horn             | Nigo et al. (2016)[31]                        |
| 26          | Male  | Intraventricular lesion                                                  | Splavski et al. (2016)[32]                    |
| 40          | Male  | Left basal ganglia lesion                                                | Kato et al. (2014)[33]                        |
| 52          | Female| Left basal ganglia lesion                                                | Mineura et al. (1993)[34]                     |
| 54          | Male  | Left caudate nucleus lesion                                              | Provincia et al. (1988)[35]                   |
| 12          | Female| Left frontal lesion                                                      | Shalabi et al. (2015)[36]                     |
| 29          | Female| Left frontal lesion                                                      | Zhao et al. (2017)[37]                        |
| 79          | Male  | Left parietal lesion                                                     | Morisako et al. (2020)[38]                    |
| 81          | Male  | Left temporo-occipital and paraventricular lesions                       | Gupta et al. (2017)[39]                       |
| 32          | Male  | Right frontal lesion                                                     | Harder et al. (2003)[40]                      |
| 63          | Male  | Right frontoparietal lesion                                              | Goldbrunner et al. (1996)[41]                 |
| 66          | Female| Right frontoparietal lesion                                              | Novak and Katzin (1995)[42]                   |
| 23          | Male  | Right frontotemporal and right cerebellar lesions                        | Manenti et al. (2013)[43]                     |
| 46          | Male  | Right parietal lesion                                                    | Takeshita et al. (1999)[44]                   |
| 89          | Female| Right periventricular lesion                                             | Liu et al. (2003)                             |
| 54          | Male  | Right superior temporal gyrus                                            | Dulai et al. (2007)[15]                       |
| 42          | Female| Right temporal lesion                                                    | Dulai et al. (2007)[15]                       |
| 13           | Male | Right temporal lesion                                                    | Momota et al. (2015)[45]                      |
| 67          | Female| Multifocal lesions                                                       | Behbahanian and Lyons (2011)[46]              |
| 56          | Male  | Multifocal lesions                                                       | Clark et al. (2010)[47]                       |
| 56          | Female| Multifocal lesions                                                       | Dulai et al. (2007)[15]                       |
| 31          | Female| Multifocal lesions                                                       | Liu et al. (2019)[48]                         |
| 29          | Male  | Multifocal lesions                                                       | Lotan et al. (2012)[49]                       |
| 36          | Female| Multifocal lesions                                                       | Pulsoni et al. (1999)[50]                     |
| 17          | Male  | No radiographic abnormality                                              | Lai et al. (1991)[51]                         |
| 64          | Male  | No radiographic abnormality                                              | Lai et al. (1991)[51]                         |
| 76          | Female| Parasagittal lesion                                                      | Comes et al. (2019)[52]                       |
| 10          | Male  | Parasagittal lesion                                                      | Gualco et al. (2010)[53]                      |

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. 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. 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. Complete or partial loss of CD5 and CD7 is common phenotypic aberrancies; however, loss of CD3 is rare. In most cases of T-cell PCNSLs, the neoplastic cells are CD8 positive and CD4 negative. 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.

T-cell PCNSL has a poor prognosis. 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.[8] 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.[6]

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.[1,30] 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.

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**Declaration of patient consent**

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

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**Conflicts of interest**

There are no conflicts of interest.

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