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# Dural Marginal Zone Lymphoma Presented as Brain Herniation: A Case Report

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## Abstract

Marginal zone lymphoma (MZL) is a rare primary Dural neoplasm with distinct histopthological features and challenging radiological appearances. The authors are reporting a case of a primary Dural lymphoma that was initially mimicked a subdural hematoma presented with new onset headache and visual changes. Ophthalmological exam revealed bilateral grade III papilledema that warranted urgent imaging. Initial CT scan without contrast was interpreted as sub-acute subdural hematoma. Based on CT findings, MRI was requested which revealed an enhancing Dural mass for which surgery was performed. Histopathology revealed marginal zone lymphoma of the Dura. All further work up was negative for systemic lymphoma.

**Keywords: Primary dural lymphoma (PDL); Primary central nervous system lymphoma (PCNSL); Marginal zone lymphoma (MZL)**

## Case Presentation

A 38 year old, African American female presented to the primary care physician with a chief complaint of new onset headache and blurred vision of three months duration. The headache had worsened during the past two weeks. She also had visual changes, described as a curtain going down with flashes of light, and reported attacks of gait imbalance and left facial droop. The patient was referred to Ophthalmology clinic for further assessment, which revealed bilateral grade III papilledema. The Ophthalmologist referred the patient to the emergency department for further neurological evaluation and brain MRI. Initial non-contrast brain CT scan showed a left crescent-shaped slightly hyper dense lesion causing rightward midline shift resulting in subfalcine and uncal herniation which was interpreted as a subacute subdural hematoma (Figure1). Neurosurgery was consulted and Brain MRI was performed which showed a dural based lobulated T2WI isointense mass with trapped CSF (Figure 2) homogenous avid post contrast enhancement (Figure 3 A & B), and restricted diffusion (Figure 4 A & B) There were no associated calvarial bony changes or adjacent parenchymal vasogenic edema. The differential diagnosis based on MRI was atypical meningioma, hemangiopericytoma or Dural metastasis. Patient underwent craniotomy and resection of the mass and histopathology revealed diffuse lymphoid infiltrate with regressed germinal centers of varying sizes (Figure 5) with predominant CD20 B cells and fewCD3 positive T cells (Figure 6). CD20 highlights the B cells within and between the follicles (Figure 7). These findings are consistent with marginal zone lymphoma and the presence of germinal centers suggests Nodal type (Figure 7). The patient underwent CT neck, chest, abdomen and pelvis to evaluate for systemic disease, which was negative. Extensive laboratory work up including CSF analysis, viral screen and bone marrow biopsy were also all negative for systemic lymphoma.

## Discussion

Dural masses result from variable tumors and disorders and maybe either primary or secondary. The most common primary dural neoplasm is meningioma followed by hemangiopericytma. Secondary neoplasms of the dura result from intracranial masses or systematic malignancy [1]. PDL is a rare subtype of PCNSL. Primary CNS lymphoma is neoplastic process of leptomeninges, brain, spinal cord, ventricles and eyes without evidence of nodal or extra nodal disease outside CNS. Secondary CNS involvement by lymphoma is much more common [2].

PCNSL can occur in both immune compromised and immune competent patients and represents approximately 2.7% of all primary and secondary CNS malignancies. PDL accounts for less than

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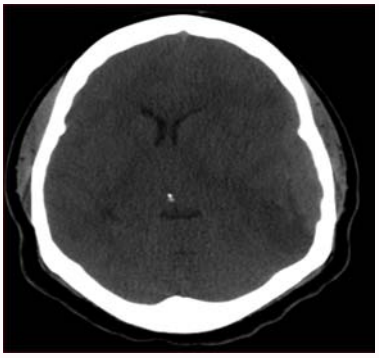

**Figure 1:** Axial non contrsted Brain CT Scan: Left elliptical shaped isodense extraxial lesion( Arrow).

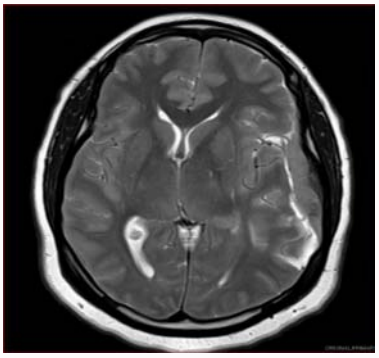

**Figure 2:** 3T Brain MRI Axial T2 WI: Left convexity elliptical extra axial isointense lesion(Arrow).

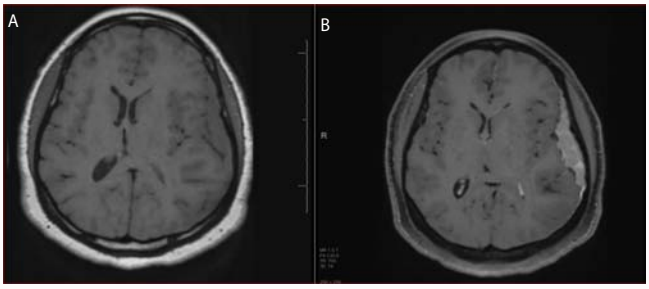


**Figure 3:** (A) 3T Brain MRI Axial T1 WI without Gadolinium: Isointense left convexity extra axial lesion ( Arrow). (B) 3T Brain MRI Axial T1 WI post Gadolinium: Homogenous avidly enhancing left convexity extra axial lesion.

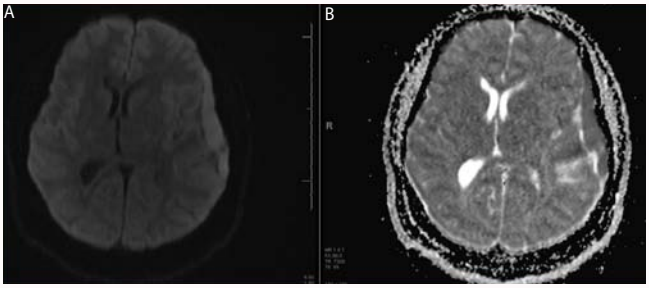


**Figure 4:** (A**)** 3T Brain MRI DWI sequence. (B**)** ADC Map Sequence. Restricted diffusion pattern seen in the left convexity extra axial lesion ( Arrows ).

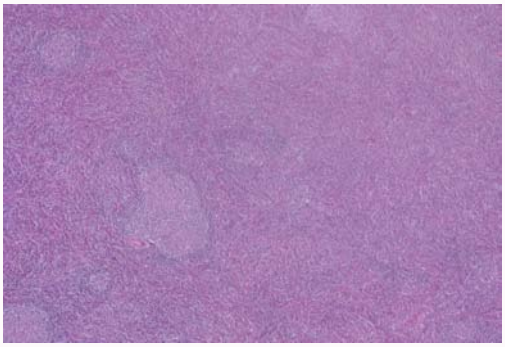

**Figure 5:** Diffuse Lymphoid infiltrate with regressed germinal centers of varying sizes (Arrow) (x4 magnification).

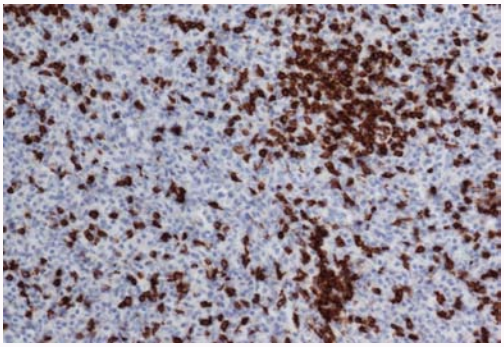

**Figure 6:** CD3 positive T cells are much fewer than CD20 B cells (x20).

1% of all CNS lymphomas [3,4]. Bhagavathi et al [5]. reported an increased incidence of PCNSL over the past three decades. It is more common in middle-aged females with a male-to-female ratio of 1:2 to1:7 and a median age of 56 years. It tends to occur in a younger age group in immune compromised patients [3,4]. The etiological factors predisposing to PCNSL are not completely understood; the only known risk factor is congenital or acquired immunodeficiency [6].

PCNSL is subdivided based on histopathological findings. Diffuse large B-cell lymphoma, accounting for 90%, is most common followed by T-cell lymphoma and then Marginal zone lymphoma [7]. PCNSL is solitary in 65% and multifocal in 35% of cases [4].

Lymphoma that arises from the dura is low-grade B-cell marginal zone lymphoma. MZL is a non-Hodgkin lymphoma, which most commonly occurs in the gastrointestinal tract where it called "mucosa associated lymphoid tissue (MALT). It is treated differently and has a different prognosis than other types of PCNSL [8].

The pathogenesis of the PDL is not totally understood, as the Dura is free of any lymphoid tissue [6]. Few presumptive theories were suggested (a) Dural lymphoma is the Consequence of seeding from an undiagnosed systemic MALT lymphoma; (b) meningothelial cells at the arachnoid membrane and Dural venous sinuses are embryologically similar to epithelial cells at other sites in which MALT lymphomas arise, and (c) Attraction of the polyclonal lymphocytes by inflammatory conditions of the Dura can give rise to MALT lymphoma [9,10].

Dural lymphomas appear as hyper- or isoattenuated masses on CT due to high cellularity and almost all demonstrate iodinated contrast enhancement. Negative CT scan does not rule out PCNSL [11,12]. On MRI, PDL shows iso- to hypointensity on T2WIand vivid post Gadolinium enhancement sometimes with heterogeneity [13]. It is easy to mistake for meningioma or subdural hematoma as these both show hyperdensity on CT scan [13]. MR imaging utilizing

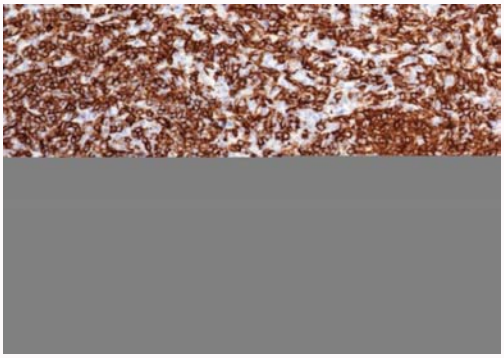

**Figure 7:** CD20 highlights the B cells within and between the follicles (x20).

DWI perfusion as lymphoma demonstrates diffusion restriction due to high cellularity and spectroscopy manifested by lipid peak and increased Cho/Cr ratio might have potential to aid the differentiation between lymphoma and other dural tumors [14].

PDL has a favorable prognosis compared to parenchymal PCNSL subtypes or Dural metastasis of systemic lymphoma. Due to the rarity of the disease, yet there is no standardized treatment plan; however, disease limited to a single site has a good response to local treatment such as surgery or focal radiation [15]. In conclusion, MZL is a rare tumor with vague nature of the clinical and the radiological characteristics. The author's main objective in this case is to raise the level of awareness and preparedness of the radiologists about this dural tumor that warrants further workup to plan proper management for such cases.

The uniqueness of this case report comes from the unusual presentation, which in this case was brain herniation and was initially interpreted as subdural hematoma despite of the absence of trauma and the young age of the patient. This case also demonstrates the importance of proper communication and cooperation between specialties and its critical role in providing high-quality service. Finally as radiologist we owe it to our patients and ourselves to practice evidence-based medicine to support our clinical decisions which ultimately will result in a better patient`s care.

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