Primary dural lymphoma: a comprehensive literature review and report of a case

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ABSTRACT
Primary dural lymphoma (PDL) is a subtype of primary central nervous system (CNS) lymphoma (PCNSL) with only an extra-axial dural location. It accounts for less than 1% of all CNS lymphomas. PDL is a sporadic CNS tumor, and in the preoperative period, because of imaging characteristics, it is usually mimicking a meningioma. Usually, PDL is a low-grade B-cell lymphoma with a relatively good response to surgical resection with or without radiotherapy. Here we reviewed 102 case reports of PDL in the literature. Then, we present the case of our patient with PDL and explain the complexity of our treatment approach.

Key words: primary dural lymphoma, review, meningioma, CNS

Introduction
Primary central nervous system (CNS) lymphoma (PCNSL) is an extranodal non-Hodgkin lymphoma of the CNS without systemic involvement [1]. Primary dural lymphoma (PDL) is a subtype of PCNSL with only extra-axial dural location; it accounts for less than 1% of all CNS lymphomas [2].

PDL is a sporadic CNS tumor, and in the preoperative period, because of imaging characteristics, it is usually mimicking a meningioma [3]. On the one hand, meningioma is the most common primary brain tumor, and on the other hand, a dural-based PCNSL is a rare intracranial tumor. Misdiagnosis could be a prevalent problem in treating patients with PDL [4, 5].

Nowadays, the mainstay of treatment for patients with PCNSL is gaining a tissue diagnosis (e.g., stereotactic biopsy), followed by high-dose methotrexate (MTX) induction chemotherapy and then consolidation therapy (e.g., whole-brain radiotherapy, WBRT) to treat the residual tumor and improve overall survival [6]. Usually, PDL is a low-grade marginal zone B-cell lymphoma (MZL) with a relatively good response to surgical resection with or without radiotherapy [7].

Here we present our patient with PDL, explain the complexity of our treatment approach, and review all reported PDL cases in the literature.

Methods
We obtained and reviewed data related to all reported cases of PDL in the literature, retrospectively. We did not exclude any case reports. All reported cases, even those with limited data, were included, but we reported missing data as not available (N/A). We found 35 case reports and case series in the literature that reported on patients with PDL. Also, we reported our patient with PDL to explain the possible complexity in facing a patient with PDL.
Results

We reviewed overall 30 men and 71 women with PDL aged 19–85 years (mean 53.85 years old). Detailed data of all patients are included in Table 1. Convexity (61%), cavernous (7.9%), and tentorial (7.9%) were the most common sites of PDL, followed by other sites (7.9%) and the falcine (6.9%). Ninety percent of all reported cases had a single lesion. Considering the histologic type of reported PDL, the most common types were marginal zone lymphoma (62.3%), diffuse large B-cell lymphoma (23.7%), B-cell lymphoma (not determined subgroup) (6.9%), and Follicular cell lymphoma (3.9%). The most common signs and symptoms in patients were headache (33.6%), followed by seizure (27.7%), cranial nerve deficits (18.8%), and visual deficit (16.8%). The most common treatment approach in the literature was surgical resection and radiotherapy (31.6%), followed by surgical resection and chemotherapy (16.8%). All detailed results of this review can be seen in Table 2.

An illustrative case

A 56-year-old female was admitted to the emergency department with acute loss of consciousness (LOC). On neurological examination, she had a Glasgow coma scale (GCS) score of 12 with bilateral reactive pupils. She had no neurological deficit. A computed tomographic (CT) scan of the brain revealed a bilateral frontal parasagittal mildly hyperdense extra-axial mass with extensive bi-frontal brain edema (Fig. 1A, B). On magnetic resonance imaging (MRI) of the brain and venography (MRV), the mass resembled a giant parasagittal meningioma with a completely occluded anterior superior sagittal venous sinus (SSS) (Fig. 1C–H); because of inappropriate bi-frontal edema in comparison to the lesion size, cortical vein thrombosis was considered as an important concomitant event, preoperatively. After a short course of treatment with intravenous (IV) anticoagulant and hyper-hydration, the patient was scheduled for surgical resection of the tumor.

In the operating room, the patient was placed in the supine position, and we approached her via bi-coroinal incision and bi-frontal one-piece craniotomy. The tumor was an extra-axial parasagittal meningioma, which was resected gross totally. Adjacent anterior SSS, falx and dura were resected simultaneously. The dura was replaced with a pedunculated pericranial galeal patch. The adjacent parasagittal brain was very fragile and pale, but we could dissect it from the tumor well. After surgery, the patient felt good with no deficit and had GCS 13. On the first post-operation day, the patient acutely deteriorated, and with GCS 5 and right fixed mydriasis and severe bi-frontal malignant edema on a CT scan (Fig. 2B, C), she underwent an emergent decompressive craniectomy. Then, the patient felt good and was discharged with GCS 15 and no deficit on the 10th post-operation day. After one month, she was scheduled for bone flap replacement. The histopathologic exam revealed a low-grade B-cell lymphoma (not determined subgroup) (Fig. 3). Our radio-oncologist treated the patient as a PDL based on negative results for systematic involvement with lymphoma, i.e., negative chest and abdominopelvic CT scan (a course of focal radiotherapy). On one year follow-up, the patient felt good with no sign of recurrence on the following brain MRI (Fig. 2D, E).

Discussion

PDLs have distinct clinical-pathological entities that separate them from intraparenchymal lymphoma. They are primarily low-grade and have a favorable prognosis. They include marginal zone lymphoma (MZL), small cell lymphoma, diffuse large B-cell lymphoma (DLBCL), and lymphoblastic and follicular cell lymphoma, in order of prevalence [8].

The pathogenesis of PDL is not well understood due to the lack of lymphoid tissue in the dura. There are 2 hypotheses for their development. One is the presence of meningotheial cells throughout the arachnoid membrane, and the other is chronic environmental antigenic stimulation, and resultant inflammatory conditions, which could precede the malignant transformation of lymphoid cells [9–11].

Meningothelial cells are analogous to epithelioid cells at other sites where MZL arises. These cells are present in the arachnoid membrane but are concentrated in the arachnoid villi with dural venous sinuses. Interestingly, most case reports localized MZL lymphoma in these regions [12–14].

However, a previous history of primary autoimmune disease, meningeal infiltration, or infection was primarily absent in our literature review. A limited number of case reports with underlying autoimmune diseases were reported, including Hashimoto thyroiditis, Grave’s disease, Sjogren’s syndrome, systemic lupus erythematosus (SLE), and relapsing-remitting multiple sclerosis (MS) [15–20].

Clinical presentation and diagnostic findings

PDL, especially MZL, most of the time presents as a single mass lesion without systemic involvement and has insidious long-lasting symptoms; there were only 7 case reports with multiple lesions which originated from the convexity [11].
| Age (year)/gender | Symptoms at onset                                             | Lesion location                                      | Number of lesions | Systemic Pathology | Treatment | Follow up |
|------------------|---------------------------------------------------------------|------------------------------------------------------|-------------------|-------------------|-----------|-----------|
| de la Fuente et al., 2017 | Generalized tonic-clonic seizure | Lt tentorium | 1 | One had CSF+ and one patient had para-aortic lymphadenopathy and bilateral lung nodules | MZL | STR + Focal RT | 12.1y |
| 47M | Generalized tonic-clonic seizure | Lt tentorium | 1 | | MZL | STR + Focal RT | 1.1y |
| 66M | Seizures, progressive gait disorder | Lt tentorium | 1 | | MZL | GTR + Focal RT | 7.2y |
| 41F | Headache, focal seizures, Rt visual field cut | Lt tentorium | 1 | | MZL | STR + WBRT + Focal RT | 11.3y |
| 51F | Long h/o headache with new worsening | Lt frontal | 1 | | MZL | STR + Focal RT | 6.8y |
| 49F | FocalLt face numbness and paresthesiaLt ear tinnitus and otalgia | Lt tentorium (compressingL brainstem and cerebellar hemisphere) | 1 | | MZL | Biopsy + chemotherapy | 8.6y |
| 69F | Walking difficulty | Lt temporo-parietal convexity | 1 | | MZL | Biopsy + WBRT + Focal RT | 5.7 y |
| 51F | Seizure, Lt homonymous hemianopsia | Rt occipital | 1 | | MZL | GTR of both lesions + NA | NA |
| 41F | Headache, focal seizures, Lt parieto-occipital | Lt parieto-occipital | 1 | | MZL | STR + WBRT + Focal RT | 4.6y |
| 51F | Headache | Rt temporal, R frontal | 2 | | MZL | STR + NA | NA |
| 49F | Focal seizures | Rt fronto-parietal | 1 | | MZL | STR + WBRT + Focal RT | 0.9y |
| 72F | Generalized tonic-clonic seizure | Rt temporo-parietal | 1 | | MZL | Partial resection + WBRT + Chemotherapy | 17.2y |
| 33F | Generalized tonic-clonic seizure, Lt facial weakness | Lt frontal and Lt parietal | 2 | | MZL | STR of sphenoid lesion only + WBRT + Focal RT | 9m |
| 39F | Vision loss, focal R-sided paresthesia | Lt frontal, Rt sphenoid region/orbital apex | 2 | | MZL | Biopsy + WBRT | 3.1y |
| 49F | Headache, seizures, visual loss | Rt temporal, Rt frontal | 2 | | MZL | STR + Focal RT | 1.3y |
| 39F | Headache | Rt frontal | 1 | | MZL | STR + Focal RT | 1.3y |
| 47M | Seizures | Lt frontal | 1 | | MZL | STR + Focal RT | 5.3y |
| 34M | Seizures | Rt temporal | 1 | | MZL | Biopsy + Focal RT | 4.7y |
| 67M | Seizures | Rt frontal | 1 | | MZL | Biopsy + focal RT | 3y |
| 51F | Focal paresthesia and numbness | Lt cavernous sinus | 1 | | MZL | GTR + Focal RT | 2.5y |
| 57M | Seizures, headache | Rt temporal | 1 | | MZL | Biopsy + Focal RT | 1.9y |
| 59F | Headache | Supraocular region | 1 | | MZL | Biopsy + Chemotherapy | 10m |
| 48F | Cranial nerve palsy | Cavernous sinus (bilateral) | M | | MZL | GTR + Focal RT | 8m |
| 77F | Gait disturbance | Rt cerebellopontine angle | 1 | | MZL | STR + Focal RT | 2m |
| Age (year)/gender | Symptoms at onset | Lesion location | Number of lesions | Systemic Pathology | Treatment | Follow up |
|------------------|------------------|-----------------|-------------------|-------------------|-----------|----------|
| Jazy et al., 1980 | 59M three episodes of bizarre seizure activity, consisting of visual and auditory hallucination | Rt temporal convexity | 1 | DLBL | Resection + WBRT + Focal RT | 16m |
| Scott, et al., 1990 | 21F Seizure with behavioral change and headache. hydrocephalus, and infarction of both dentate nuclei. | NA | NA | CSF+ | Biopsy + IT chemotherapy + WBRT | 15m |
| Kumar et al., 1997 | 40F Focal numbness, visual field defects | Rt cavernous sinus | 1 | NA | MZL | RT | 5.3y |
| | 62F seizures | Biparietal dura | NA | NA | MZL | Chemotherapy | 2m |
| | 52F Seizures, focal numbness | Lt frontal dura | 1 | NA | MZL | RT + Chemotherapy + IT chemotherapy | 7m |
| | 43F Dizziness, headache, blurred vision, focal numbness | Lt tentorial | 1 | NA | MZL | RT | 9m |
| | 57F seizures | Lt anterior falk cerebri | 1 | NA | MZL | RT | 14m |
| Kambham, et al., 1998 | 39F Hearing loss, pain, weakness | Lt cerebellar pontine angle | 1 | NA | MZL | STR | 4y |
| | 62F Recent onset headache | Lt parieto-occipital area | 1 | NA | MZL | RT | 6m |
| Hodgson, et al., 1999 | 57F Headache | Rt sphenoid wing | 1 | FCL | Resection + WBRT + Focal RT | 6m |
| Altundag, et al., 2000 | 66F Syncope, seizure | Rt parietal | 1 | NA | MZL | Resection + RT | 12m |
| Amaker, et al., 2000 | 49F Intermittent headache, vomiting, decreased memory, apathy, and right-sided weakness | Lt frontal | 1 | — | T cell rich LBCL | Resection + chemotherapy | 14m |
| Sanjeevi, et al., 2001 | 46F Chronic headache, decreasing visual acuity | Lt cavernous sinus | 1 | — | MZL | STR + RT | 15m |
| Estevez, et al., 2002 | 70F bilateral temporal headache, decrease in Rt hearing and visual acuity | Paragigittal convexity | — | — | MZL | RT | 1y |
| Goetz, et al., 2002 | 64F Lt hemiparesis | Rt frontoparietal | 1 | — | MZL | STR + RT | 3m |
| Lehman, et al., 2002 | 63F Focal seizure, Rt trigeminal neuralgia | Falcotentorial | 1 | — | MZL | STR + RT | NA |
| Beriwal, et al., 2003 | 67F Neck pain | Lt cerebellar hemisphere | 1 | — | FCL | Resection + RT | 18m |
| Itoh et al., 2001 | 28F Tinnitus, nausea, headache, bilateral papilledema | CP angel | 1 | — | MZL | GTR | 2y |
| Age (year)/gender | Symptoms at onset                                      | Lesion location                      | Number of lesions | Systemic Pathology | Treatment                                      | Follow up |
|-------------------|-------------------------------------------------------|--------------------------------------|-------------------|--------------------|------------------------------------------------|-----------|
| 47M               | Seizure, visual field defects, memory loss            | Lt tentorial                         | 1                 | MZL                | STR + RT                                       | 8m        |
| 33M               | growing lump in the right frontal area                | Rt frontal                           | 1                 | —                  | Resection + RT + chemotherapy                   | 30m       |
| 53M               | persistent headaches and a generalized tonic-clonic seizures | atrium of the right lateral ventricle | 1                 | —                  | Resection + IT chemotherapy                     | 14m       |
| 64F               | Headache, Lt facial weakness                         | Rt temporoparietal                   | 1                 | CSF+               | STR + IT chemotherapy + chemotherapy            | 6.9y      |
| 33F               | Simple partial seizures and blurry vision            | Lt temporal and Lt frontal           | 2                 | CSF+               | Biopsy + WBRT + IT chemotherapy                | 7.5y      |
| 35M               | Headache, dizziness, focal paresthesia, and numbness  | Lt tentorium, Lt frontoparietal      | 2                 | CSF+               | Biopsy + WBRT + IT chemotherapy                | 4.9y      |
| 47M               | Tonic-clonic seizure                                  | Lt tentorium                         | 1                 | MZL                | STR + focal RT                                 | 2.5y      |
| 39F               | Visual loss, focal paresthesia                        |Lt frontal, Rt sphenoidal             | 2                 | —                  | STR + WBRT + Focal RT                           | 1.6y      |
| 49F               | Seizures and focal sensory symptoms                   | Rt parietal                          | 1                 | CSF+               | STR + WBRT + Focal RT                           | 0.9y      |
| 51F               | headache                                             | Bilateral frontal                    | 2                 | —                  | Biopsy + WBRT                                  | 1.1y      |
| 50F               | Headache, seizures, visual loss                       | Rt frontal                           | 1                 | MZL                | WBRT                                           | 0.7y      |
| 56F               | NM                                                   | falx                                 | 1                 | MZL                | Resection                                      | NA        |
| 49M               | Seizures                                             | Lt frontal                           | 1                 | MZL                | Resection + chemotherapy + RT                  | 7.6y      |
| 66M               | Seizures                                             | Rt frontal                           | 1                 | MZL                | Resection + RT                                 | 13m       |
| 29F               | NM                                                   | subdural                             | 1                 | MZL                | Resection + RT                                 | 3y        |
| 61F               | Headache, drowsiness, NV                             | Rt frontotemporal                    | 1                 | MZL                | Resection + NM                                 | 21m       |
| 62F               | Ataxia                                               | Lt occipital                         | 1                 | MZL                | Resection + RT                                 | 25m       |
| 47F               | Facial droop, numbness, dysarthria                    | Parietal                             | 1                 | MZL                | Resection + NM                                 | NA        |
| 57F               | Rt arm pain                                           | Lt frontoparietal                    | 1                 | MZL                | Resection + chemotherapy                       | 5.5y      |
| 70F               | Visual deficit                                       | Tentorium                            | 1                 | MZL                | Resection + RT                                 | 3.3y      |
| 59F               | Unsteady gait, visual deficit                         | Falx                                 | 1                 | MZL                | Resection + RT                                 | 2.8y      |
| 53F               | Headache, visual deficit                             | Sella, Suprasella                    | 1                 | MZL                | Resection + RT                                 | 11m       |
| 48F               | Headache, ear pain                                   | Tentorium, falx                      | 1                 | MZL                | Resection + Chemotherapy + RT                  | 20m       |
| 52F               | right hemifacial paresthesia                          | Rt parieto occipital                 | 1                 | DLBCL              | Biopsy + chemotherapy                          | 22m       |
| Age (year)/gender | Symptoms at onset | Lesion location | Number of lesions | Systemic | Pathology | Treatment | Follow up |
|-------------------|-------------------|----------------|-------------------|----------|-----------|-----------|----------|
| Yamada et al., 2006 | 59F severe frontal headaches | Bilateral frontal | 1 | — | DLBCL | Resection + chemotherapy | 30m |
| Galarza et al., 2006 | 61M generalized headache | vertex | 1 | — | DLBCL | Resection + chemotherapy + WBRT | 23m |
| Sacho et al., 2010 | 46F sudden collapse, reduced level of consciousness, and focal seizures | Rt parietal subdural | 1 | — | DLBCL | Resection + chemotherapy | Dead |
| Said et al., 2011 | 42F Rt hemiparesia, hemiparesthesia, retroorbital pain & headache | Lt vertex | 1 | — | DLBCL | Resection + chemotherapy | 34m |
| Parekh, et al., 1993 | 65F Scalp lump, Rt hemiparesia | Lt parietal | 1 | — | NHL | Resection + RT | 6y |
| Landys, et al., 1995 | 62M Headaches, malaise, unsteady gait | Frontoparietal | 1 | — | NHL | Resection + chemotherapy | 5y |
| Paige & Bernstein, 1995 | 51M Scalp mass, Headaches | Bilateral occipital | 1 | — | LBCL | Resection + RT + chemotherapy | NA |
| 71M enlarging painless mass | Lt temporal | 1 | — | LBCL | Resection + RT + chemotherapy | NA |
| Curty, et al., 1997 | 19M Scalp lump, Headaches | Rt parietal | 1 | CSF + bone | BCL | Resection + chemotherapy | NA |
| Pardhanani, et al., 2000 | 77M Ocular proptosis | Lt orbitofrontal | 1 | — | LBCL | Biopsy + RT | died |
| Age (year)/gender | Symptoms at onset | Lesion location | Number of lesions | Systemic Pathology | Treatment | Follow up |
|-------------------|-------------------|----------------|-------------------|--------------------|-----------|-----------|
| Karschnia et al., 2020 | Headaches [10 of 20 patients (50%)], cranial nerve deficits [affecting cranial nerve II, III, IV, or VI in most cases; 7 of 20 (35%)], limb weakness | Bilateral frontal | 1 | Bone+ DLBCL | Biopsy + chemotherapy | 0.2y |
| 64F | | Lt frontal | 1 | — | MZL | GKR | 3.7y |
| 60F | | Rt frontal | 1 | Bone+ DLBCL | GTR + chemotherapy | 5.2y |
| 61F | | Rt tentorium & Rt hemisphere | 1 | CSF+ MZL | Biopsy + steroid | 3.8y |
| 73F | | Sella turcica | 1 | — | MZL | Biopsy + Chemotherapy | 2.1y |
| 48F | | Lt frontal | 1 | Bone+ DLBCL | Biopsy + Chemotherapy | 0.1y |
| 68M | | Rt wall of cavernous sinus | 1 | CSF+ | T cell NHL | Biopsy + chemotherapy | 0.5y |
| 54F | | Lt frontal | 1 | Bone+ | FCL | GTR + chemotherapy | 9.3 |
| 71F | | Lt frontal, Lt parietal | 2 | Bone+ MZL | STR | 0.1y |
| 46F | | Rt frontal | 1 | Bone+ CSF+ | MZL | GTR + chemotherapy | 15.7y |
| 38F | | Lt frontal | 1 | Bone+ DLBCL | Biopsy + chemotherapy | 1.8y |
| 73F | | Rt wall of cavernous sinus | 1 | Bone+ CSF+ | DLBCL | chemotherapy | 1.6y |
| 75F | | Rt frontotemporal | 1 | Bone+ | DLBCL | Biopsy + chemotherapy | 0.5y |
| 85M | | Clivus | 1 | Bone+ DLBCL | Biopsy + chemotherapy | 4.7y |
| 76M | | Rt sphenoid wing | 1 | Bone+ | DLBCL | GTR + chemotherapy | 4.6y |
| 55M | | Lt frontal | 1 | Bone+ DLBCL | GTR + chemotherapy | 6.3y |
| 62M | | Lt middle cranial fossa | 1 | Bone+ CSF+ | FCL | STR + chemotherapy | 15.4y |
| 57M | | Lt Meckel’s cave | 1 | NA | Unspecified BCL | Resection + chemotherapy + RT | NA |
| Adbel Aziz & van Loveren, 1999 | Facial numbness & pain | Rts. Meckel’s cave | 1 | — | Lympho-blastic BCL | Resection + chemotherapy | 5m |
| Saraceni et al., 2016 | Headache & lack of coordination | Rt parietal | 1 | — | Lympho-blastic BCL | Resection + chemotherapy | 5m |
| Raguz et al., 2018 | Intermittent headache | Rt frontal | 1 | BM+ CSF- | DLBCL | Resection + chemotherapy | 4m |
| Kulkarni et al., 2012 | Painless progressive blurred vision | Rt optic canal extension to Rt cavernous | 1 | — | Low-grade BCL | Resection + RT | NA |
| Dobran et al., 2020 | Personality & mood change | Rt frontal | 1 | — | DLBCL | Resection + RT | 3y |
| 64F | | Rt handed & lat hemianopsia | 1 | — | DLBCL | Resection | 8y |
| 26F | | Lt arm weakness | 1 | — | BCL | Resection | 2y |
| Fattahi et al., 2022 | Acute loss of consciousness (LOC) | Bilateral frontal parasagittal | 1 | — | BCL | Resection + RT | 1y |

BM — bone marrow; CSF — cerebrospinal fluid; DLBCL — diffuse large B-cell lymphoma; F — female; FCL — follicular cell lymphoma; GTR — gross total resection; IT — intra-thecal; Lt — left; M — male; m — month; MZL — marginal zone lymphoma; NA — not available; NHL — non-Hodgkin’s lymphoma; RT — radiotherapy; Rt — right; STR — subtotal resection; WBRT — whole-brain radiotherapy; y — year
Table 2. Summary of table one information

| Age          | Range 19–85y | Mean 53.85y |
|--------------|--------------|-------------|
| Sex          | Male 30 (29.7%) | Female 71 (70.3%) |
| Site of lesion | Convexity 62 (61%) | Cavernous 8 (7.9%) |
|              | Tentorial 8 (7.9%) | Falcine 7 (6.9%) |
|              | Sellar 3 (2.9%) | Cerebellopontine angle 3 (2.9%) |
|              | Sphenoid wing 2 (1.9%) | Other 8 (7.9%) |
| Number of lesions | Single 91 (90%) | Multiple 10 (10%) |
| Pathology    | Marginal zone lymphoma 63 (62.3%) | Diffuse large B-cell lymphoma 24 (23.7%) |
|              | B-cell lymphoma (not determined subgroup) 7 (6.9%) | Follicular cell lymphoma 4 (3.9%) |
|              | Lymphoblastic cell lymphoma 1 (0.9%) | T cell rich B-cell lymphoma 1 (0.9%) |
|              | T cell lymphoma 1 (0.9%) | |
| Clinical signs and symptoms | Headache 34 (33.6%) | Seizure 28 (27.7%) |
|              | Focal 23 | Tonic-colonic 5 |
|              | Cranial nerve deficit 19 (18.8%) | Visual deficit 17 (16.8%) |
|              | Focal neurological deficits 12 (11.8%) | |
|              | Just motor 5 | | Just sensory 4 |
|              | Sensory + motor 3 | | |
| Neuropsychiatric problems 10 (9.9%) | |
| Treatment approach | Surgical resection (S) alone 12 (11.8%) | Radiotherapy (R) alone 6 (5.9%) |
|              | Chemotherapy (C) alone 2 (1.9%) | Biopsy (B) alone 1 (0.9%) |
|              | S + R 32 (31.6%) | S + C 17 (16.8%) |
|              | B + C 10 (9.9%) | B + R 7 (6.9%) |
|              | R + C 1 (0.9%) | S + C + R 10 (9.9%) |
|              | B + R + C 2 (1.9%) | Corticosteroid alone 1 (0.9%) |
Figure 1. Preoperative axial brain CT scan (A, B) revealed an extra-axial bi-frontal parasagittal mass with peritumoral edema with iso-signal on T1 and T2 sequence (C, D) of MRI. Also, the lesion had bright enhancement (E–G) on MRI with gadolinium enhancement of the adjacent dura mater known as dural tail. On MRV, we can see the superior sagittal sinus completely occluded (H) in the location of the tumor.
The most frequent complaints among the patients with PDL were headaches and seizures. Other accompanying symptoms of PDL originated from convexity. Frontal, parietal, temporal, and occipital were relevant neurological deficits according to their site of origin, including focal sensory, motor deficit, visual disturbance, and ataxia, respectively. Case reports with PDL originating from the skull base, including the sella and parasellar region (3 cases), cavernous sinus (8 cases), cerebellopontine angle (3 cases), and Meckel’s cave (1 case), were rare and their symptoms were determined by involved surrounding cranial nerves. The most pathologic subtype of skull base PDLs was peculiarly diffuse large B-cell lymphoma (DLBL) [1, 9, 15, 17, 21, 22]. There is just 1 case report of PDL, which originates from the atrium of the lateral ventricle; its clinical presentation was headache and generalized seizure, and its subtype was MZL [16].

In many earlier reports, PDLs were non-tender, not pulsatile, subcutaneous mass with a permeation pattern of growth. However, this feature (being a non-tender, non-pulsatile subcutaneous mass) is still an accompanying symptom. There were some case reports with less common clinical presentations; 2 cases primarily present ocular symptoms. One of them presented with progressive proptosis, which originates from the orbitofrontal lobe, and the other patient had a unilateral painless progressive blurred vision — his lesion had originated from the optic canal and extended toward the cavernous sinus [1, 23, 24]. There were also case reports of PDL with psychiatric manifestations such as bizarre behavior, hallucination, and unsteady mood [2, 25].

PDL is often initially interpreted as meningioma due to their clinical and radiographic features having a lot in common. Both appear as an iso-hypo intensity signal mass on T1 weighted magnetic resonance (MR) images with homogenous enhancement and hyperintensity signal mass on T2 weighted MR images. Dural tail sign, calvarial hyperostosis, or infiltration, en plaque thickening of the sphenoid bone, bone erosion or destruction, and invasion of the superior sagittal sinus are their common features [11, 26–28].

Figure 2. Postoperative brain CT scan (A) of the patient after 1 day revealed diffuse bi-frontal edema (B) which elevates the fixed bone flap (C). On one year follow-up, MRI with gadolinium (D, E) revealed no remnant or recurrence of the tumor.
Figure 3. Histopathologic exam of the case. Diffuse meningeal infiltration by cellular sheets of lymphocytes, plasma cells, and lymphoplasmacytoid cells with a vague focal nodular growth pattern (A, H&E, ×100). A higher-power view (B) showed mainly small lymphocytes and plasma cells. Also, many small and intermediate-sized lymphoid cells are positive for CD20 (C. Immunohistochemistry, anti-CD20 antibody, ×200). We can see the predominant population of cells with lymphoplasmacytoid and plasma cell morphologies showing positive immunoreactivity for CD138 (D. Immunohistochemistry, anti-CD138 antibody, ×200). Image E shows many of the cells in the previous figure showing cytoplasmic immunoreactivity for Kappa light chain (E. Immunohistochemistry, anti-kappa antibody, ×200). Fewer than 1% of the cells are immunoreactive for lambda light chain (F. Immunohistochemistry, anti-lambda antibody, ×200). Image G shows immunostaining for CD3 highlights scattered reactive T cells (G. Immunohistochemistry, anti-CD3 antibody, ×200). Also, we can see the low proliferation capacity of infiltrating lymphoid cells (H. Immunohistochemistry, MIB-1 antibody, ×200).
Few studies demonstrated in some detail images that were more favorable for PDL, such as calcification, the ratio of vasogenic edema to the tumor, the intensity of dural tail enhancement compared to lesion enhancement, fuzzy tumor-brain interface, and pattern of diffusion restriction. PDL in the apparent diffusion coefficient (ADC) map has a lower signal intensity than meningioma; however, the low signal intensity on the ADC map might also be observed in atypical and malignant meningioma. None of those mentioned above details was sufficient to verify a definite diagnosis [10, 21, 29].

There were 2 case reports in which PDLs were primarily misdiagnosed with acute subdural hematoma (SDH). In one of these reports, according to the preoperative image, a subdural hematoma was presumed, but during the operation, the suspicious diagnosis was substituted by meningioma. This patient had massive cerebral edema surrounding the lesion. The craniotomy and evacuation of the lesion were done. After 5 weeks, the patient returned with recurrent mass. Due to massive cerebral edema, the surgeons did not replace the bone flap, and despite all of their efforts to reduce intracranial pressure, the patient eventually died. Ultimately the histologic assessment revealed DLBCL [20]. The other case report with acute SDH had a mild presentation with no devastating manifestations; the pathology subtype in that patient was MZL [12].

Some case reports of MZL were misdiagnosed as pseudolymphomatous lesions, like plasma cell granuloma, pseudolymphoma, inflammatory pseudotumor, etc. Differentiation of lymphoma from inflammatory processes may be only possible after examining cytologic properties in the frozen section. However, Itoh and his colleagues demonstrated that the cluster of differentiate 20 (CD20) staining pattern is essential to confirm MZL diagnosis, and immunohistochemical assessment only is insufficient [22].

Neurological staging of PDL is essential to choosing a better treatment option, so all patients with presumed PDL should undergo an evaluation to exclude systemic involvement beyond the CNS. These evaluations include a lumbar puncture (seeking lymphoma cells in CSF), computed tomography of the chest, abdomen and pelvis, as well as a bone marrow biopsy. There were 16 case reports in which CSF was involved. The most popular pathology subtype in these studies was DLBCL [1, 11].

Therefore, when encountering a young patient with a short duration of neurological symptoms, rapid progression in symptoms, or systemic symptoms, caution should be taken in choosing conservative management with suspicion of meningioma diagnosis due to overlooking PDL, in which cases resection, in addition to adjuvant therapy, is necessary [10].

Many patients with PDL were reported to have chronic or acute neurologic symptoms before diagnosis [30]. In our case, the presentation of the tumor was entirely acute with LOC, and the patient had no symptoms before the day of admission. Generally, based on the location of the PDL, we could expect some previous neurological symptoms, but the presence or lack of these symptoms usually could not guide us to a preoperative diagnosis of PDL. Because of acute loss of consciousness and a cut-off point presentation in SSS on MRV, initially we suspected this extensive bi-frontal vasogenic edema is due to cortical vein thrombosis in the territory of the closed SSS, so she was treated by intravenous anticoagulant and hyper-hydration.

**Treatment**

According to previous reports, various therapeutic strategies were applied for PDL. The favorable clinical course of PDL is comparable with PCNSL, which had a better prognosis and less aggressiveness [31]. In PDL, compared to other PCNSL, the role of systemic chemotherapy in relapsing is unknown, so if leptomeningeal is involved, IT chemotherapy or WBRT should be added to surgery as adjuvant therapy because most of the time, due to infiltrative nature and relapse, gross total resection (GTR) is impossible [1, 2, 32].

The most popular treatment option for low-grade MZL with a single site of origin was resection combined with adjuvant therapy, chemotherapy, or radiotherapy. The dura mater is outside the blood brain barrier (BBB), so chemotherapy seems an excellent option to reach it without passing BBB11. DLBL had a poor prognosis in which 5-year survival after chemotherapy and radiotherapy was less than 10%. This poor prognosis may be due to a high proliferation fraction that causes rapid growth and recurrence. Treatment was unsatisfying with high doses of MTX either alone or with radiotherapy [20].

Like in our case, PDL represented a large frontal parasagittal mass in one study with occlusion of the anterior third of the superior sagittal sinus and severe cerebral edema. In that study, thallium accumulation in scintigraphy in both early and late phases determined tumor aggressiveness, and the bicornal craniotomy was done with bone replacement and systemic chemotherapy, used due to their mentioned-above benefits connected with eradication of the remnant (not passing BBB) [18].

Although most therapeutic strategies for MZL were effective in achieving complete remission during their clinical follow-up, in a study by de la Fuente and his colleagues, 4 patients had progression, 2 of whom had local recurrent tumors (at the resection site, just one of them received suboptimal focal radiotherapy) [21]. In another
study by Iwamoto and his colleagues, in 3 of 8 patients, relapse occurred after 6 years. However, they did not confirm that relapse occurred from the exact clone as the PDL; however, histopathologically they were similar. One patient in their study developed treatment-induced leukoencephalopathy after high doses of MTX and WBRT, which seems to suggest that so chemotherapy may be unnecessary [8, 11].

Berivial and his colleagues described a well-defined ovoid mass overlying the left cerebellopontine angle with follicular subtype. They treated their patient with radiotherapy after resection, which is the standard treatment for early-stage of follicular lymphoma in other sites [33].

One study reported a PDL with T cell rich B-cell lymphoma. This subtype of PDL appeared iso-intense in all MRI sequences, and angiography revealed prominent neovascularization so that the patient underwent obliteration of neovascular blush, and the following day embolization craniotomy was done [27].

They are limited reports of complications following surgery, such as hematoma, hygroma, infection, etc. There was just one study that reported wound infection at the site of craniotomy that responded well to antibiotic therapy without bone flap removal.

Considering the treatment approach to our patient, the patient was comatose with anisocoria, and we had to operate on her, on postoperative day 1, for decompressive craniectomy despite gross total resection of the large mass and expansible duralpasty with pedunculated patch and complete intraventricular treatments for relieving brain edema. As a result, we think that in patients with extensive brain edema, suspected preoperative PDL, or an unusual meningioma diagnosis, it is better to not place the bone flap at the time of surgical resection of the tumor, as it was done in former studies. Also, based on preoperative imaging, if histopathology shows it could be PDL, we could treat the patient with other options: biopsy followed by radiotherapy, etc. We recommend designating a study to reach the goal of PDL management without unwanted surgery-related complications.

Conflict of interest

All authors declare no conflicts of interest.

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