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REVIEW ARTICLE

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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

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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.

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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 sinuous (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-coronal 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 meningothelial 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].

Table 1. Results of primary dural lymphoma literature review

	Age (year)/ /gender	Symptoms at onset	Lesion location	Number of lesions	Systemic	Pathology	Treatment	Follow up
de la Fuente et al., 2017	47M	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
	66M	Seizures, progressive gait disorder	Left fronto-parietal	1		MZL	Biopsy + chemotherapy	1.1y
	41F	Headache, focal seizures, Rt visual field cut	Lt parieto-occipital	1		MZL	GTR + Focal RT	7.2y
	51F	long h/o headache with new worsening	Lt frontal	1		MZL	STR + WBRT + Focal RT	11.3y
	49F	Focal Lt face numbness and paresthesia Lt ear tinnitus and otalgia	Lt tentorium (compressing L brainstem and cerebellar hemisphere)	1		MZL	STR + Focal RT	6.8y
	69F	Walking difficulty	Lt temporo-parietal convexity	1		MZL	Biopsy + chemotherapy	8.6y
	51F	Seizure, Lt homonymous hemianopsia	Rt occipital	1		MZL	Biopsy + WBRT + Focal RT	5.7 y
	59M	Headache	Rt temporal, R frontal	2		MZL	GTR of both lesions + NA	NA
	49F	Focal seizures	Rt fronto-parietal	1		MZL	STR + WBRT + Focal RT	4.6y
	72F	Generalized tonic-clonic seizure	Long lesion extending along the falx cerebri	1		MZL	STR + NA	NA
	33F	Generalized tonic-clonic seizure	Lt frontal and Lt parietal	2		MZL	STR + WBRT + Focal RT	0.9y
		Headache, L facial weakness	Rt temporo-parietal	1		MZL	Partial resection + WBRT + Chemotherapy	17.2y
	39F	Vision loss, focal R-sided paresthesia	Lt frontal, Rt sphenoid region/orbital apex	2		MZL	STR of sphenoid lesion only + WBRT + Focal RT	9m
	49F	Headache, seizures, visual loss	Rt temporal, Rt frontal	2		MZL	Biopsy + WBRT	3.1y
	39F	Headache	Rt frontal	1		MZL	STR + Focal RT	1.3y
	30F	Facial pain	Cavernous sinus	1		MZL	STR + Focal RT	9y
	67F	Headache	Rt occipital	1		MZL	STR + NA	NA
	47M	Seizures	Lt frontal	1		MZL	Biopsy + Focal RT	5.6y
	34M	Seizures	Rt temporal	1		MZL	GTR + Focal RT	5.3y
	67M	Seizures	Rt frontal	1		MZL	Biopsy + focal RT	4.7y
	51F	Focal paresthesia and numbness	Lt cavernous sinus	1		MZL	Biopsy + focal RT + Chemotherapy	3y
	57M	Seizures, headache	Rt temporal	1		MZL	GTR + Focal RT	2.5y
	59F	Headache	Suprasellar region	1		MZL	Biopsy + Focal RT	1.9y
	48F	Cranial nerve palsy	Cavernous sinus (bilateral)	M		MZL	Biopsy + Chemotherapy	10m
	77F	Gait disturbance	Rt cerebellopontine angle	1		MZL	GTR + Focal RT	8m
	50 F	Cranial nerve palsy	Cavernous sinus	1		MZL	STR + Focal RT	2m

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Table 1. cont. Results of primary dural lymphoma literature review

	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	CSF+	NA	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	22m
	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 falx 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	Parasagittal 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

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Table 1. cont. Results of primary dural lymphoma literature review

	Age (year)/ /gender	Symptoms at onset	Lesion location	Number of lesions	Systemic	Pathology	Treatment	Follow up
Rottneck et al., 2004	47M	Seizure, visual field defects, memory loss	Lt tentorial	1	—	MZL	STR + RT	8m
Abdullah, et al., 2005	33M	growing lump in the right frontal area	Rt frontal	1	—	Lympho-blastic BCL	Resection + RT + chemo-therapy	30m
Kelley, et al., 2005	53M	persistent headaches and a generalized tonic-clonic seizure	atrium of the right lateral ventricle	1	—	MZL	Resection + IT chemo-therapy	14m
Iwamoto, et al., 2006	64F	Headache, Lt facial weakness	Rt temporoparietal	1	CSF+	MZL	STR + IT chemothera-py + chemotherapy	6.9y
	33F	Simple partial seizures and blurry vision	Lt temporal and Lt frontal	2	CSF+	MZL	Biopsy + WBRT + IT chemotherapy	7.5y
	35M	Headache, dizziness, focal paresthesia, and numbness	Lt tentorium, Lt frontoparietal	2	CSF+	MZL	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	—	MZL	STR + WBRT+ Focal RT	1.6y
	49F	Seizures and focal sensory symptoms	Rt parietal	1	CSF+	MZL	STR + WBRT+ Focal RT	0.9y
	51F	headache	Bilateral frontal	2	—	MZL	Biopsy + WBRT	1.1y
	50F	Headache, seizures, visual loss	Rt frontal	1	—	MZL	WBRT	0.7y
Tu, et al., 2005	56F	NM	falx	1	—	MZL	Resection	NA
	49M	Seizures	Lt frontal	1	—	MZL	Resection + chemothera-py + RT	7.6y
	66M	Seizures	Rt frontal	1	—	MZL	Resection + RT	13m
	29F	NM	subdural	1	—	MZL	Resection + RT	3y
	61F	Headache, drowsiness, N/V	Rt frontotemporal	1	—	MZL	Resection + NM	21m
	62F	Ataxia	Lt occipital	1	—	MZL	Resection + RT	25m
	47M	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 + Chemothera-py + RT	20m
↓ Brito et al., 2014	52F	right hemifacial paresthesia	Rt parieto occipital	1	—	DLBCL	Biopsy + chemotherapy	22m

Table 1 cd. Results of primary dural lymphoma literature review

	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, hemiparesthsia, retro orbital 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

Table 1. cont. Results of primary dural lymphoma literature review

	Age (year)/ /gender	Symptoms at onset	Lesion location	Number of lesions	Systemic	Pathology	Treatment	Follow up
Karschnia et al., 2020	64F	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 or dysesthesias [7 of 20 (35%)], gait instability, or vertigo [5 of 20 (25%)], word-finding difficulties or confusion [5 of 20 (25%)], and painless growth of a subcutaneous mass	Bilateral frontal	1	Bone+	DLBCL	Biopsy + chemotherapy	0.2y
	60F		Lt frontal	1	—	MZL	GKRS	3.7y
	61F		Rt frontal	1	Bone+	DLBCL	GTR + chemotherapy	5.2y
	73F		Rt tentorium & Rt hemisphere	1	CSF+	MZL	Biopsy + steroid	3.8y
	48F		Sella turcica	1	—	MZL	Biopsy + Chemotherapy	2.1y
	68M		Lt frontal	1	Bone+	DLBCL	Biopsy + Chemotherapy	0.1y
	54F		Rt wall of cavernous sinus	1	CSF+	T cell NHL	Biopsy + chemotherapy	0.5
	71F		Lt frontal	1	Bone+	FCL	GTR + chemotherapy	9.3
	46F		Lt frontal, Lt parietal	2	Bone+	MZL	STR	0.1y
	38F		Rt frontal	1	Bone+ CSF+	MZL	GTR + chemotherapy	15.7y
	73F		Lt frontal	1	Bone+	DLBCL	Biopsy + chemotherapy	1.8y
	75F		Rt wall of cavernous sinus		Bone+ CSF+	DLBCL	chemotherapy	1.6y
	85M		Rt frontotemporal	1	Bone+	DLBCL	Biopsy + chemotherapy	0.5y
	76M		Clivus	1	Bone+	DLBCL	Biopsy + chemotherapy	4.7y
	55M		Rt sphenoid wing	1	Bone+	DLBCL	GTR + chemotherapy	4.6y
	62M		Lt frontal	1	Bone+	DLBCL	GTR + chemotherapy	6.3y
	57M		Lt middle cranial fossa	1	Bone+ CSF+	FCL	STR + chemotherapy	15.4y
Adbel Aziz & van Loveren, 1999	40F	Facial numbness & pain	Lt Meckel's cave	1	NA	Unspecified BCL	Resection + chemotherapy + RT	NA
Saraceni et al., 2016	42M	Headache & lack of coordination	Rt parietal	1	—	Lymphoblastic BCL	Resection + chemotherapy + RT	5m
Raguz et al., 2018	34M	Intermittent headache	Rt frontal	1	BM+ CSF–	DLBCL	Resection + chemotherapy	4m
Kulkarni et al., 2012	39F	Painless progressive blurred vision	Rt optic canal extension to Rt cavernous	1	—	Low-grade BCL	Resection + RT	NA
Dobran et al., 2020	49M	Personality &mood change	Rt frontal	1	—	DLBCL	Resection +RT	3y
	64F	Rt handed & lat hemianopsia	Lt occipitoparietal	1	—	DLBCL	Resection	8y
	26F	Lt arm weakness	Rt frontoparietal	1	—	BCL	Resection	2y
Fattahi et al., 2022	56F	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%)

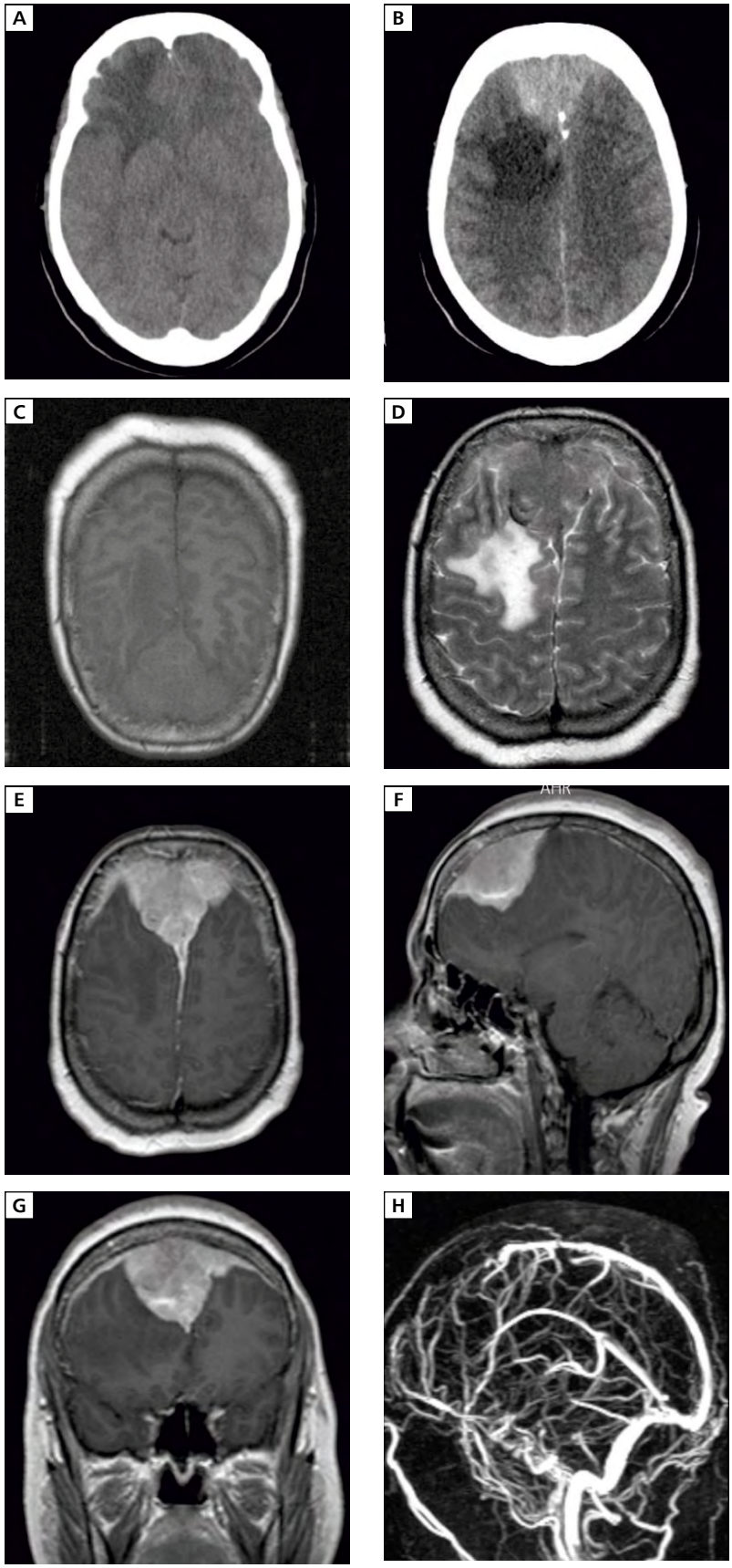

Figure 1. Preoperative axial brain CT scan (**A, B**) revealed an extra-axial bi-frontal parasagittal mass with peritumoral edema with iso-signal on T1 and T2 sequence (**C, D**) of MRI. Also, the lesion had bright enhancement (**E–G**) on MRI with gadolinium enhancement of the adjacent dura mater known as dural tail. On MRV, we can see the superior sagittal sinus completely occluded (**H**) in the location of the tumor

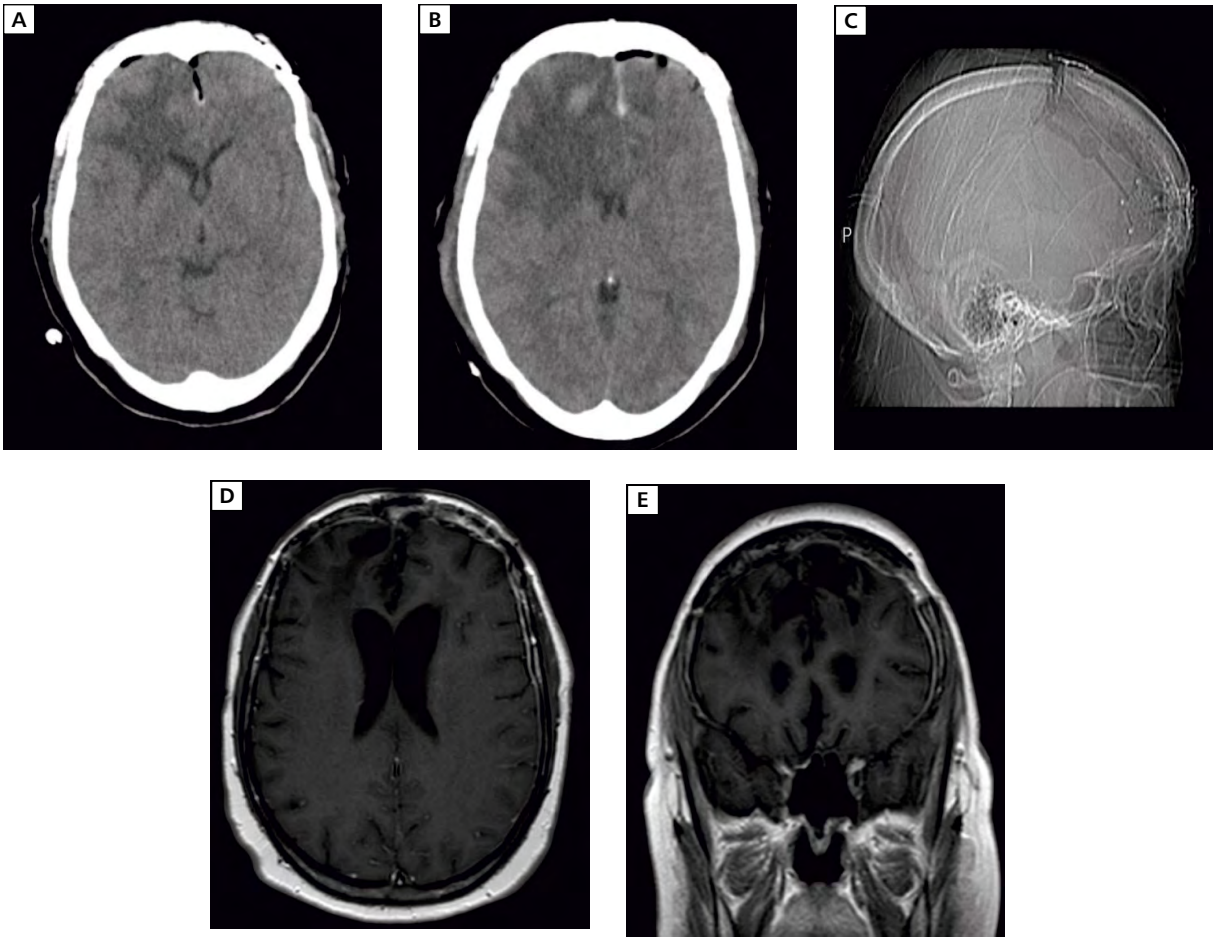

Figure 2. Postoperative brain CT scan (**A**) of the patient after 1 day revealed diffuse bi-frontal edema (**B**) which elevates the fixed bone flap (**C**). On one year follow-up, MRI with gadolinium (**D, E**) revealed no remnant or recurrence of the tumor

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].

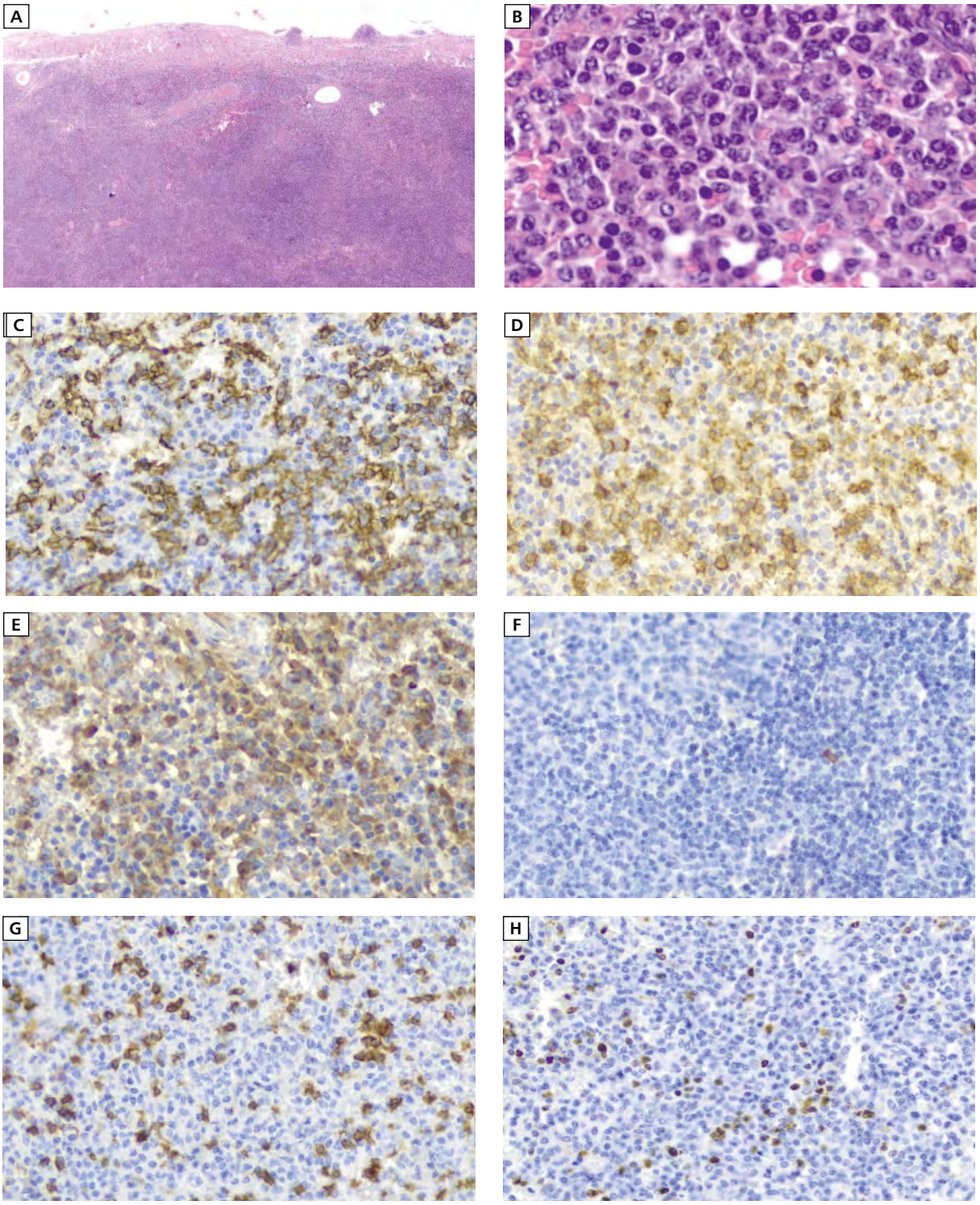

Figure 3. Histopathologic exam of the case. Diffuse meningeal infiltration by cellular sheets of lymphocytes, plasma cells, and lymphoplasmacytoid cells with a vague focal nodular growth pattern (**A**, H&E, ×100). A higher-power view (**B**) showed mainly small lymphocytes and plasma cells. Also, many 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 bicoronal craniotomy was done with bone replacement and systemic chemotherapy, used due to their mentioned-above benefits connected with eradiation 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].

Beriwal 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 expansile duraplasty with pedunculated patch and complete intravenous 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 replace 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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