



Intraventricular Lymphoplasmacyte-Rich Meningioma: A Case Report

İntraventriküler Lenfoplazmositten Zengin Menenjiyom: Bir Olgu Sunumu

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ABSTRACT

Lymphoplasmacyte-rich meningioma (LPM) is one of the rarest variants of meningioma and those LPMs that arise in the intraventricular space are even rarer. LPMs are classified as grade I (benign) tumors with a low proliferative rate and diagnosis is made through the histological identification of high numbers of inflammatory cells (lymphocytes and plasma cells) in the resected tumor tissue. In the current case, magnetic resonance imaging of a 37-year-old woman who presented at our neurosurgery department following a generalized tonic-clonic seizure revealed a partially mortified intraventricular mass, which had caused pronounced peritumoral edema and had a relatively rough surface. Surgical resection was performed. Histological analysis revealed large numbers of inflammatory cells, confirming the diagnosis of LPM, but also indicated that the lesion was positive for the proliferation marker Ki-67. Follow-up magnetic resonance imaging 3 months after surgery revealed no residual tumor or recurrence.

KEYWORDS: Lymphoplasmacyte-rich Meningioma, Lymphocyte, Plasmacyte, Intraventricular, Ki-67

ÖZ

Lenfoplazmositten zengin menenjiyom (LPM), en nadir menenjiyom çeşitlerinden biridir ve intraventriküler boşluktan köken alan LPM'ler daha da nadirdir. LPM'ler düşük proliferatif hızı sahip derece I (benign) tümörler olarak sınıflandırılır ve tanı rezeksiyon yapılmış tümör dokusunda yüksek sayıda enflamatuvar hücrenin (lenfositler ve plazma hücreleri) histolojik olarak tanımlanması yoluyla konur. Mevcut olguda genel bir tonik-klonik nöbetten sonra beyin ve sinir cerrahisi bölümümüze gelen 37 yaşında bir kadında manyetik rezonans görüntüleme, peritümöral ödeme neden olmuş ve nispeten kaba bir yüzeye sahip kısmen gangrene bir intraventriküler kitle gösterdi. Cerrahi rezeksiyon yapıldı. Histolojik analiz LPM tanısını doğrulayacak şekilde yüksek sayıda enflamatuvar hücre gösterdi ama aynı zamanda lezyonun proliferasyon işareti Ki-67 pozitifliğini de ortaya koydu. Cerrahiden 3 ay sonra kontrol manyetik rezonans görüntüleme herhangi bir rezidüel tümör veya nöks göstermedi.

ANAHTAR SÖZCÜKLER: Lenfoplazmositten zengin menenjiyom, Lenfosit, Plazmosit, İnaventriküler Ki-67

INTRODUCTION

Lymphoplasmacyte-rich meningioma (LPM) is a rare type of meningioma that is characterized histopathologically by massive infiltrates of inflammatory cells, such as lymphocytes and plasma cells. Despite its World Health Organization (WHO) classification as grade I (benign), the invasion of adjacent brain tissue is frequently seen by magnetic resonance imaging (MRI) (13). Nonetheless, the outcome of surgical resection without adjuvant therapy is generally favorable (25).

The symptoms of LPM vary according to the location, but some systemic hematological abnormalities, including hyperglobulinemia and iron refractory anemia, have been documented (7). Associated amyloid material and increased serum immunoglobulins have been described, though rarely (9).

Since it was first described in 1971 by Banerjee and Blackwood (2), 36 cases of LPM have been reported. None

have been intraventricular in origin. Indeed, intraventricular meningiomas are extremely rare compared to those of other locations, accounting for only 0.5–3% of all meningiomas (22). Most intraventricular tumors arise in the lateral ventricle and over 90% occur in the trigone (3,17). Here we report an LPM that originated in the intraventricular trigone.

CASE REPORT

A 37-year-old woman was admitted to our neurosurgery department 5 hours after suffering a generalized tonic-clonic seizure. She had no prior history of seizure activity and a neurological examination showed no abnormalities. MRI revealed an intraventricular mass that had caused marked peritumoral edema. The mass measured approximately 3 cm in diameter. Compared with gray matter, the lesion was slightly hypointense on T1-weighted MRI and hyperintense on T2-weighted MRI. It was strongly enhanced after Gd-diethylenetriamine pentaacetic acid administration (Figure 1A–C). The lesion

appeared to have a relatively rough surface (Figure 1E) and was partially mortified (Figure 1A).

The patient underwent a right occipitotemporal craniotomy for symptomatic improvement. A grayish-pink mass originating from the intraventricular trigone and measuring $3 \times 3 \times 4$ cm was completely removed. The neoplasm closely adhered to the lateral ventricular walls and choroid plexus but had a complete tumor capsule. Superficially, the resected tumor mass was gritty. Hematoxylin and eosin (H&E) staining revealed multiple nests of meningotheial cells and abundant collagen fibers with a dense infiltration of lymphocytes and plasma cells (Figure 2A). Immunohistochemical staining (Figure 2B–J) revealed that the lesion was positive for epithelial membrane antigen (EMA), vimentin, Ki-67, kappa, lambda, CD3, CD20, and CD138, but negative for neuron-specific enolase (NSE), glial fibrillary acidic protein (GFAP), and progesterone receptor (PR).

Following surgery, the patient had intermittent fever with a maximal temperature of 38.5°C . Red blood cells ($1,000 \times$

$10^6/\text{L}$) and white blood cells ($53 \times 10^6/\text{L}$) were found in the cerebrospinal fluid (CSF) by routine CSF analysis. Blood and CSF cultures were sterile. The intermittent fever resolved with symptomatic treatment 4 days after surgery. The rest of the postoperative course was uneventful and the outcome was favorable. The patient has returned to normal activities without sequelae or epilepsy. Follow-up MRI revealed no residual tumor or recurrence 3 months after surgery (Figure 1D–F).

DISCUSSION

An Introduction to LPM

LPM was histologically classified as a rare variant of meningioma by the WHO in 1993 (12). It features extensive chronic inflammatory infiltrates that often overshadow the inconspicuous meningotheial component in the pathology findings. The categorization of LPM as a distinct clinicopathologic entity remains controversial, since its behavior often resembles that of an inflammatory process (5,16). Indeed, some re-

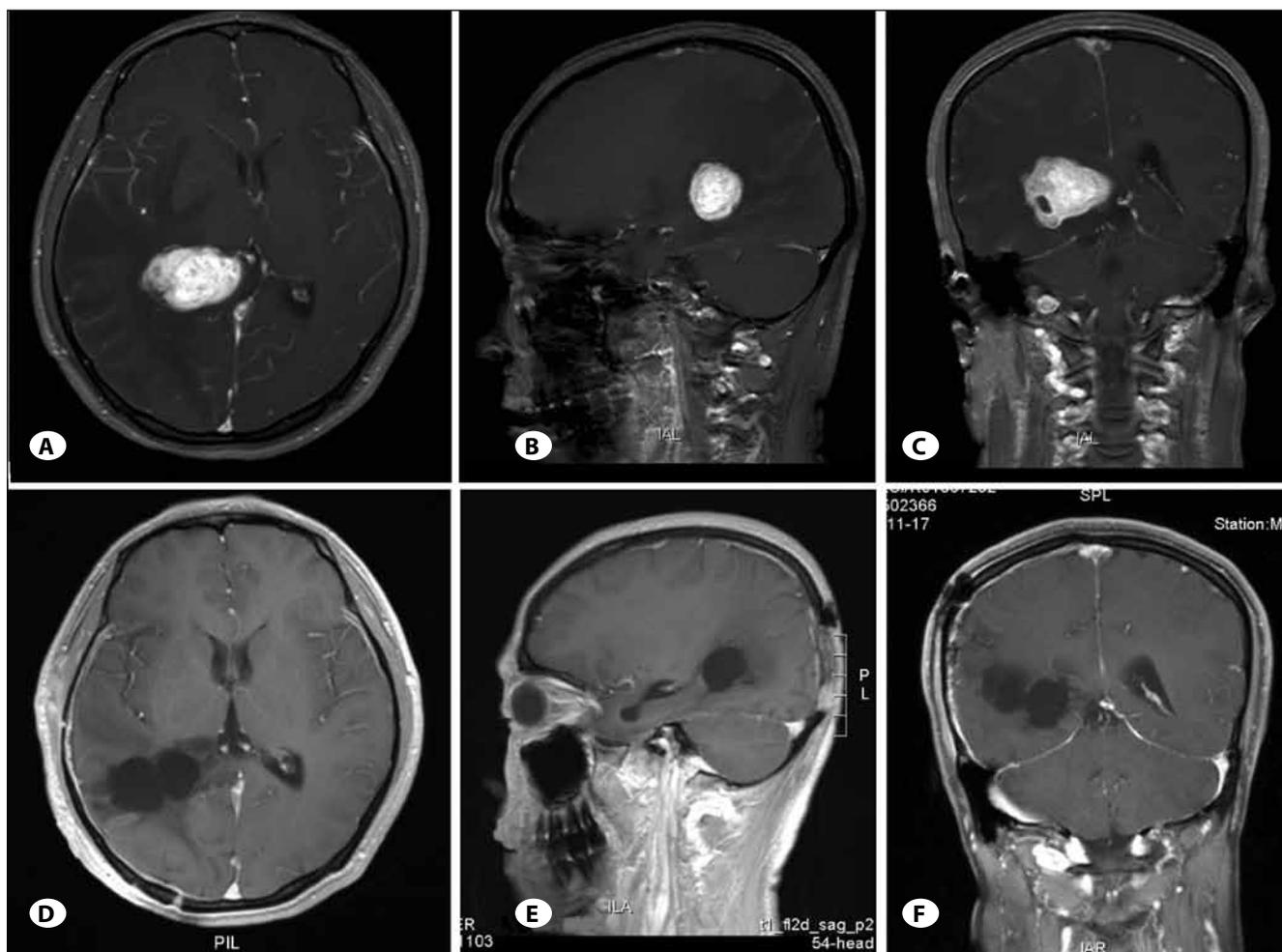


Figure 1: Preoperative and postoperative MRI results. Preoperative MRI of the brain with Gd-diethylenetriamine pentaacetic acid contrast agent revealed an enhanced irregular lesion (A, axial view; B, sagittal view; C, coronal view). MRI scanning at 3 months after the operation showed that the tumor was completely resected; no recurrence was observed (D, axial view; E, sagittal view; F, coronal view).

searchers have suggested that the nature of LPM is that of a primary meningioma with prolonged immunological stimulation by plasma cells or a meningothehal reaction secondary to chronic inflammation (25).

Clinical Findings

LPM belongs to WHO grade I meningiomas, which means that the risk of recurrence and aggressive growth is low. In the present case, however, immunohistochemical staining revealed that the tumor cells were positive for the proliferation

marker Ki-67. The preoperative MRI revealed that the tumor had a relatively rough surface and was partially mortified. This is not the first report of a LPM showing positive staining for Ki-67: Nohara et al. (21) reported a 12-year-old boy who presented with a LPM in the posterior fossa. Histological examination showed that the tumor had invaded the normal brain tissue and that the Ki-67 labeling index, determined by using a MIB-1 monoclonal antibody, was relatively high. The authors concluded that LPM can exhibit an atypical invasive nature (21).

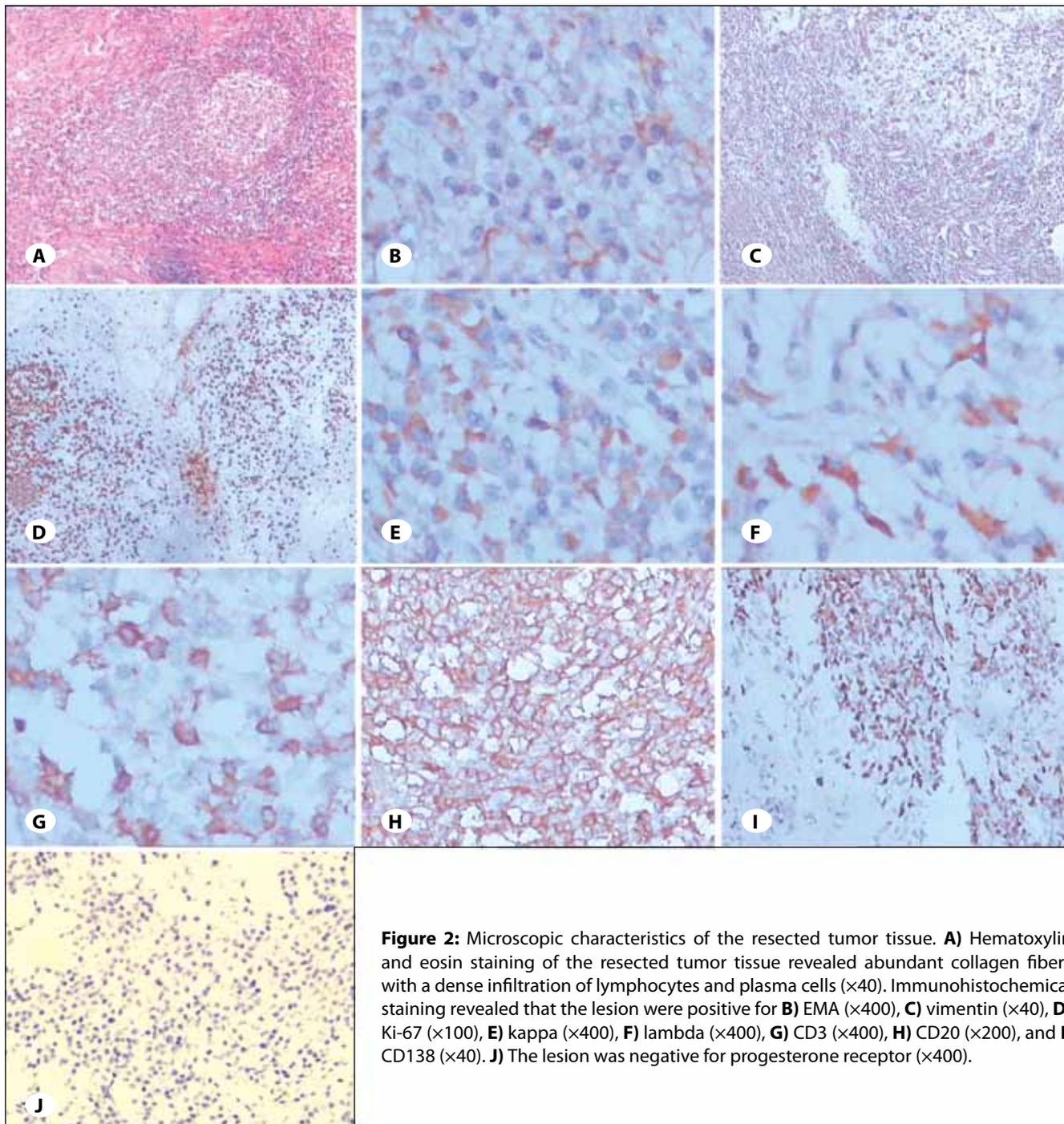


Figure 2: Microscopic characteristics of the resected tumor tissue. **A)** Hematoxylin and eosin staining of the resected tumor tissue revealed abundant collagen fibers with a dense infiltration of lymphocytes and plasma cells (×40). Immunohistochemical staining revealed that the lesion were positive for **B)** EMA (×400), **C)** vimentin (×40), **D)** Ki-67 (×100), **E)** kappa (×400), **F)** lambda (×400), **G)** CD3 (×400), **H)** CD20 (×200), and **I)** CD138 (×40). **J)** The lesion was negative for progesterone receptor (×400).

Literature Review

In a review of the literature conducted via PubMed with the keywords "lymphocyte", "plasmacyte", and "meningioma", 23 papers (Table I), including the present case, reported LPMs (10, 13, 20, 25), with 37 cases in total. With 21 female and 16 male patients, the female:male ratio was 1.3:1. The age of onset ranged from 4 to 72 with a mean of 35.6 years. Multiple lesions are more frequent in LPM than in typical meningioma with 16.2% of these 37 cases involving multiple lesions, a rate that is only 1–3% for typical meningiomas (5). The most frequent locations of LPM are the convexity, and the cerebellopontine angle (CPA).

With regard to tumor appearance, analysis of the MRI results and the pathology of the LPM suggests that its appearance is different from ordinary meningioma (13). LPM tumor features include flat growth along the meninges, an irregular shape, an unclear boundary, prominent edema, a notable sensitivity to contrast enhancement, invasion of adjacent brain tissues, and a characteristic inflammation.

Surgical resection was the treatment of choice in the 37 cases and few showed tumor progression after resection. There is no standard adjuvant therapy for residual or recurrent tumor after surgical resection. Gamma knife radiosurgery with a low

Table I: Review of the Literature via Pubmed with the Keywords "Lymphocyte", "Plasmacyte" and "Meningioma"

Author	Year	Age	Gender	Location	Treatment	Outcome
Banerjee et al. (2)	1971	71	M	Subfrontal	Resection	Well
Horten et al. (9)	1979	15	F	CPA	Resection, radiation, chemotherapy	Dead
	1979	53	F	Foramen magnum	Resection	Well
	1979	52	F	Falx	Resection	Well
	1979	22	M	Parasagittal	Resection	Well
	1979	4	M	CPA	Resection	Well
Stam et al. (24)	1980	59	M	Falcotentorial	Resection	Not described
Mirra et al. (18)	1983	11	F	Multiple	Resection	Well
	1983	39	F	Spinal canal	Resection	Expansion
Gi et al. (7)	1990	8	M	Multiple	Resection, radiation	Recurrence
Loiseau et al. (15)	1995	11	F	Multiple	Resection	Well
Yamaki et al. (26)	1997	22	M	Multiple	Resection	Well
	1997	24	F	Clival	Resection	Recurrence
Mizushima et al. (19)	1997	64	F	Parasagittal	Resection	Well
Katayama et al. (11)	1997	47	F	Convexity	Resection	Well
Yoneyama et al. (27)	1999	36	M	Convexity	Resection	Recurrence
	1999	41	F	Convexity	Resection	Well
	1999	22	M	Falx	Resection	Well
Pittella et al. (23)	2001	47	M	Spinal canal	Resection	Dead
Bruno et al. (5)	2004	45	F	Convexity	Resection	Well
Loh et al. (14)	2006	22	F	Sphenoid ridge	Resection	Well
Hirunwiwatkul et al. (8)	2007	24	M	Multiple	Resection	Well
Nohara et al. (21)	2007	12	M	CPA	Resection	Well
Ghosal et al. (6)	2007	25	M	Convexity	Resection	Well
Avninder et al. (1)	2008	22	F	Foramen magnum	Resection	Well
Bodi et al. (4)	2008	72	F	Convexity	Resection	Well
Liu et al. (13)	2010	38 (mean)	M4 F3	Not clear	Not described	Not described
Kanno et al. (10)	2011	55	F	Jugular foramen	Resection	Well
Wang et al. (25)	2011	62	F	Multiple	Resection, radiation	Well
Nakayama et al. (20)	2012	37	F	Convexity	Resection	Well
Present case	2013	37	F	Intraventricular	Resection	Well

CPA: cerebellopontine angle, F: female, M: male.

peripheral dose may have an effective role in the treatment of LPM (25).

In the present case, meningioma was suspected due to the clinical presentation and MRI examination. The rough edge and pronounced peritumoral edema were indicative of an aggressive meningioma, a diagnosis that was supported by the close adherence of the tumor to the lateral ventricular walls and choroid plexus and its intact tumor capsule. The H&E pathology results, however, indicated that it was an inflammatory pseudotumor. A review of the literature suggested LPM, which was confirmed by positive immunohistochemical staining for EMA and vimentin. Although the patient developed a fever after surgery, blood and CSF analysis did not indicate a bacterial infection. Two possible causes of fever are a febrile response after surgery or blood left in the subarachnoid space after the craniotomy, but the relationship between the inflammation caused by LPM and the fever is still unclear. There was no recurrence of the tumor after 3 months of follow-up.

In conclusion, we have reported a case of intraventricular LPM in a 37-year-old woman who presented with occasional seizures. The definitive diagnosis of LPM relied on histopathological findings. LPM is very rare, particularly when located in the intraventricular space. We believe that total excision remains the treatment option of choice for such patients.

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