Intraventricular Lymphoplasmacyte-Rich Meningioma: A Case Report

İntraventriküler Lensoplazmositten Zengin Menenjيوم: 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, Plasmaeyte, Intraventricular, Ki-67

ÖZ


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 × 3 × 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 meningothelial 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 × 10⁶/L) and white blood cells (53 × 10⁶/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 meningothelial 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-
searchers have suggested that the nature of LPM is that of a primary meningioma with prolonged immunological stimulation by plasma cells or a meningothelial 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](image)

**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).
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   |
| Horten et al. (9)     | 1979 | 53  | F      | Foramen magnum        | Resection                          | Well   |
| Horten et al. (9)     | 1979 | 52  | F      | Falx                  | Resection                          | Well   |
| Horten et al. (9)     | 1979 | 22  | M      | Paraventricular        | Resection                          | Well   |
| Horten et al. (9)     | 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   |
| 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   |
| Yamaki et al. (26)    | 1997 | 24  | F      | Clival                | Resection                          | Recurrence |
| Muzushima et al. (19) | 1997 | 64  | F      | Paraventricular        | Resection                          | Well   |
| Katayama et al. (11)  | 1997 | 47  | F      | Convexity             | Resection                          | Well   |
| Yoneyama et al. (27)  | 1999 | 36  | M      | Convexity             | Resection                          | Recurrence |
| Yoneyama et al. (27)  | 1999 | 41  | F      | Convexity             | 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   |
| Nakai et al. (10)     | 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   |

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 this region. There was no recurrence of the tumor after 3 months of follow-up.

In our experience, 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.

REFERENCES