Clinical, Radiologic, and Pathologic Features of 56 Cases of Intracranial Lymphoplasmacyte-Rich Meningioma

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■ OBJECTIVE: **Intracranial lymphoplasmacyte-rich meningioma (LPRM) is rarely reported because of its extremely low incidence, and current understanding of this disease is poor. We analyzed the incidence and clinical, radiologic, pathologic, and prognostic features of intracranial LPRMs.**

■ METHODS: **Approximately 10,908 intracranial meningiomas were surgically resected in Beijing Tiantan Hospital between June 2009 and November 2016. All pathologically proven LPRM cases were identified. Statistical analysis was performed to determine which parameters were associated with prognoses and tumor resection.**

■ RESULTS: **LPRMs accounted for approximately 0.51% of intracranial meningiomas (56 of 10,908). Patients with LPRM had a mean age of 44.6 years and there was no significant gender preference (28 male and 28 female). Gross total resection was achieved in 45 cases, subtotal resection in 9 cases, and partial resection in 2 cases. At discharge, Karnofsky Performance Scale scores and neurologic function improved in 42 cases (75.0%) and worsened in 14 cases (25.0%). At the 41.5 months median follow-up (range, 5–97 months), 5 patients had tumor recurrence, and the 3-year and 5-year progression-free survival rates were 94.6% and 92.9%, respectively. One patient died of tumor recurrence. At the latest follow-up, Karnofsky Performance Scale scores improved in 48 cases (85.7%), worsened in 7 cases (12.5%), and 1 patient died (1.8%). Univariate analysis showed that the extent of tumor resection and tumors located in the skull base were significantly associated with short-term outcomes. Skull base location was significantly associated with increased risk of worse long-term outcomes. Tumor size ≥45 mm, poorly defined tumor border, and skull base location significantly limited the extent of tumor resection.**

■ CONCLUSIONS: **Intracranial LPRM is a rare subtype of meningioma with no gender difference and low recurrence. Long-term survival is expected, although tumors located in the skull base and bone destruction are independent risk factors of poor long-term outcomes. Microsurgical treatment of skull base LPRM remains a formidable challenge because of poorly defined borders and critical neurovascular structure encasement. Radical tumor resection, which induces severe neurologic deficits, is unnecessary.**

INTRODUCTION

Meningiomas represent the most common primary intracranial tumors, accounting for 26%–35% of all primary brain tumors, with annual incidence rates ranging from 1.3 to 7.62 in 100,000 individuals.[1-5] They originate

Key words
- Clinical features
- Intracranial
- Lymphoplasmacyte-rich meningioma
- Radiology
- Surgical outcomes

Abbreviations and Acronyms
CI: Confidential interval
CT: Computed tomography
EMA: Epithelial membrane antigen
GTR: Gross total resection
IHP: Idiopathic hypertrophic pachymeningitis
KPS: Karnofsky Performance Scale
LPRM: Lymphoplasmacyte-rich meningioma
MRI: Magnetic resonance imaging
OR: Odds ratio
OS: Overall survival
PCG: Plasma cell granuloma
PFS: Progression-free survival
WHO: World Health Organization

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from arachnoid cap cells and/or arachnoid trabeculae and are classified into 3 grades and 16 subtypes according to the 2016 World Health Organization (WHO) classification for brain tumors, among which 80%–90% are benign and the remaining 10%–20% are atypical and malignant meningiomas.[6,7]

Lymphoplasmacyte-rich meningioma (LPRM), which was first described by Banerjee and Blackwood in 1971, has been classified as grade I by WHO since 1993.[8,9] LPRM is a rare, benign variant of meningioma, characterized by massive inflammatory cell infiltration and a variable proportion of meningothelial tumorous elements. Fewer than 70 LPRMs have been reported in the English literature, most of which were isolated case reports except for a few studies.[10,11] Given the paucity of reported cases, the clinical, radiologic, and pathologic features of LPRMs remain unclear. In addition, factors associated with prognosis after surgical resection need to be further studied.

To further elucidate the clinical, radiologic, and pathologic features and prognosis of this rare disease, we present an analysis of a consecutive cohort of 56 patients with histologically proven intracranial LPRM from one of the largest neurosurgical centers in China.

METHODS

Patient Population

Reviewing the records of Beijing Tiantan Hospital for 2009–2016 identified 56 patients with pathologically confirmed LPRM among a total of 10,908 patients with meningioma.

Clinical and Radiologic Records

The clinical data and operation records of LPRM cases were retrospectively reviewed. The following information was recorded: patient age and sex, symptoms at disease onset, duration of disease, preoperative diagnosis, tumor size, tumor location, computed tomography (CT) and magnetic resonance imaging (MRI) features, extent of tumor removal, and surgical outcomes. Tumor size was assessed according to measurements of the maximum tumor diameter on MRI. The extent of resection was recorded as Simpson grade 1–4 according to the operation record and postoperative MRI. Peritumoral edema was correlated with tumor size according to the modified score by Regelsberger et al.,[12] which classifies edema as follows: grade 0, no edema; grade I, equal to or smaller than the tumor size; grade II, surpassing the tumor size; and grade III, nearly hemispheric edema. Radiographic evaluation was performed by 2 board-certified neurosurgeons.

Pathologic Examination

Fresh paraffin-embedded tumor tissue was cut into 5-μm slices and stained with hematoxylin-eosin. Immunohistochemical staining was performed for differential diagnoses. Two independent neuropathologists reviewed the microscopic pathologies of the LPRMs according to the WHO grading system. Immunohistochemical analyses included epithelial membrane antigen (EMA), vimentin, S-100, leukocyte common antigen, CD38, CD3, CD20, glial fibrillary acidic protein, and Ki-67.

Surgical Outcome and Postoperative Follow-Up

Follow-up data for all patients with LPRM were obtained by office visits or telephone interviews. Postoperative complications, progression-free survival (PFS) and overall survival (OS) were recorded. Short-term outcome was assessed by Karnofsky Performance Scale (KPS) scores at discharge. Long-term outcome was assessed by KPS scores at the latest follow-up. The progression of LPRM was defined according to radiologic studies performed after tumor removal. PFS was defined as the time between initial surgery and tumor progression on radiologic studies. OS was defined as the time between initial surgery and death.

Statistical Analysis

The continuous variables and categorical data were presented as the means and percentages, respectively. The χ^2 test was used to assess potential risk factors for short-term and long-term surgical outcomes and extent of tumor resection. Multivariate logistic regression analyses were used to identify independent risk factors of surgical outcomes and extent of tumor resection. Statistical analysis was performed using SPSS version 19.0 software (IBM Corp., Armonk, New York, USA). Probability values were obtained from 2-sided tests, with statistical significance defined as $P < 0.05$.

RESULTS

Incidence and Clinical Features of Intracranial LPRMs

The incidence of intracranial LPRM among intracranial meningioma was 0.51% (56 of 10,908 cases). Clinical data are summarized in Table 1. The ages of the 56 patients (28 males and 28 females) at symptom onset ranged from 15 to 66 years, and the mean age was 44.6 ± 12.0 years (mean ± standard deviation). The duration from symptom onset to hospital admission ranged from 2 weeks to 6 years; the median duration was 2 months. The initial manifestations included headache or increased intracranial hypertension (24 patients), limb weakness (8 patients), dizziness (7 patients), blurred vision (7 patients), numbness (6 patients), seizures (6 patients), memory loss (3 patients), diplopia (3 patients), facial paralysis (3 patients), hearing loss (2 patients), and hyposmia (2 patients). Seven patients were asymptomatic; their tumors were found incidentally on neuroimaging.

Of the 56 LPRMs, 54 were primary lesions and only 2 were recurrent lesions. Six of the 56 LPRM cases (10.7%) were misdiagnosed before surgery (Figure 1) as schwannoma, myeloid sarcoma, medulloblastoma, teratoma, craniopharyngioma, and lymphoma (1 case each, 1.8%). Six patients received postoperative radiotherapy. The median preoperative KPS score was 90 (range, 70–100).

Radiologic Features of Intracranial LPRMs

Radiologic data are summarized in Table 2. Non–skull base tumors were located in the frontal convexity (19 cases), parietal convexity (10 cases), lateral ventricle (3 cases), pineal region (2 cases), posterior fossa convexity (1 case), and fourth ventricle (1 case). Skull base LPRMs were located in the sphenoid ridge (6 cases), tuberculum sellae/parasellar (5 cases), cerebellopontine angle (3 cases), petroclival region (3 cases),