Dural Marginal Zone Lymphoma Confused with Meningioma en Plaque

We report a case of dural marginal zone lymphoma which showed the usual radiological findings resembling meningioma. A 59-year-old woman presented with headache. Initial computed tomography and magnetic resonance images showed a frontal convexity meningioma. The patient underwent a craniotomy and subtotal (Simpson grade II) resection of tumor was done. Pathological examination confirmed an extranodal marginal zone B-cell lymphoma of Mucosa-Associated Lymphoid Tissue (MALT). The lesion was composed of a lymphoid mass with irregularly shaped follicles surrounded by many monomorphic small lymphocytes and a stained marginal zone for B-cell markers CD20 and CD29a. The natural history of primary CNS lymphoma and MALT type lymphoma are different. B-cell MALT lymphoma can mimic meningioma in its radiological features. Accordingly, MALT lymphoma of the CNS must be considered in the differential diagnosis of meningioma.

KEY WORDS : B-cell lymphoma of MALT · Meningioma.

INTRODUCTION

Central nervous system (CNS) lymphoma is a non-Hodgkin’s lymphoma that involves the brain, spinal cord, or ocular structures. The role of surgery in the management of CNS lymphoma is limited. Combining chemotherapy and radiotherapy has improved the survival of patients with CNS lymphoma, but long term survival is still quite limited.

Primary CNS non-Hodgkin’s lymphoma is rare, comprising 1% of intracranial tumors. CNS lymphoma occurs typically as the result of secondary spread from systemic lymphoma4,9,10). Lymphomas involving the dura are typically secondary, occurring in 5-9% of all patients with non-Hodgkin’s lymphoma. On rare occasions, primary meningeal lymphomas have been described. Recently, these have been classified as mucosa-associated lymphoid tissue (MALT) type lymphoma or marginal zone B-cell MALT lymphoma2,3). MALT lymphoma of the cranial dura mater is rare, but can be cured with surgery. We report a rare case of MALT type lymphoma, which was diagnosed as en plaque meningioma preoperatively.

<table>
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<th>Case</th>
<th>Reference</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Presentation</th>
<th>Therapy</th>
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<td>M</td>
<td>Rt frontal convexity</td>
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<td>Radiation/Chemotherapy</td>
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<td>8</td>
<td>41</td>
<td>M</td>
<td>Left frontal convexity</td>
<td>Headache, nausea, vomiting</td>
<td>Total resection</td>
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<td>63</td>
<td>F</td>
<td>Both supratentorial and infratentorial convexity</td>
<td>Focal seizure</td>
<td>Subtotal resection</td>
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<td>36</td>
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<td>Both parietal convexity</td>
<td>Headache</td>
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<td>47</td>
<td>M</td>
<td>Lt tentorium</td>
<td>Rt visual field disturbance, seizure</td>
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CASE REPORT

A 59-year-old woman presented with headache for three days. The patient had been treated for hypertension and diabetes mellitus for over 10 years. Physical and neurological examinations were essentially normal. Results of routine laboratory data were within normal limits. A computed tomographic scan of her brain revealed an extra-axial, broad-based large mass in the high frontal convexity with no calcification or hemorrhage. Magnetic resonance imaging identified an extensive homogeneously enhanced extra-axial dura-based mass as a meningioma, which showed isosignal intensity on T1-weighted images and low signal intensity on T2-weighted images (Fig. 1). Cerebral angiography revealed a mass with vascular staining fed by both superficial temporal arteries (Fig. 2).

A bifrontal craniotomy was performed. The mass was attached to the dura. Dissection of tumor from normal parenchyma was difficult because of severe adhesion. The mass was near totally removed with excision of the affected dura and a small remnant of the dura along the sagittal sinus wall was coagulated. Intraoperative gross examination of the surgical specimen showed a thickened dura, and a yellowish solid extra-axial and highly vascularized mass. Grossly, the lesion was compatible with meningioma. The patient’s postoperative course was uneventful. Histopathologically, the tissue consisted of well-defined lymphoid follicles surrounded by monomorphic small lymphocytes with dense chromatin nuclei. Marginal zones stained for B-cell marker, suggesting a diagnosis of B-cell lymphoma: CD20/CD79a immunostain was positive, CD3/CD5 was negative and Ki67 immunocytochemistry was less than 10%. The diagnosis of marginal zone B-cell MALT lymphoma was confirmed by histological and immunophenotyping results according to the WHO classification (Fig. 4).

The patient was discharged with no postoperative neurological deficits. We recommended brain radiation therapy, but was refused. There was no evidence of recurrence at follow-up MRI (Fig. 3), and the patient remains disease-free with an unremarkable neurological examination at the 22-month follow-up.
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**DISCUSSION**

Primary CNS lymphoma represents about 1% of intracranial
tumors. CNS involvement may be predominantly paren-
chymal or leptomeningeal. These tumors may consist of
solitary, multifocal or diffuse masses. Primary CNS lymphoma
typically presents with clinical symptoms and signs of
increased intracranial pressure and/or cortical dysfunction5,9).
The clinical presentation includes personality change,
cerebellar signs, headaches, seizures, cranial nerve palsy,
and motor dysfunction. The clinical course is usually rapid.
The time between the onset of symptoms and the diagnosis
is often short.

Extranodal marginal zone B-cell MALT type lymphomas,
also known as low-grade B-cell MALT lymphomas11,12,
can occur in a wide variety of extranodal sites, in particular,
gastrointestinal tract, thyroid, salivary gland, lung, and,
less commonly, the ocular adnexa, thymus, skin, soft tissue,
breast, tongue, tonsil, gallbladder, liver, urogenital tract,
and dura6,7). Known predisposing conditions are chronic
inflammation such as Helicobacter-associated gastritis7, Borrelia
burgdorferi infection (skin), and autoimmune diseases
such as Hashimoto’s thyroiditis and Sjogren’s syndrome.
Extranodal marginal zone B-cell lymphoma has also been
reported in the brain, where it may involve the parenchyma,
leptomeninges, or dura. Nine cases of intracranial dura-

**CONCLUSION**

Primary CNS non-Hodgkin’s lymphoma is malignant
and prognosis remains poor. Primary MALT type lymphomas
of the cranial dura mater are rare, but can be cured with
local therapy and respond well to radiation and chemotherapy.
Therefore, MALT lymphoma of the CNS must be considered
in the differential diagnosis of meningioma and other extra-
axial pathology.

References

1. Abdullrah S, Morgenstern D, Rosado MF, Losos IS : Primary lympho-
blastic B-cell lymphoma of the cranial dura mater: a case report and
review of the literature. Leuk Lymphoma 46: 1651-1657, 2005
2. Bacon CM, DU MQ, Dogan A: MALT Lymphoma: a practical
ML: Non-Hodgkin’s lymphoma of mucosa-associated lymphoid
tissue. Oncologist 11: 1100-1117, 2006
5. Costa H, Franco M, Hahn MD: Primary lymphoma of the central
nervous system: a clinical-pathological and immunohistochemical
study of ten autopsy cases. Arq Neuropsiquiatr 64: 976-982, 2006
6. Fariaha P, Gascoyne RD: Molecular pathogenesis of mucosa-associated
Prax 95: 1163-1168, 2006
E703-704, 2006
10. Hunt MA, Jahnke K, Murillo TP, Neuwelt EA: Distinguishing primary
central nervous system lymphoma from other central nervous system
diseases: a neurosurgical perspective on diagnostic dilemmas and