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Case Report

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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 lympoma of MALT · Meningioma.

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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 lymphoma[4,9,10]. 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 lymphoma[2,3]. 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. Histopathological features and management were discussed including review of reported cases (Table 1)[1,8,14-17,19].

Table 1. Review of primary CNS MALT type lymphomas that mimics meningioma

Case	Reference	Age	Sex	location	Presentation	Therapy
1	1	33	M	Rt frontal convexity	growing lump in the right frontal	Radiation/ Chemotherapy
2	8	41	M	Left frontal convexity	Headache, nausea, vomiting	Total resection Radiation/ Chemotherapy
3	13	63	F	Both supratentorial and Infratentorial	Focal seizure	Subtotal resection
4	14	36	F	Both parietal convexity	Headache	Radiation/ Chemotherapy
5	15	64	F	Rt fronto-parietal convexity	Lt hemiparesis	Subtotal resection Radiation
6	16	73	F	Lt fronto-parietal convexity	Rt arm weakness, partial seizure, dysphasia	Chemotherapy
7	17	47	M	Lt tentorium	Rt visual field disturbance, seizure	Subtotal resection/Radiation

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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).

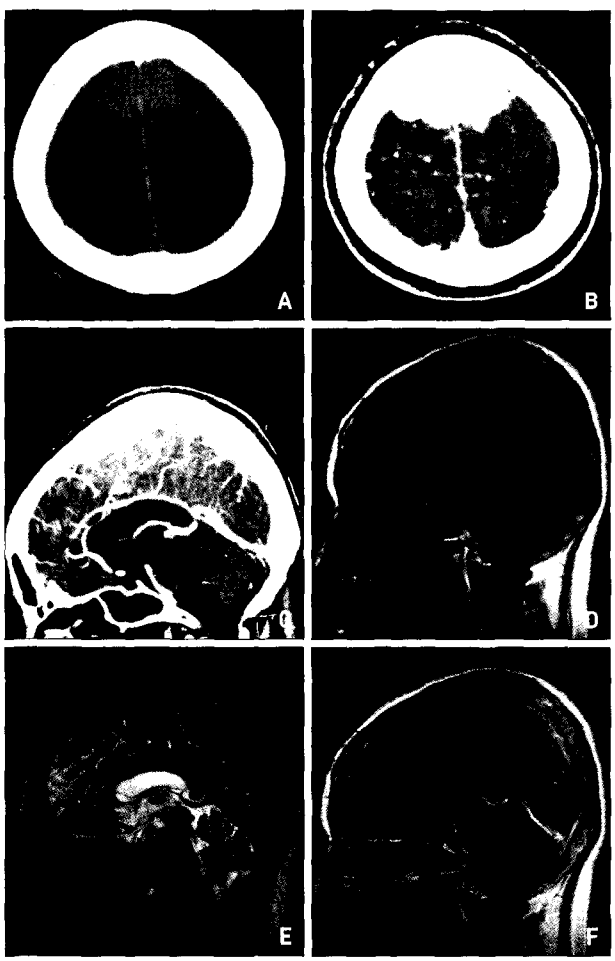

Fig. 1. Computed tomography shows a slightly high-density mass in high frontal convexity (A) and broad-based mass of dura with contrast enhancement at axial (B) and sagittal (C) image. Magnetic resonance image (MRI) scans show a mass which demonstrates iso-signal intensity in T1-weighted (D) images, low signal intensity in T2-weighted (E) magnetic resonance images. T1-weighted gadolinium-enhanced MRI shows a homogenously enhancing lesion. The mass had a broad base in the dura (F).

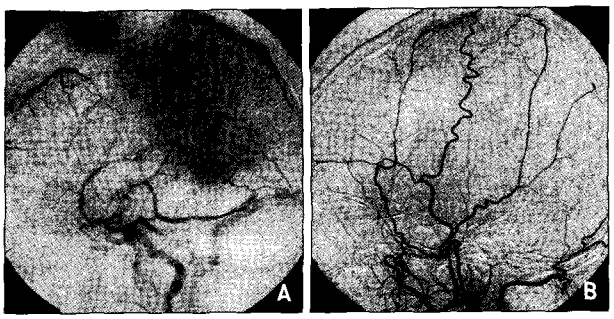

Fig. 2. Internal cerebral angiogram shows filling defect at the superior sagittal sinus by compression of the tumor (A). Tumor stains from the branches of the right superficial temporal artery (B).

Fig. 3. Follow-up magnetic resonance image (MRI) shows remaining dural thickening at the surgical bed and no enhanced mass at T1-weighted gadolinium-enhanced coronal (A) and sagittal (B) MRI.

A bifrontal craniotomy was performed. The mass was attached to the dura. Dissection of tumor from normal parenchyme 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 lympocytes 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.

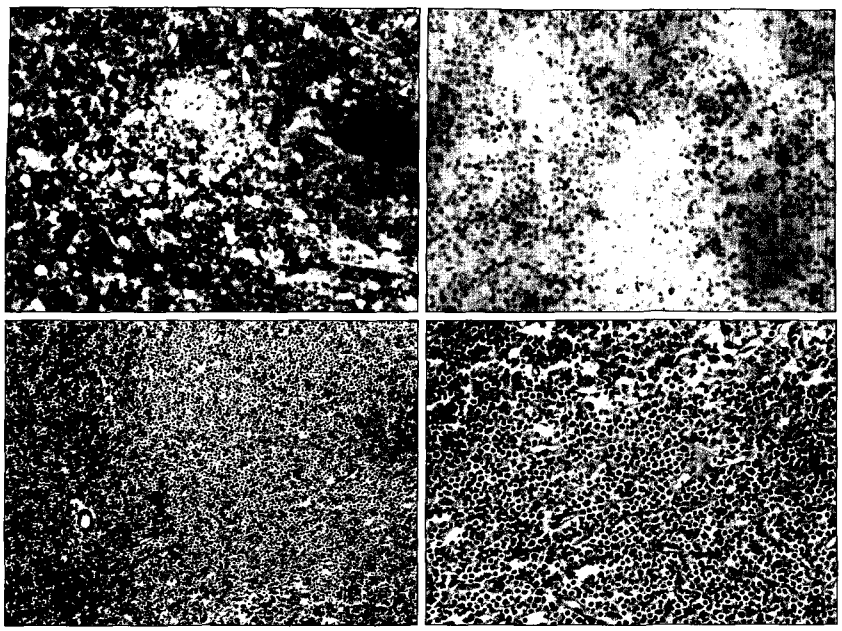

Fig. 4. Immunohistochemical staining demonstrates positive staining for CD 20 (A) and CD 79a (B). Microphotography finding of the surgical specimen, showing infiltration of neoplastic lymphoid cell, monomorphic population of small B lymphocyte with dense chromatic neucleus. H&E×200 (C), × 400 (D).

broad-based dural mass, radiographically indistinguishable from a meningioma. Dura-based MALT lymphoma of the CNS differs from the majority of primary CNS lymphomas in pathologic findings. On immunohistochemical analysis, MALT lymphoma express pan-B-lymphocyte markers (CD19, CD20 and CD79a), and surface immunoglobulin, but do not express CD3, CD5, CD10, CD45. To confirm its diagnosis, the predominance of CD20 B-cells and kappa light chain restriction is essential.

MALT lymphoma of the CNS is a slow-growing lymphoma and is curable by local excision, radiation, and systemic chemotherapy. It is therefore important to distinguish this entity from other types of B-cell non-Hodgkins lymphoma.

DISCUSSION

Primary CNS lymphoma represents about 1% of intracranial tumors. CNS involvement may be predominantly parenchymal 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 dysfunction[5,9]. 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 lymphomas[11,12], 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 dura[6,7]. Known predisposing conditions are chronic inflammation such as Helicobacter-associated gastritis[17], 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-based low-grade B-cell MALT lymphoma have been reported in the literature to date. Our case presented as a solitary

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.

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