CASE REPORT

Sphenoid Ridge Lymphoplasmacyte-rich Meningioma

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There are numerous histologic variants of meningioma. Among the more uncommon are intracranial masses composed of meningiomatous and plasma cell-lymphocytic elements. We report a 22-year-old woman with lymphoplasmacyte-rich meningioma who initially presented with dizziness and progressive headache. Neuroradiologic images revealed typical meningiomas of the sphenoid ridge with extensive perifocal edema. Complete macroscopic removal of the tumor was performed. Histologic examination revealed a meningioma with massive infiltrates of plasma cells and lymphocytes. Brain computed tomography on the 6th postoperative day revealed total removal of the tumor with marked reduction of brain edema. Complete resolution of symptoms occurred with no evidence of tumor recurrence during 2 years of follow-up. [*J Formos Med Assoc* 2006;105(7):594–598]

Key Words: brain tumor, meningioma

Lymphoplasmacyte-rich meningioma is a designation adopted by the new World Health Organization Classification of Tumors of the Central Nervous System[1] to describe a tumor characterized by the components or features of a common meningioma accompanied by massive infiltration of plasma cells and lymphocytes. Because of its atypical histologic and clinical presentation, some investigators have argued against diagnosis of this particular type of tumor as a meningioma.[2,3] Meningiomas presenting as tumors with the features of a lymphoplasmacyte-rich meningioma are very unusual.[4–8] The pathophysiology associated with this form of tumor is poorly understood. We report a young Taiwanese woman with lymphoplasmacyte-rich meningioma.

Case Report

This 22-year-old woman presented with 10 days of progressive headache, dizziness, and nausea. Computed tomography (CT) (Figure 1A) revealed a sphenoid ridge isodense tumor mass with homogeneous enhancement (Figure 1B). Laboratory findings were normal. No hematologic abnormalities were noted. Left temporoparietal craniotomy was performed, revealing a grayish-red, firm tumor attached to the sphenoidale dura. Complete macroscopic removal of the tumor was performed. Brain CT on the 6th postoperative day (Figure 1C) revealed total removal of the tumor with marked reduction of brain edema. The patient's recovery was

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characterized by complete resolution of her symptoms, and no evidence of tumor recurrence was detected at the 6-month follow-up (Figure 1D).

Microscopic examination of the surgically resected tumor disclosed inflammatory granulation tissue with lymphocyte and plasma cell infiltration (Figure 2A). Clusters of cells forming epithelioid foci and lobules of meningothelial cells, surrounded by dense lymphoplasmacytic infiltrates, were also noted (Figure 2B). The meningothelial component exhibited a strong immunoreactivity for epithelial membrane antigen (EMA) (Figure 2C). Most of the type T lymphocytic cells present in this tumor were strongly positive for CD45 (Figure 2D).

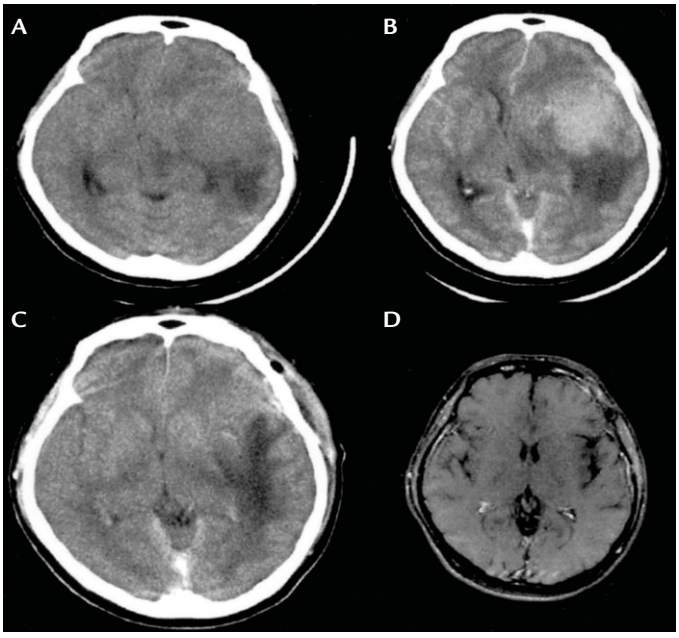

Figure 1. Neuroradiologic images of a lymphoplasmacyte-rich meningioma in a young woman. (A) Non-enhanced CT scan shows an isodense tumor mass over the left temporoparietal area with severe brain edema. (B) CT with contrast medium shows homogeneous enhancement. (C) CT performed on the 6th postoperative day reveals total removal of the tumor and marked reduction in brain edema. (D) MRI performed 6 months postoperatively shows no tumor recurrence.

Discussion

The clinical features of 18 cases of lymphoplasmacyte-rich meningioma that have been reported to date, including the case described in this report, are

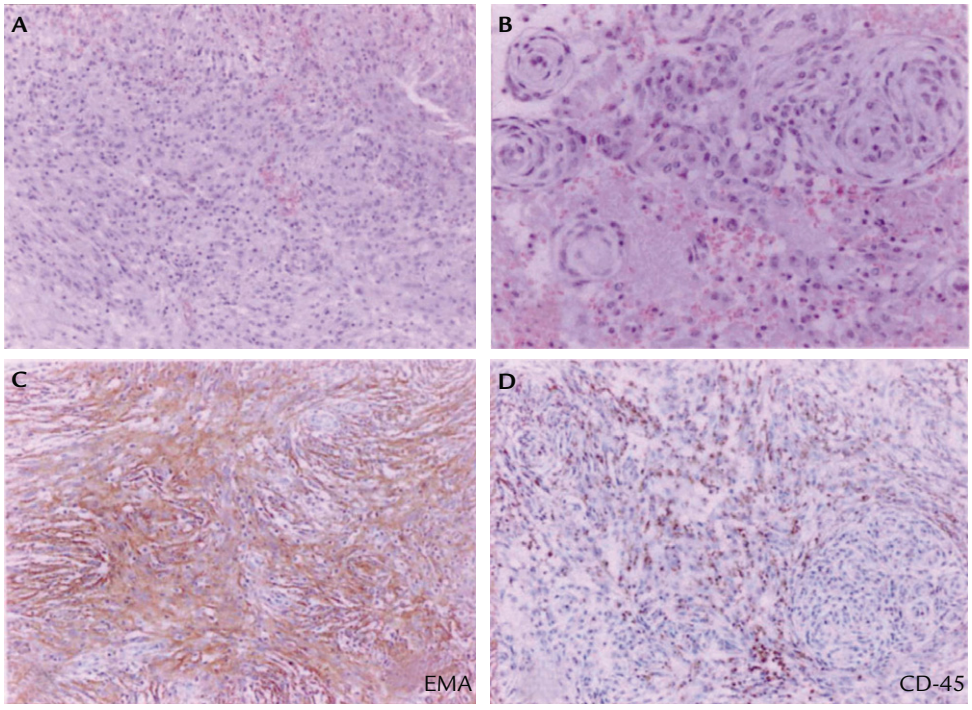

Figure 2. Microscopic appearance and immunohistochemical properties of the lymphoplasmacyte-rich regions of the meningioma. (A) Tumor specimen showing inflammatory granulation tissue with lymphocyte and plasma cell infiltration (hematoxylin & eosin, HE, original magnification ×100). (B) Stained section showing lobules of meningothelial cells, surrounded by dense lymphoplasmacytic infiltrates (HE, original magnification ×200). (C) Strong immunoreactivity of the meningothelial component for epithelial membrane antigen (immunohistochemical stain ×100). (D) Strong immunoreactivity for CD45 exhibited by most of the type T lymphocytic cells present in the tumor (immunohistochemical stain ×100).

Table. Clinical features of 18 patients with lymphoplasmacyte-rich meningioma

Authors, year	Age (yr), sex	Site	CT scan	Treatment	Outcome
Banerjee & Blackwood, 1971	71, M	Anterior skull base		Resection	8 mo, no recurrence
Horten et al, 1979	15, F	CPA		Resection Radiation Chemotherapy	2 yr, dead
	53, F	Foramen magnum		Resection	7 mo, well
	52, F	Falx		Resection	5 mo, well
	22, M	Falx		Resection	5 yr, well
	10, M	Bilatera CPA		Resection	2 mo, no change
Stam et al, 1980	59, M	Falcotentorial	Edema –	Resection	
Mirra et al, 1983	11, F	Frontal convexity	Edema –	Resection	1 yr, well
		Temporal convexity		Resection	
		CPA		Resection	
	39, F	Spinal canal (C3–7)		Resection	3 yr, enlargement
Loiseau et al, 1995	11, F	Parietal convexity	Edema +	Resection	11 mo, well
Yamaki et al, 1997	22, M	Parasellar	Edema –	Resection	7 yr, regression
		Clival		None	
		CPA		None	7 yr, regression
		Spinal canal (C1–5)		Resection	7 yr, regression with partial enlargement
	24, F	Clival	Edema –	Resection	8 yr, enlargement; 10 mo later, regression
		Clinoid		Resection	12 yr, no recurrence
		Middle fossa		None	10 mo, regression
		Jugular formen		Resection	7 mo, regression
Mizushima et al, 1997	64, F	Parasagittal and falx	Edema +	Resection	4 yr, no recurrence
Katayama et al, 1997	47, F	Parietal convexity	Edema +		
Yoneyama et al, 1998	36, M	Parietal convexity	Edema –	Resection	5 yr, no recurrence
	41, F	Frontal convexity	Edema +	Resection	5 yr, no recurrence
	22, M	Falx	Edema +	Resection	8 mo, no recurrence
Present case	22, F	Sphenoid ridge	Edema +	Resection	11 mo, no recurrence

CPA = cerebellopontine angle.

presented in the Table. Two clinical features are notable that are unique to these tumors. First, the age of onset in these patients ranges from 10 to 71 years with a mean of 34.5 years, indicating a predilection for a younger age group. Second, according to statistics compiled by the Brain Tumor Registry, female predominance among patients with this meningioma variant is less marked when compared with that among patients suffering from meningiomas of every type.[1] The male to female ratio of the former is 1:1.4 whereas that of the latter is 1:1.9. However, the number of reported cases of lymphoplasmacyte-rich meningioma is too small to identify the clinical features that can help to definitively differentiate these meningiomas from typical meningiomas. It should also be noted that many other neoplasms or intracranial masses can mimic a lymphoplasmacyte-rich meningioma with respect to exhibition of cortical mass and biologic abnormalities. Examples of such neoplasms include chordoid meningioma, multiple myeloma, solitary plasmacytoma, plasma cell granuloma, giant lymph node hyperplasia, sinus histiocytosis, and lymphomatoid granulomatosis.

Chordoid meningiomas have a peculiar chordoid pattern characterized by massive lymphoplasmacytic infiltrates in the interlobular stroma.[9] Even though direct involvement of the central nervous system, independent of skull or spine lesions, is unusual in multiple myeloma, rare cases that mimic meningiomas have been reported.[10] In our patients, the lack of bone involvement excluded the diagnosis of multiple myeloma.

Solitary plasmacytoma is usually observed in women during the 5th decade of life and histologic examination shows a predominance of plasma cells.[11,12] Histologically, plasma cell granulomas are mainly composed of heavy infiltrates of plasma cells, lymphocytes, and nests of epithelial-like cells. These epithelial-like cells can be distinguished from meningothelial cells by immunostaining for epithelial membrane antigen, which is positive in meningiomas (Figure 2C), and show negative staining in plasma cell granulomas.[13,14]

Plasma cell-type giant lymph node hyperplasia shares some of the biologic aspects of lymphoplasmacyte-rich meningioma; however, pathologic examination shows benign lymphoid tissue but never a meningothelial component. The diagnosis of a solitary brain localization of sinus histiocytosis[15,16] and a lymphomatoid granulomatosis with isolated involvement of the brain[17,18] can be easily excluded by histologic examination as well.

Various hypotheses have been advanced to explain the infiltration of lymphoplasmacytes. For example, these lesions have been proposed to develop from the collision of a plasmacytoma with a meningioma or from a plasmacytoma of the meninges with a leptomeningeal reaction.[4] The inflammatory cell reaction to the meningioma has also been proposed to constitute a mechanism of host resistance.[5] However, the relationship between inflammatory lesions and meningiomas remains uncertain. Immunohistochemical study in this case suggests that the lymphoplasmacytic infiltrate was not neoplastic in nature but rather represented an inflammatory reaction to the meningioma, suggesting that a biologic reaction was responsible for the marked brain edema. The tumor had not disturbed the venous circulation, and the brain edema disappeared after resection of the tumor.

The significance of blood abnormalities found in lymphoplasmacyte-rich meningioma is unclear. Stam et al concluded that the plasma cell infiltrates were not tumoral in origin because of the abundant production of almost all classes of immunoglobulins.[8] Blood abnormalities are thought to be dependent on the plasmacyte infiltrates in these patients because abnormalities disappear after complete removal of the tumor and reappear with relapse.[19] In our patient, no hematologic abnormalities were noted.

The prognosis for patients with lymphoplasmacyte-rich meningioma is unknown. Our patient did not receive postoperative radiotherapy and had no evidence of tumor recurrence during 24 months of follow-up. Further study of lymphoplasmacyte-rich meningiomas is required to determine the prognosis of patients with lymphoplasmacyte-rich meningioma and to clarify the significance of the meningeal reaction as a manifestation of the host–tumor relationship at the meninges.

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