CASE REPORT

Spindle cell hemangioendothelioma of the brain

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Summary A 23-year-old healthy male patient presented with an absence seizure for the first time. A contrast-enhanced brain computed tomography scan and magnetic resonance imaging revealed a tumor with perifocal edema in the superior part of the left frontal lobe. The tumor was treated with a total resection via stereotactic craniotomy. The pathologic report documented spindle cell hemangioendothelioma of the brain. Spindle cell hemangioendothelioma is a rare vascular tumor featured by a histologic manifestation intermediate between benign hemangioma and malignant angiosarcoma. It is hoped that our report will contribute to further understanding of the neuropathology and natural history of this unusual tumor.

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1. Introduction

Spindle cell hemangioendothelioma is a rare vascular tumor that is featured by a histologic manifestation intermediate between benign hemangioma and malignant angiosarcoma.1-8 Enzinger and Weiss were the first to describe hemangioendothelioma as a heterogeneous group with at least three subtypes, including epithelioid hemangioendothelioma, spindle cell hemangioendothelioma, and malignant endovascular angioendothelioma.2 Clinically, spindle cell hemangioendothelioma presents as single or multiple cutaneous and subcutaneous nodules, and usually involves the lung, heart, lymph nodes, liver, or bone. Because central nervous system cases are rare, we describe a case of intra-axial spindle cell hemangioendothelioma of the brain occurring in a 23-year-old male patient.1

2. Case history

This previously healthy 23-year-old male patient presented with an absence seizure for the first time on November 9,
2009. On examination, he was conscious and alert, and all vital signs were normal. The cranial nerves were intact. Muscle power was 5/5 in four limbs. Sensation was normal. All laboratory tests were within normal limits. A contrast-enhanced brain computed tomography (CT) scan revealed a tumor with high density in the superior part of the left frontal lobe (Fig. 1). Contrast-enhanced brain magnetic resonance imaging (MRI) revealed a mass with perifocal edema (Figs. 2 and 3). Based on the findings of a homogeneously enhanced, well-defined lesion affecting the brain, a provisional diagnosis of cavernous angioma was made.

On November 13, 2009 a left frontal skull craniotomy with resection of tumor was performed in a stereotactic procedure. On operation, there was a 1.0-cm-sized, dark red soft tissue mass on the brain cortex. The tumor was removed completely en bloc. After surgery, the patient only took anticonvulsants for 1 week to prevent epilepsy. There was no seizure episode following the operation. The patient was subsequently referred to a radio-oncologist and followed up at the outpatient department.

Figure 1 An axial contrast-enhanced brain computed tomography (CT) scan revealing a tumor with high density in the superior part of the left frontal lobe (arrow).

Figure 2 Axial T2-weighted magnetic resonance image with gadolinium demonstrating a well-defined homogeneously hypointensity mass in the left frontal lobe (arrow).

Figure 3 Coronal T1-weighted magnetic resonance image with gadolinium demonstrating a well-defined homogeneous mass in the left frontal lobe (arrow).

Figure 4 This section shows closely packed cells with plump oval or spindled nuclei and scant cytoplasm. The nuclei display hyperchromatism and mild pleomorphism but no mitosis. Several small capillary lumina containing red blood cells are also seen (H&E ×200).
The pathologic examination of the tumor revealed spindle cell hemangioendothelioma. Grossly, the excised specimen was a well-circumscribed, oval dark brown and firm mass, measuring 1.0 × 0.8 × 0.7 cm in size. Microscopically, the tumor contained numerous prominent, mostly thin-walled blood vessels that varied in size (Fig. 4). Immunohistochemical staining for factor VIII-related antigen, CD31, and CD34 was positive in the endothelial cells lining the vascular spaces and the few epithelioid cells, but was generally sparse in the cellular spindled areas (Figs. 5 and 6). Glial fibrillary acidic protein (GFAP) staining showed positive staining in the gliotic tissue of the peripheral tumor area, indicating the central nervous system origin of the tumor (Fig. 7).

3. Discussion

Spindle cell hemangioendothelioma was first described as a low-grade sarcoma with histologic features intermediate between hemangioma and Kaposi's sarcoma. It is difficult to make a specific classification when a given tumor does not conform to the features of a single subtype. Distinct subtypes are described including epithelioid hemangioendothelioma, spindle cell hemangioendothelioma, and malignant endovascular papillary angioendothelioma. This tumor is characterized by the presence of cellular spindled areas alternating with cavernous vascular spaces. In our case, the tumor was composed of numerous thin-walled vessels, in-between which are cells with spindle-shaped nuclei and indistinct cytoplasmic borders. In this case, diagnosis of this vascular tumor, the positive CD31 immunohistochemical stain, a stain specific for endothelial cells, confirmed the presence of endothelial cells as did positive immunoperoxidase stain for factor VIII and CD34. Classification of this patient's spindle cell hemangioendothelioma was challenging. Epithelioid hemangioendothelioma is characterized by rounded, eosinophilic epithelioid cells. Spindle cell hemangioendothelioma is typically described as being composed of two cellular components. Vascular spaces lined by endothelial cells separate cellular areas of spindle cells which are distinguished by the focal presence of rounded endothelial cells which form nests or line vascular channels. Both tumor cell types feature vascular changes in the cytoplasm of some cells. In this case, the tumor did not distinctly possess the features of one tumor cell type; nonetheless, it appears to be most consistent with spindle cell hemangioendothelioma.

Spindle cell hemangioendothelioma of the brain has been rarely reported. Epithelioid hemangioendothelioma has been described in extradural intracranial locations. Soft tissue spindle cell hemangioendothelioma is clinically indolent and apparently histologically benign, but the local recurrence rate is as high as 60%. Aggressive behavior of a benign-looking soft tissue spindle cell hemangioendothelioma with lymph node metastasis requiring radiotherapy has been described previously. Immunohistochemical staining to identify cell proliferation markers, such as MIB-1 for Ki-67 antigen, as indicators of malignant behavior are useful in tumors of the central nervous system, particularly astrocytomas.

The prognosis and treatment of hemangioendothelioma have not been well established. Hemangioma and hemangioendothelioma are moderately radiosensitive tumors. Hemangiomas involving the skeletal system have been treated effectively with local radiotherapy for many decades. Local radiotherapy is also useful in treating inaccessible hemangioendothelioma with good long-term local control. Life-threatening hemangiomas of infancy have been palliated by systemically administered...
α interferon and this modality of therapy may also be of benefit in instances of recurrent hemangioendothelioma, although no data exist regarding the effectiveness of α interferon against hemangioendothelioma.9

4. Conclusions

Spindle cell hemangioendothelioma is a low-grade tumor. Prognosis and treatment of hemangioendothelioma have not been well established. Involved-field radiotherapy is the only modality commonly used following incomplete tumor resection or tumor recurrence. Our patient underwent a gross total resection, but it is too early to state whether this will be of benefit either with regard to survival or delaying time to tumor recurrence. However, the patient has remained symptom-free since completion of surgery, and there has been no evidence of recurrent tumor at follow-up by brain MRI.

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References