

Primary Rhabdomyosarcoma Associated With Tumoral Hemorrhage

—Case Report—

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Abstract

A 42-year-old female presented with right hemiparesis. Computed tomography (CT) demonstrated a slightly heterogeneous high density mass over the left frontal convexity with peritumoral edema, with homogeneous enhancement by contrast material. Magnetic resonance imaging showed the left frontal convexity tumor as a heterogeneous low intensity area on the T₁-weighted image and a heterogeneous high intensity area on the T₂-weighted image, with homogeneous enhancement and dural tail sign after intravenous administration of gadolinium-diethylenetriaminepenta-acetic acid. After admission, the patient suddenly lost consciousness. CT demonstrated aggressive increase in the bulk of the tumor and large peritumoral hemorrhage. The tumor was totally removed. The histological diagnosis was rhabdomyosarcoma. Combined immunohistochemistry and electron microscopy techniques should be used to differentiate rhabdomyosarcoma from malignant meningioma.

Key words: brain, rhabdomyosarcoma, magnetic resonance imaging, immunohistochemistry, electron microscopy

Introduction

Intracranial primary rhabdomyosarcoma occurs either as a primary tumor or as an extension from the primary tumor located in the head and neck. Intracranial primary rhabdomyosarcoma is very rare. The clinical findings in patients with intracranial rhabdomyosarcoma are similar to other brain tumors, manifesting as a space-occupying lesion or focal neurological deficits. However, the characteristic findings of diagnostic imaging are not well known. The diagnosis of rhabdomyosarcoma is based on positive immunostaining for desmin and myoglobin and the ultrastructural features.

We report a case of primary intracranial rhabdomyosarcoma associated with peritumoral hemorrhage and unique neuroimaging characteristics.

Case Report

A 42-year-old female was admitted on November 5, 2000, because of a one-month history of slowly

progressive right hemiparesis. On admission, the patient had no neurological abnormality except for mild right hemiparesis. Chest radiography showed no abnormalities, and all laboratory findings were within normal limits. Computed tomography (CT) demonstrated a slightly heterogeneous high density mass over the left frontal convexity with peritumoral edema, with homogeneous enhancement by contrast medium. Magnetic resonance (MR) imaging showed the left frontal convexity tumor as a heterogeneous low intensity area on the T₁-weighted image and a heterogeneous high intensity area on the T₂-weighted image, with homogeneous enhancement and a cyst formation after intravenous administration of gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA). Coronal MR imaging with Gd-DTPA showed the "dural tail sign" around the tumor attachment (Fig. 1). Right internal and external carotid angiography demonstrated an avascular mass. The CT, MR imaging, and angiography findings suggested atypical meningioma, because the mass was an extraaxial tumor with intratumoral heterogeneous radiological characteristics combined with the presence of the dural tail sign.

She suddenly experienced headache and projec-

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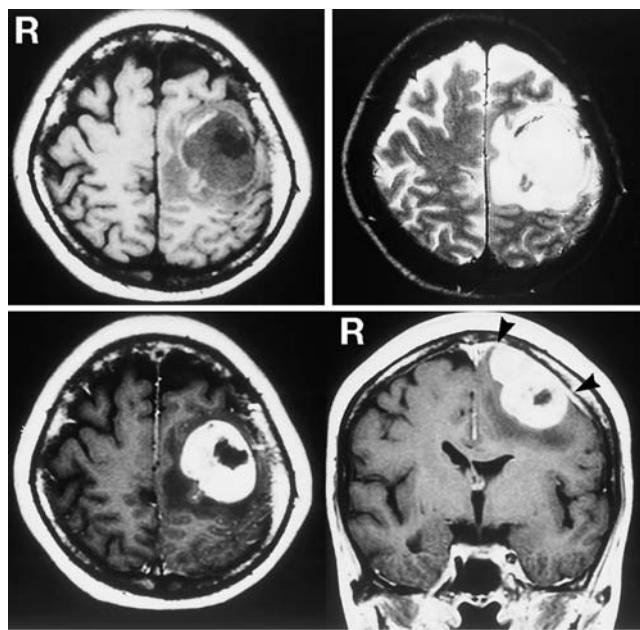


Fig. 1 Magnetic resonance images showing the mass as heterogeneous low intensity on the T₁-weighted image (upper left), and heterogeneous high intensity on the T₂-weighted image (upper right). The mass showed a cyst formation and dural tail sign (arrowheads) after intravenous injection of gadolinium-diethylenetriaminepenta-acetic acid (lower row).

tile vomiting and then lost consciousness on November 12, 2000. CT demonstrated that the bulk of the tumor had aggressively increased and the tumor was associated with large tumoral hemorrhage (Fig. 2). She underwent emergency craniotomy. The dural surface was intact and the tumor was slightly attached to the convexity dura mater. The tumor was grayish brown and showed a tendency to hemorrhage, and the demarcation of the tumor from the brain parenchyma was partially unclear. The tumor was totally removed.

Histological examination showed complicated fusiform or spindle tumor cells, forming intersecting fascicles of cells, which included massive necrotic and hemorrhagic tissues. Abundant nuclear mitotic figures and primitive capillaries were also recognized (Fig. 3A). Immunohistochemical staining revealed positive reaction for vimentin and myoglobin in the tumor cells, but negative reaction for glial fibrillary acidic protein and epithelial membrane antigen (Fig. 3B). Electron microscopy demonstrated that the large cells contained myofilament bundles (actin and myosin filaments) interrupted by Z-discs and minute bundles of filament with dense bodies in

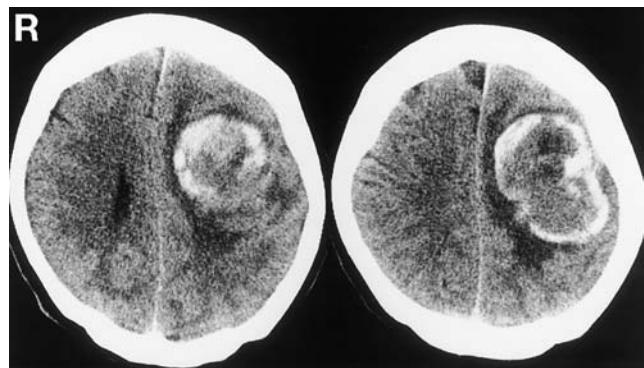


Fig. 2 Computed tomography scans showing aggressive increase in the bulk of the tumor and large tumoral hemorrhage.

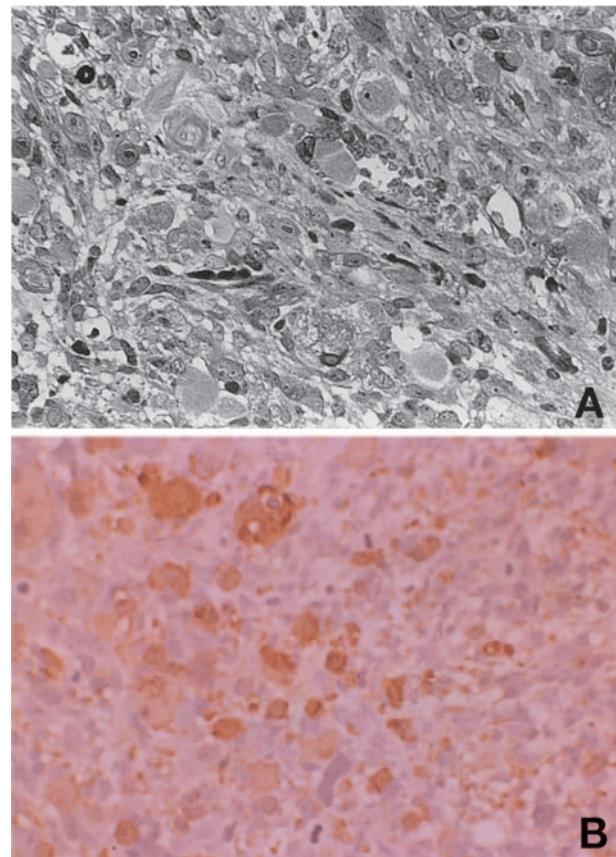


Fig. 3 A: Photomicrograph of the tumor showing complicated fascicles of fusiform or spindle tumor cells with abundant nuclear mitotic figures and primitive capillaries. HE stain, $\times 200$. B: Immunohistological staining for myoglobin showing positive reaction in the rhabdomyoblastic cells. $\times 400$.

the cytoplasm of the small cells (Fig. 4). Based on these findings, the histological diagnosis was rhabdomyosarcoma. Gallium whole body scintigraphy detected no extracranial tumor, so the tumor was considered to be a primary intracranial rhabdomyosarcoma.

Postoperative MR imaging showed radical removal of the tumor. The postoperative course was uneventful. The patient fully recovered consciousness with mild right hemiparesis. She was transferred to another institute for adjuvant therapy.

Discussion

Only 35 cases of the primary intracranial rhabdomyosarcoma have been reported since 1934.^{5,10,13,19,23)} Age at presentation ranged from 1 to 68 years with a slight male predominance (male: female ratio 1.6:1).¹²⁾ Children accounted for 72% of the reported cases.²³⁾

The neuroradiological characteristics of primary intracranial rhabdomyosarcoma are still unclear. Rhabdomyosarcoma shows similar CT and MR findings to meningioma.^{9,11)} Only few cases of this rare clinical entity described with CT and/or MR imaging findings have been reported (Table 1).^{3,8,10-12,22,23,25,26)} Rhabdomyosarcoma appears as a homogeneously or heterogeneously enhanced mass on CT but angiography usually reveals an avascular or hypovascular mass.²³⁾ Rhabdomyosarcomas are frequently accompanied by marked adjacent parenchymal edema, but calcification is rarely present. In our case, the rhabdomyosarcoma was located in the frontal convexity and appeared as a homogeneously enhanced mass on CT and MR imaging with a cyst formation, but angiography showed an avascular mass. Findings of homogeneous enhancement on CT and MR imaging with hypovascularity by angiography might differentiate primary intracranial

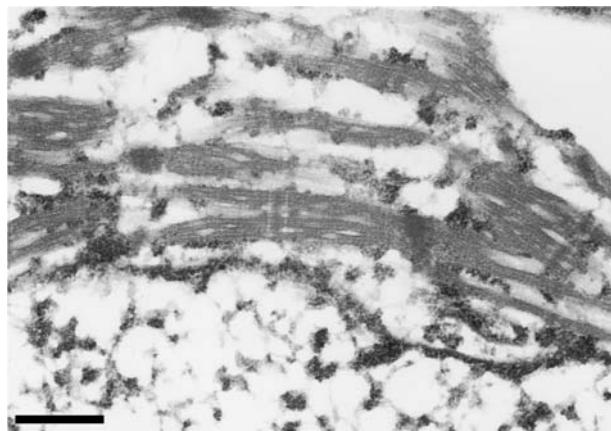


Fig. 4 Electron micrograph of a differentiating myoblast displaying myofilament bundles (actin and myosin filaments) and well-organized formation of Z bands. Bar = 4 μ m.

Table 1 Imaging findings of primary intracranial rhabdomyosarcoma

Author (Year)	Age/Sex	Location of tumor	CT	CT with contrast medium	MR imaging with contrast medium
Namba et al. (1979) ²²⁾	17/M	tentorial apex	ND	hetero	ND
Chiba et al. (1981) ⁸⁾	51/F	frontal	HD	hetero	ND
Olson et al. (1985) ²³⁾	16/M	tentorial apex	ND	homo	ND
	3/F	CP angle	ND	homo	ND
	1/F	CP angle	ND	homo	ND
	1/M	middle fossa	ND	hetero	ND
Dropcho and Allen (1987) ¹⁰⁾	11/F	frontal	ND	homo	ND
Bhatia et al. (1989) ³⁾	10/F	frontoparietal	ND	homo	ND
Tomei et al. (1989) ²⁶⁾	14/F	pineal area	HD	hetero	ND
	14/M	frontotemporal	HD	homo	ND
Hanna et al. (1993) ¹¹⁾	2/F	parietal	HD	hetero	ND
	0/M	frontal	HD	hetero	ND
	5/F	trigone	ND	ND	hetero
Hawkins et al. (1999) ¹²⁾	44/M	frontal	hetero HD	ND	ND
Sugita et al. (2000) ²⁵⁾	80/F	frontal	ND	ND	hetero
	48/F	cerebellum	ND	ND	hetero
Present case	42/F	frontal	hetero HD	homo	homo

CP: cerebellopontine, CT: computed tomography, HD: high density, hetero: heterogeneous, homo: homogeneous, MR: magnetic resonance, ND: not done.

rhabdomyosarcoma from meningioma.

Our patient suffered massive tumoral hemorrhage. Intracranial rhabdomyosarcoma and other sarcomas should be added to the differential diagnosis of tumors manifesting with spontaneous hemorrhage.¹⁰⁾ In the present case, the histological study showed the tumor was rich in primitive capillaries, but the preoperative angiographic study revealed the tumor as an avascular mass. The discrepancy between the histological and the angiographic findings suggests circulation disturbance in the tumor and may be related to the episode of tumor bleeding. Abundant primitive capillaries in rhabdomyosarcoma might be also related to the bleeding tendency. The patient and family should be informed of the possibility of sudden clinical deterioration due to spontaneous tumor bleeding.

Most rhabdomyosarcomas are densely cellular and consist of small cells in which myoblastic components may not be recognized by hematoxylin and eosin staining.⁶⁾ Primary rhabdomyosarcoma of the central nervous system should be differentiated from medullomyoblastomas,²¹⁾ and from other types of brain tumors containing myocystic components, such as teratomas, gangliorhabdomyosarcomas, gliomyosarcomas, mixed mesenchymomas, medullopitheliomas, and intracranial extension or metastasis from rhabdomyosarcoma located in the tissues outside the central nervous system.^{1,2,13-15,17,24)}

Immunohistochemical staining for myoglobin can be used to identify rhabdomyoblastic cells.²³⁾ Myoglobin and desmin are specific for striated muscle differentiation. Myosin is expressed earlier in embryonal myoblastic development than myoglobin, and may be more likely to occur in poorly differentiated tumor cells.¹⁶⁾ Only vimentin and cytokeratin staining cannot differentiate mesenchymal from neuroectodermal cells and are not specific for rhabdomyosarcoma.⁴⁾ Myoglobin is a specific marker for striated muscle and rhabdomyosarcoma.^{14,16)} Immunohistochemical methods can help to establish the differential diagnosis and to characterize rhabdomyosarcoma.^{9,14,18)}

Electron microscopy may be of great help in confirming the diagnosis of rhabdomyosarcoma, particularly when the presence of myofilaments or myofibrils is shown in cells.^{20,27,28)} Myofilaments, either in disarray or in parallel with primitive Z band material, may be observed. In our case, rhabdomyosarcoma was difficult to differentiate from malignant meningioma by light microscopy, but the findings of immunohistochemistry and electron microscopy established the final histological diagnosis as rhabdomyosarcoma. We think that immunohistochemistry and electron microscopy are essen-

tial for the correct differential histological diagnosis of this rare clinical entity from malignant meningioma.

The prognosis for patients with primary intracranial rhabdomyosarcoma is poor, with survival beyond 24 months being exceptional.⁷⁾ Patients with the longest survival have received aggressive treatment consisting of radical gross removal, and high dose (50-60 Gy) whole brain irradiation followed by systemic chemotherapy such as methotrexate, vin-cristine, actinomycin D, and cyclophosphamide.^{10,12)}

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