Ganglioglioma Presenting as a Meningioma: Case Report and Review of the Literature

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OBJECTIVE AND IMPORTANCE: Gangliogliomas are intra-axial, avascular masses located predominately in the temporal lobe. A ganglioglioma that mimics a meningioma in that it is extra-axial and has a significant extracranial vascular supply has not been reported previously.

CLINICAL PRESENTATION: A 12-year-old girl presented with a right temporoparietal mass. A neurological examination revealed nothing abnormal, and the girl's symptoms were limited to headaches.

INTERVENTION: Magnetic resonance imaging revealed an extra-axial mass, and cerebral catheter angiography revealed a blood supply mainly from the posterior division of the right middle meningeal artery. Intraoperative findings confirmed the extra-axial location of the tumor, and histological analysis revealed that the tumor was a ganglioglioma.

CONCLUSION: This report confirms that gangliogliomas can present as extra-axial, vascular masses that are similar to meningiomas.

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Key words: Angiography, Ganglioglioma

Gangliogliomas are rare intra-axial tumors that originate at various sites throughout the craniospinal axis. Most commonly, they present in children as avascular temporal lobe masses that induce seizures. We describe a case of an extra-axial ganglioglioma fed by the posterior division of the middle meningeal artery. A review of the literature reveals only one previous report of a ganglioglioma that presented as a vascular mass (2), but no description of a ganglioglioma mimicking an extra-axial mass supplied by the external carotid arterial circulation similarly to meningiomas.

CASE REPORT

A 12-year-old girl who experienced increasingly painful headaches for several months was referred to our institution. Her medical history was significant only for mild, controlled asthma, and the physical examination revealed nothing abnormal. Magnetic resonance imaging revealed a right posterior temporal convexity extra-axial mass (*Fig. 1*). In addition, preoperative angiography was performed to assess the possibility for embolization. A right external carotid artery injection (*Fig. 2*) demonstrated supply from the posterior division of the middle meningeal artery. The parietooccipital artery of the right posterior cerebral artery supplied a minor component of the tumor (*Fig. 3*). The posterior division of the right middle meningeal artery subsequently was embolized.

Intraoperative findings confirmed the extra-axial location of the tumor. Microscopic pathological findings revealed the tumor subtype to be ganglioglioma (*Fig. 4*). There were no perioperative complications, and the patient was discharged 3 days after the operation. Neither radiotherapy nor chemotherapy was initiated postoperatively. At the 10-month follow-up examination, the patient was doing well and displayed no evidence of tumor recurrence.

DISCUSSION

Epidemiology

Gangliogliomas are relatively uncommon tumors that occur primarily in children and in the temporal lobe. Cushing, in a report of a series of patients with brain tumors, estimated the incidence of gangliogliomas to be 0.3% (3a). Case series published since then have confirmed the low incidence of these tumors. Demierre et al. (4) reported an incidence of 0.6% among 998 patients undergoing biopsy for intracranial tumors. Kalyan-Raman and Olivero (6) reported an incidence of 1.3% in their series. Other series have reported an incidence of 3.8% in patients with brain tumors (14), although other reviewers have stated incidences as high as 6.25% in adults and 10% in children (3). Gangliogliomas in the spinal cord are a rarer entity. Hamburger et al. (5) published descriptions of 2 cases and reviewed 66 known cases. They estimated the incidence of spinal cord gangliogliomas to be less than 1%. Other case reports have illustrated gangliogliomas in unusual locations such as the trigeminal nerve, the suprasellar region, the optic apparatus, and the pineal gland (1, 3). The leptomeningeal subarachnoid spread of tumors has been described as well (13).

Presentation and treatment

Gangliogliomas occur in children and young adults, although one reported case series lacked children (age range, 18–58 yr) (6). Patients who present with this disorder typically have a history of seizure, which may reflect the lesion's tendency to be located in the temporal lobes. In the series of Zentner et al. (14), for instance, 92% of the patients presented with seizures, most of which were resistant to medical therapy. Radical resection remains the treatment of choice and often leads to a good long-

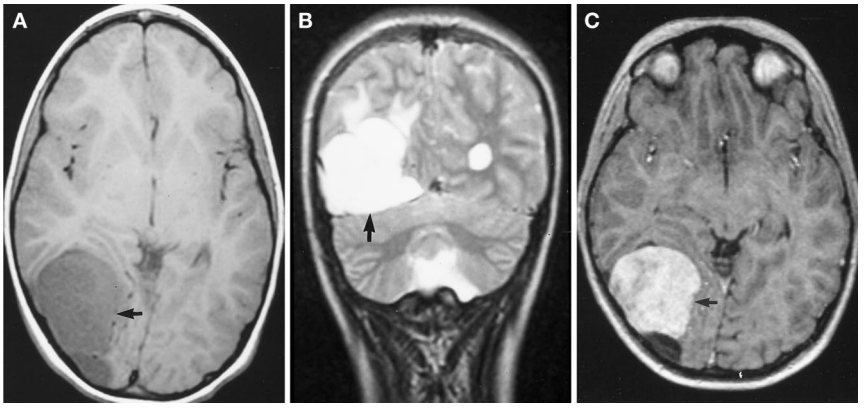

FIGURE 1. Initial magnetic resonance images showing a right posterior temporal convexity extra-axial mass in a 12-year-old girl. *A*, T1-weighted axial scan. *B*, T2-weighted coronal image. *C*, T1-weighted axial scan with contrast.

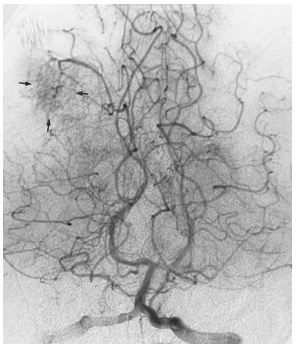

FIGURE 3. Angiogram at left vertebral artery injection site showing minor vascular supply from right parieto-occipital branch artery. *Arrows* point to tumor blush.

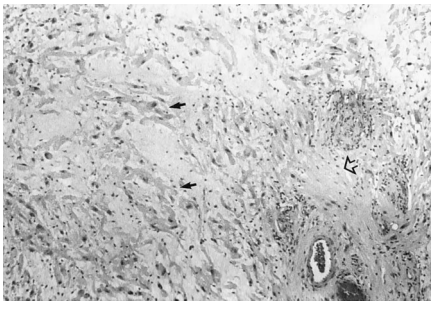

FIGURE 4. Photomicrograph of tumor section showing clusters of irregularly distributed dysplastic neurons (*solid arrows*) and an area composed of benign astrocytes (*open arrowhead*). Perivascular lymphocytic infiltrates are also present (hematoxylin and eosin; original magnification, ×40).

term prognosis. In the series of 58 patients described by Lang et al. (8), event-free survival after surgery was 95% for cerebral tumors but only 36% for spinal cord gangliogliomas. The control of seizures after resection is excellent. In 40 follow-up cases, Zentner et al. (14) reported a seizure-free rate of 80%, with the remaining 20% experiencing a significant reduction in seizures. Postoperative radiation after subtotal resection is controversial but is not indicated after gross total resection (3).

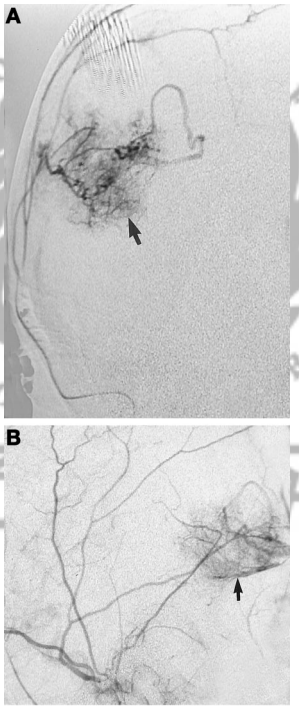

FIGURE 2. Right external carotid artery angiograms with contrast enhancement. *A*, anteroposterior view. *B*, lateral view. *Arrows* indicate tumor arterial blush.

Histopathology

Gangliogliomas are defined by histological criteria. The first cellular component is the ganglion cells, which resemble cortical neurons but are arranged haphazardly. Neoplastic astrocytic cells are a second feature, although cells with oligodendroglial morphology have been described. These glial fibrillary acidic protein-positive astrocytes are intermingled with the ganglion component. Other important features include the absence of perineuronal satellitosis of the astrocytic cells and a lobular pattern due to stromal reaction and lymphocytic infiltrate (9). Immunoreactivity for glial fibrillary acidic protein, neuron-specific enolase, and synaptophysin has been documented and reflects the various components of the tumor. Malignant transformation has been described, mostly because of anaplasia of the astrocytic component. Markers of malignancy (vascular hyperplasia, pleomorphic astrocytes, mitotic figures, and necrosis) are present in as many as 32% of gangliogliomas (9). However, histological grade is not associated with clinical outcome (8).

Imaging

The radiographic appearance of gangliogliomas is variable, but certain characteristics prevail. Imaging studies reveal a well-circumscribed lesion situated in the peripheral cortex, especially in the temporal lobe (4, 12, 14). Computed tomography has revealed the presence of calcification in 41% (14) to 83% (4) of patients and hypodense lesions in 59% (14) to 83% (4) of cases. In one study, magnetic resonance imaging revealed that these lesions were either partially or completely cystic in 57% of 40 reported cases (14). The cystic appearance varies from a single large cyst with a mural nodule to a multicystic mass. Provenzale et al. (12), however, stated that cystic components were more common in children (83%; mean age, 5.5 yr) than in adults (63%; mean age, 25.6 yr). The solid component of the lesion is low to isointense to gray matter on T1-weighted images and hyperintense to gray matter on T2-weighted images (12),

although 32% of tumors were reported to be isointense to hypointense on T2 images in the case series of Zentner et al. (14). A congenital ganglioglioma with high signal intensity on T1-weighted images and hypointense signal intensity on T2-weighted images also has been reported (10). The enhancement pattern varies on magnetic resonance imaging scans, but the signal intensity is usually moderate and homogeneous (11, 12, 14). Perifocal edema and significant mass effect are usually not present.

Other radiographic tools used in diagnosis include positron-emission tomography and angiography. Kincaid et al. (7), for instance, used nuclear medicine in a case series of 11 gangliogliomas. These researchers illustrated tumor hypometabolism in low-grade tumors visualized on [^{18}F]2-fluoro-2-deoxy-D-glucose positron emission tomographic studies and increased activity in two high-grade gangliogliomas seen on ^{201}Tl-enhanced single-photon emission computed tomographic scans. They concluded that nuclear medicine studies have an excellent correlation in preoperatively predicting the histological grade of a ganglioglioma. Provenzale et al. (12) concurred in their series that tumor hypometabolism usually corresponds with low-grade gangliogliomas.

Angiographic appearance

The vascular characteristics of gangliogliomas are rarely reported. Castillo (3) mentioned that angiography is not indicated in the primary evaluation of gangliogliomas. Demierre et al. (4), however, described avascular masses located by performing catheter angiography in 67% of cases, whereas the remainder revealed only "irregular" vessels on the arterial and venous phases. In 1987, Kalyan-Raman and Olivero (6) published the largest series of gangliogliomas studied angiographically. In that series, 4 of 10 cases were avascular, and the remainder were hypovascular masses with a small area of abnormal vascularity. One previous case report described a markedly vascular ganglioglioma that was supplied entirely by the internal carotid artery system (2). To our knowledge, our report is the first to describe a histologically proven ganglioglioma supplied primarily by the external carotid arterial system. This pattern of external arterial feeders indicates that this tumor was extra-axial in nature, which is corroborated by radiographic and intraoperative data. If so, this article would constitute the first to report an extra-axial supratentorial ganglioglioma.

CONCLUSION

Gangliogliomas at many sites along the craniospinal axis have been described. Although variability in radiographic appearance is acknowledged, certain characteristics (temporal location, cystic and calcified components) do exist. An extra-axial location and an angiographically robust appearance can be added to the possible characteristics of these uncommon tumors. Although we do not recommend catheter angiography in all suspected cases of gangliogliomas, certain cases—especially those that mimic typical dura-based lesions such as meningiomas—may benefit from preoperative evaluation of arterial supply. However, a robust vascular supply to a ganglioglioma, although atypical, does not necessarily portend a prognosis of malignancy.

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COMMENTS

Siddique et al. report an unusual case of a ganglioglioma mimicking a meningioma. Because this tumor was large and in a child, there would be little controversy regarding the need for surgery to be performed. Certainly, the preoperative angiogram suggested a meningioma, and embolization was appropriate. This case illustrates a frequent observation in tumor operations—the situation is not always as it seems, and a biopsy of the tumor is almost always necessary.

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Siddique et al. describe a case of a 12-year-old girl who presented with a large temporoparietal mass and headache. Preoperative imaging revealed the mass to be extra-axial, and this location was confirmed during the operation. The histological examination of the specimen revealed the diagnosis of ganglioglioma.

The authors indicate that this lesion presented as a meningioma. This diagnosis was made primarily on the basis of the extra-axial location of the tumor and on

the meningeal blood supply from the external carotid artery circulation. The angiogram is striking and highly suggestive of meningioma. Although these features are typical of meningiomas, they are not enough to substantiate the diagnosis. Moreover, some radiographic features in this case are atypical for meningiomas. The magnetic resonance imaging scan demonstrates an inhomogeneous enhancing lesion with an associated cyst. The lesion is hyperintense on the T2-weighted image. Certainly, meningiomas can be hyperintense on T2-weighted images, but they are not usually hyperintense to this degree. Although no computed tomographic scans are presented, the magnetic resonance imaging scans do not demonstrate any hyperostotic changes. Finally, and most important, no evidence of a "dural tail" or any other enhancing dural attachment exists.

Although we disagree that the ganglioglioma described in this case report presented as a meningioma, we acknowledge the unusual presentation of the tumor, the features of which Siddique et al. recognized and reported. This report adds useful information to the literature regarding these rare primary brain tumors—namely, that they may appear in an extra-axial location on preoperative imaging and that they may receive blood supply through the external carotid artery circulation via the meningeal branches.

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Siddique et al. present the first reported case of a supratentorial extra-axial ganglioglioma. The extra-axial location of this child's tumor was first defined by magnetic resonance imaging and angiography and then confirmed by intraoperative findings. The diagnosis was made histologically. Although this case admittedly represents a rare presentation of a rare tumor, it is significant in that it introduces extra-axial location and marked vascularity as possible characteristics of gangliogliomas. Siddique et al. state that such presenting characteristics do not necessarily portend a malignant prognosis. Obviously, to bolster this assertion, it is important to continue the follow-up beyond the 10-month follow-up examination and document the patient's continued good condition.

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