Ganglioglioma presenting as a vascular lesion in a 10-year-old boy

Case report

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The authors present the case of a 10-year-old boy admitted for evaluation of a generalized seizure and a history of headaches. Computerized tomography (CT) and gadolinium-enhanced magnetic resonance (MR) imaging demonstrated a large nonhomogeneous contrast-enhancing mass of the left frontal lobe, with a large cystic component. Cerebral angiography revealed the lesion to be highly vascular and fed entirely by the internal carotid artery system. The patient underwent craniotomy and the lesion was completely removed. Neuropathological study revealed that the tumor was a ganglioglioma. On review of the literature, it was found that gangliogliomas often present in the second and third decade, are known to have cystic components, and are contrast-enhancing on CT and MR imaging; however, they are classically known to be avascular on angiography. This case of a markedly vascular ganglioglioma emphasizes that these tumors should be included in the differential diagnosis of vascular supratentorial lesions.

KEY WORDS • brain neoplasm • ganglioglioma • magnetic resonance imaging • cerebral angiography

Gangliogliomas are rare tumors making up 0.4% of all brain tumors and 1.2% to 7.6% of pediatric cerebral neoplasms.1,1,13 The lesions most often occur in the temporal lobe but have been found elsewhere in the cerebrum. A frequent presentation is seizures.12 These tumors are normally contrast-enhancing on computed tomography (CT) and magnetic resonance (MR) imaging and often have cystic as well as solid components; however, they are classically avascular on angiography.6,9 The case is reported of a highly vascular frontal lobe lesion in a 10-year-old boy, which had the pathological features of a ganglioglioma. The pathology is discussed and the literature reviewed.

Case Report

This 10-year-old right-handed boy was transferred to our care from the emergency room with a new-onset generalized seizure. He had been well until 5 years prior to admission, when he suffered a mild head injury. At that time, there was no change in his level of consciousness, but he experienced headache with nausea and vomiting. A skull x-ray film was normal, but an electroencephalogram (EEG) demonstrated excessive delta-wave abnormalities over the left frontal region. The patient continued to experience intermittent headaches over the left frontal region, which were considered to be migraines. His developmental history was normal, as was his family history.

Examination. Physical examination revealed mild bossing over the left frontal region. The neurological examination was remarkable for an isolated right pronator drift. No papilledema, extraocular paresis, or frontal lobe syndrome was found. Chest x-ray and abdominal ultrasound studies were normal, as were laboratory investigations.

A CT scan demonstrated a 5 × 5 × 8-cm mass lesion in the left frontal lobe. The tumor was mainly cystic, with a large 5 × 4-cm enhancing component adjacent to the frontal convexity dura. There was a shift of midline structures from left to right, with partial subfalcine herniation (Fig. 1). Left internal carotid angiog-
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Fig. 1. Computerized tomography scan showing a 5 × 5 × 8-cm mass in the left frontal lobe that is mainly cystic, with a large enhancing central component. A shift of midline structures is seen.

Fig. 2. Left internal carotid angiogram revealing a large frontal vascular mass with blush which persisted throughout the capillary-venous phases. Supply is principally from the left middle cerebral artery (anterior temporal and frontal polar branches). Drainage occurs via the inferior sagittal sinus and the cerebral cortical veins to the superior sagittal sinus. A left external carotid injection (not shown) demonstrated no abnormality.

Fig. 3. Sagittal magnetic resonance image with gadolinium enhancement demonstrating a contrast-enhancing lesion with a large, septated cystic component. The solid component enhanced on gadolinium injection.

Fig. 4. Intraoperative photograph revealing exposed tumor with marked vascularity, a nonhomogeneous appearance, and large draining veins, and a surrounding encephalomalacic cyst. Note the proximity of the tumor to the convexity dura.

Magnetic resonance imaging with gadolinium injection showed a contrast-enhancing lesion with a large surrounding septated cystic component. The enhancing solid component suggested a highly vascular lesion (Fig. 3). The lesion was adjacent to the frontal convexity dura.

Operation. At surgery, the left frontal convexity bossing was confirmed. The tumor was highly vascular, and demonstrated large veins draining into the sagittal sinus (Fig. 4). The arterialized blood within the cortical veins confirmed rapid shunting. The surrounding cyst was entered, and the central core was progressively devascularized. The venous drainage was coagulated last and the tumor removed en bloc. The septae within the cyst...
and attached to the surrounding encephalomalacic brain were sectioned by biopsy.

Pathological Examination. On gross inspection, the surgical specimen was firm, pink, and homogeneous with a multigranular surface resembling a cauliflower. Histological sections revealed that the central two-thirds of the lesion was composed of numerous thick and hyalinized vessels, some being small veins and arteries, many of which were completely obliterated by collagen. These vessels were surrounded by groups of cells resembling oligodendrocytes (Fig. 5a), with round and oval nuclei that were slightly hyperchromatic and polymorphic in a clear cytoplasm (Fig. 5b). Microcysts were found, and periodic acid-Schiff-positive droplets were seen accompanying the proliferation. The fibrillary processes of other cells in this region suggested astrocytes; within these proliferations, there were large neuronal cells with occasional Nissl substance. The periphery of the tumor was less vascular, but the cellularity was increased, with clusters of astrocytic, oligodendrogial-like cells and neurons (Fig. 5b). The vessels in this region were less numerous, with thin and less hyalinized walls. The major part of the tissue was loaded with dense collagen fibers (Fig. 5c). A specimen from the cyst wall of the lesion revealed astrogliosis and fibrovascular tissue with macrophages containing hemosiderin pigments. Within the tumor, immunohistochemistry showed clusters of glial fibrillary acidic protein-positive cells (Fig. 5d), often surrounding a large synaptophysin- (Fig. 5e) or neuron-specific enolase (NSE)-positive cell (Fig. 5f). Some of the cells described as oligodendrogial-like were NSE-positive, while others were completely negative.

Ultrastructural examination revealed mainly tumor cells, with round or oval nuclei, in a cytoplasm loaded with polyribosomes or filled with microfilaments. Other cells showed neurotubules and dense junctions between processes, but no well-defined synaptic complexes were found. Occasional cells showed a basal lamina around the cytoplasmic membrane. These findings suggested a mixture of oligodendrogial-like cells, astrocytes, and neuronal elements consistent with a ganglioglioma.

Postoperative Course. At 2 years postsurgery, the patient is seizure-free while taking anticonvulsant drugs.
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There was a complete tumor resection as revealed on MR imaging and there is no evidence of tumor recurrence.

Discussion

The radiological differential diagnosis of such a highly vascularized and yet cystic lesion in proximity to the dura includes meningioma, hemangiopericytoma, hemangioblastoma, and glioma. The first two lesions were considered unlikely once the external carotid angiogram failed to demonstrate increased vascularity. Our working diagnosis was supratentorial hemangioblastoma until the pathological characteristics became available.

Gangliogliomas are often characterized by a mixture of differentiated elements, oligodendrocytes, astrocytes, and neuronal cells, all of which were observed in this lesion. The immunohistochemical study showing irregularly placed NSE-positive cells as well as the ultrastructural studies demonstrating neurotubules and dense junctions confirm the diagnosis of ganglioglioma. The vascularity seen in this tumor is, however, highly unusual in a ganglioglioma, and the reason for it is not clear. Our first thoughts were that the hypervasculartiy might be related to trauma. The abnormal EEG at the time of a minor head injury 5 years previously suggests the lesion was long-standing. The hemosiderin found in the cyst wall and at the tumor surface could suggest a remote hemorrhage in or around the lesion. However, due to the lack of external carotid artery feeding vessels and of necrosis around the tumor, and the clearly neoplastic nature of the tumor vessels, it is unlikely that trauma is the sole cause of the hypervasculartiy.

High-grade gliomas may show hypervasculartiy on angiography, but it is rarely so marked as the hypervasculartiy demonstrated in Fig. 2. In addition, although the vessels were neoplastic in nature, their tendency to hyalinize and to be obliterated by collagen as well as the cellular characteristics of the tumor suggest that this lesion is benign.

The oligodendroglial component of the ganglioglioma found at pathological examination was important. There have been reports of oligodendroglioma-like lesions presenting in association with arteriovenous malformations (AVM's). These were thought by some authors to represent oligodendrogliomas and by others to represent an oligodendroglial proliferative abnormality associated with an AVM, rather than a true oligodendroglioma. In the tumor described in this paper, however, venous and arterial elements were both present but never found intermixed, as would be the case with an AVM. Unfortunately, angiogenic factors such as fibroblast growth factor and angiogenin were not assayed within the tumor, but this is being considered for future cases.

In conclusion, ganglioglioma should be included in the differential diagnosis of highly vascularized brain tumors, when the history and physical examination suggest a long-standing process and when the patient presents with seizures. The increased vascularity seen in the ganglioglioma described in this paper, we believe, does not adversely affect the usual good prognosis of completely resected lesions of this nature.

Acknowledgment

We thank Dr. Peter C. Burger of Duke University Medical Center for reviewing the pathology in this case.

References