Pleomorphic Xanthoastrocytoma Presenting with Massive Intracranial Hemorrhage

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Summary: A 46-year-old woman presented with massive left temporal lobe intraparenchymal and diffuse subarachnoid hemorrhage on CT. Lack of enhancement excluded intermediate and high-grade primary tumor or metastasis as likely causes. At surgery, a fibrovascular left temporal lobe mass adherent to the dura proved to be a pleomorphic xanthoastrocytoma. The unusual hemorrhagic presentation of this typically benign entity is thought to be related to the meningeal involvement, which itself is characteristic of this neoplasm.

Index terms: Astrocytoma; Brain neoplasms, computed tomography

We present a case of temporal lobe pleomorphic xanthoastrocytoma presenting with massive intraparenchymal and subarachnoid hemorrhage.

Case Report

A 46-year-old white woman with a history of intermittent headaches for 1 year reported “not feeling well” the night of admission. She was otherwise in good health except for an abnormal mammogram awaiting breast biopsy. On the night of admission, the patient lay down on her couch and became unresponsive. She was transported to an outlying hospital and required intubation. Blood pressure was 170/90 mm Hg. She would respond only with flexion to pain on the left and was flaccid on the right. An outside computed tomographic (CT) scan of the brain revealed left-sided temporal intraparenchymal and subarachnoid hemorrhage.

The patient was transferred to our institution on a ventilator; she had stable vital signs otherwise, having received 25 g of intravenous mannitol. Neurologically, she was unresponsive to voice and would respond by withdrawing to pain on the left more than the right. She had a positive right Babinski sign and was mildly hyperreflexic throughout. Follow-up CT was obtained, en route to which a dilated left pupil developed. This CT scan (Fig 1A and B) again revealed massive left temporal lobe hemorrhage, subarachnoid hemorrhage, and transtentorial and subfal-}

cine herniations. Because of findings of progressive transtentorial herniation, the patient was taken to surgery without a cerebral angiogram, with a provisional diagnosis of ruptured left middle cerebral artery aneurysm.

The patient underwent a left frontotemporal craniotomy. After a significant amount of blood had been removed from the middle cranial fossa, including the superior and middle portions of the temporal lobe, a tannish gray fibrous mass that was vascular was uncovered. The mass was growing into the dura mater of the floor of the middle cranial fossa and cavernous sinus. As much of the tumor as possible was resected, with specimens sent for pathologic analysis; the dura mater was then irrigated and closed.

On pathologic examination, the neoplastic astrocytes that composed this tumor were immunoreactive for glial fibrillary acidic protein, positive for S-100 protein, epithelial membrane antigen negative, and low-molecular cyto-keratin negative, with positive reticulin-staining paren- chyma representing collagen interlaced between neo- plasticallymphocytes. Cells with clear, round cytoplasmic vacuoles were present. The tumor had a pleomorphic appearance and was located superficially (Fig 1C). The diagnosis of pleomorphic xanthoastrocytoma was made. A Movat’s pentachrome stain revealed collagenous thickening and focal intramural lymphocytes in a few vessels within the tumor, but neither this nor the other stains provided a histologic explanation of hemorrhage in this case.

The patient’s postoperative course was characterized by difficulty in controlling intracranial hypertension. A left middle cerebral arterial distribution infarction developed on the third postoperative day. The patient died on the sixth postoperative day after withdrawal of life support systems.

Discussion

Pleomorphic xanthoastrocytoma is a rare glial neoplasm thought to originate from subpial astrocytes (1), representing a distinct form of supratentorial astrocytoma. It was described by Kepes et al in 1979, in 12 patients, the majority
of whom presented with seizures in the first three decades of life. All had superficial cortical tumors with extensive leptomeningeal involvement, usually in the temporal lobe (2). Subsequent radiologic investigations have supported the typical occurrence of the tumor equally within both sexes in the first three decades of life (3), a temporal lobe predilection (3, 4), and a tendency to present with seizures, although headache is also common (1, 3, 4). These neoplasms are typically cortex based with meningeal involvement, although meningeal enhancement on imaging studies is a less constant finding than enhancement of the intraxial component (3, 4).

In one radiologic series, five of seven pleomorphic xanthoastrocytomas showed leptomeningeal involvement, and one of seven showed dural adherence (3). In another report, two of two cases showed dural attachment (3), and in a third series, two of six tumors showed leptomeningeal involvement (4).

Associated intraxial cyst formation is common, occurring in three of seven patients in one series (3) and two of six in another (4). The tumor exhibits mixed attenuation on CT with variable patterns of enhancement, including gyriform (3–5). Calcification is uncommon (3). On magnetic resonance imaging, the tumor is uniformly hyperintense on T2-weighted images, with most either hypointense or isointense on T1-weighted imaging, and there is a report of T1 hyperintensity. The tumor enhances with gadolinium (3, 4). The tumor is characteristically hypovascular at angiography, although intense tumor stain has been reported (1, 4).

Histologically, xanthoastrocytoma is pleomorphic with varying admixtures of spindle...
cells, multinucleated giant cells and foamy lipid-laden xanthomatous cells, with mitosis and necrosis noticeably absent. Glial fibrillary acidic protein stains positive, indicating an astrocytic origin. Demonstration of the basal laminae by electron microscopy implicates the subpial astrocyte as the progenitor cell (6).

The catastrophic clinical presentation in this case is unusual. It is proposed that the temporal lobe and adjacent meningeal involvement by the neoplasm eventually produced erosion of an artery on the surface of the brain and caused massive intraparenchymal and subarachnoid hemorrhage. Thin-walled tumor vessels in this case could also be contributory to the hemorrhage, but this feature has not been described as occurring in pleomorphic xanthoastrocytoma (7). There was no history of bleeding diathesis or hypertension to complicate this event. The propensity for pleomorphic xanthoastrocytoma to invade both leptomeninges and pachymeninges is well known (1, 3, 4). It is therefore interesting that vascular invasion with hemorrhage is not more characteristic of these tumors. We found no reports of tumor-associated hemorrhage. In fact, their behavior is indolent, with long postoperative survival common (6). Recurrence usually is in the form of small-cell glioblastoma multiforme; these rare cases of pleomorphic xanthoastrocytoma with a rapidly fatal outcome have been reported (8).

Histologically, no atypical features were identified in this case. The massive intracranial hemorrhage destroyed brain and meningeal tissues invaded by tumor, precluding an optimal search for vascular invasion. None was apparent otherwise. The differential diagnosis for the CT findings in this clinical setting includes ruptured left middle cerebral artery aneurysm, ruptured vascular malformation, and hemorrhagic neoplasm, especially metastatic tumor, glioblastoma multiforme, or oligodendroglioma, all with hematoma replacing much of the temporal lobe. The absence of contrast enhancement associated with the temporal lobe hematoma mitigates against the possibility of aneurysm or large arteriovenous malformation. Metastatic tumor or glioblastoma multiforme would, similarly, be expected to exhibit some enhancement. Oligodendroglioma is a rarer glial tumor that could exhibit less enhancement and may be more likely than glioblastoma multiforme to bleed grossly (9). Of note, a punctate focus of calcification was present in the left temporal lobe. Although nonspecific, either vascular malformation or tumor are compatible with this.

In summary, in this case of massive intracranial hemorrhage complicating a left temporal lobe pleomorphic xanthoastrocytoma, we propose that vascular invasion by the tumor represented an event consistent with pleomorphic xanthoastrocytoma’s tendency for meningeal extension, and that resultant hemorrhage into the tumor replaced much of the temporal lobe. The extent of the tumor’s involvement in the temporal lobe remains unclear because of the presence of hematoma, which destroyed brain tissue.

References