Pleomorphic Xanthoastrocytoma: A Case Report

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Purpose: To draw attention to the radiological findings of a benign variant of cerebral astrocytoma in a young patient.

Materials and Methods: A 24-year-old man with generalized tonic-clonic seizure of 7 years' duration and normal neurological examination was examined with plain skull series, brain CT and MRI, and cerebral angiography. MR imaging was performed with a 0.5 Tesla Toshiba MRT-50A scanner (T1WI, PDWI, T2WI, 0.1 mmol/kg of Gd-DTPA, SE).

Results: 1) Plain skull series: A radiolucent lesion with a partial radiopaque rim of about 2.5 x 3 cm size in the right anterior parietal bone. 2) Brain CT scan: A cystic mass in the right frontoparietal cortex of midconvexity with pressure erosion on the adjacent skull and partial enhancement at outer and anterior portion. 3) Brain MRI: A hypointense mass containing a small, intensely enhancing isointensity anterolaterally on T1-weighted images, which was hyperintense with better delineation of bulging cortical-based appearance on T2-weighted images. No peritumoral edema. 4) Cerebral angiography: An avascular mass.

Conclusion: The diagnosis of pleomorphic xanthoastrocytoma (PXA) should be entertained in patients in whom a superficially placed enhancing intracerebral tumor containing cystic portion that seems to be in contact with the meninges develops during juvenile years.

Index Words: Brain CT, neoplasms
Brain MRI, neoplasms

INTRODUCTION

Pleomorphic xanthoastrocytoma (PXA) is a low-grade leptomeningeal glioma affecting young patients (second and third decades) with favorable prognosis, which was first described by Kepes et al. in 1979 as a clinicopathologically distinct variant of cerebral astrocytoma (1). The tumor is most often located superficially in the cerebral hemispheres, typically the temporal or temporoparietal region involving leptomeninges but not dura, and is a circumscribed tumor with a cystic component. Solid portion of the tumor shows intense contrast enhancement on both CT and MRI and surrounding leptomeningeal enhancement is seen occasionally. Seizures and headaches are common clinical features. The tumor cells display marked pleomorphism including bizarre giant cells and scanty mitotic figures with no necrosis. Many contain large amounts of lipid in their cytoplasm and are surrounded by reticulin fibers, thus simulating a mesenchymal tumor. The tumor is claimed to arise from subpial astrocyte. The astrocytic origin of the tumor is confirmed by immunoperoxidase staining for glial fibrillary acidic protein (GFAP) (1-4).

CASE REPORT

This 24-year-old man was admitted to our hospital for clinical evaluation of his generalized tonic-clonic seizure of 7 years' duration. Neurological examination was normal. Plain skull revealed a radiolucent lesion with a partial radiopaque rim of about 2.5 x 3 cm size in the right anterior parietal bone. On CT scan (Fig. 1a, b, c) a cystic mass in the right frontoparietal cortex of mid-convexity with pressure erosion of the adjacent
skull was noted. Outer and anterior portion of the lesion was enhanced after administration of contrast material. The tumor was hypointense containing a small iso-intensity laterally on precontrast T1-weighted image (Fig. 2a) and enhanced intensely anterolaterally on postcontrast T1-weighted image (Fig. 2b). On T2-weighted image (Fig. 2c) it was hyperintense and its bulging and cortical-based appearance was more distinct on this image than on T1-weighted image. There was no peritumoral edema. Right carotid angiogram showed an avascular mass in the frontoparietal region.

Right frontoparietal craniotomy disclosed a superficial frontoparietal mass. This was covered by the leptomeninges but it was not attached to the dura mater. The mass was soft and yellowish, extended into the cortex, and was relatively well delimitated from the surrounding brain. The patient recovered and was discharged after 13 days.

Histological examination (Fig. 3) revealed astrocytic proliferation with GFAP-positive multinuclear large pleomorphic cells having abundant pale acidophilic cytoplasm. Mitosis and necrosis were not present. The neoplastic cells were surrounded by reticulin fibers on reticulin stain.

**DISCUSSION**

Features supporting the diagnosis of pleomorphic xanthoastrocytoma (PXA) are: (a) clinical-young patients, with epilepsy being a prominent feature; (b) gross pathology-temporal and temporoparietal sites with superficial location involving the leptomeninges but not dura mater and with a circumscribed nature and a cystic component; (c) microscopic pathology-pleo-

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![Precontrast CT(a) shows a well-defined cystic mass in the right frontoparietal cortex. The lesion is well enhanced anterolaterally on postcontrast scan(b). Erosion of the underlying skull is well seen on the bone algorithm(c).](image-url)
morphism, numerous giant cells, GFAP stain-positive in many of the tumor cells; (d) benign behavior.

The recognition of this relatively recently identified entity is important on two counts: first, because of the striking disparity between its ominous-looking histological features and its usually favorable postoperative course; and, second, because its pleomorphic microscopic appearances have led to its designation under several different diagnostic labels such as glioblastoma multiforme, atypical fibroxanthoma, monstrella sarcoma, unclassified glioma, and possibly, giant cell astrocytoma with histiocytic infiltrate. The radiologic features are therefore especially important since they may call attention to the possibility of PXA prior to histologic evaluation and can avoid overly aggressive treatment.

So far all the examples of this tumor have been supratentorial. They were typically superficial, often occupying the cerebral cortex over a relatively restricted field only and extending in the overlying leptomeninges, but not the dura. Most of them were less than 3.5 cm in diameter and had not peritumoral edema. Nevertheless, in only one patient of Kepes’s 12 cases there was radiological and operative evidence of erosion of the adjacent skull as in our case, and we could not find other cases with the skull erosion in the literatures.

The circumscribed nature and superficial location are in contradistinction to usual gliomas, which arise deep in white matter and infiltrate widely. Although PXA is an astrocytoma and occurs in young indivi-

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**Fig. 2.** Axial T1-weighted(400/15/2) MR image(a) shows a bulging cystic mass containing a small solid portion superficially. It is strongly enhanced anterolaterally on axial Gd-DTPA T1-weighted image(b) and is hyperintense on coronal T2-weighted image(3000/120/1)(c) with more distinct cortical-based bulging contour than on T1-weighted images.

**Fig. 3.** Photomicrography of pathologic specimen(Hematoxylin-Eosin; original magnification, ×400) shows multinucleated pleomorphic cells having abundant pale acidophilic cytoplasm without evidence of mitosis and necrosis.
other tumors appears lobulated intracerebral lesions with variable calcification. On MRI, most of the tumors are well-circumscribed and cortical-based and isointense with gray matter on T1-weighted images and hyperintense on T2-weighted images. All the masses enhance with contrast material. Cystic components and gyriform and leptomeningeal enhancement are seen occasionally(2-4). The radiological differential diagnosis of a cortical-based enhancing mass should include meningioangiomatosis, PXA, hemangiopericytoma, lymphoproliferative mass, anaplastic astrocytoma, metastasis, inflammatory mass, and granulomatous disease(e.g., sarcoma, fungal disease)(2-4, 8-9). Our case fulfills all features of PXA as described by Kepes and others.

The superficial location of this tumor and its relationship to the leptomeninges facilitates surgical extirpation, but poor definition of the tumor-brain interface may preclude total removal. In the reported cases, even if the tumor is not totally removed, later surgery for residual or for local recurrence of PXA has been well tolerated and can lead to useful control of symptoms. Death or follow-up period of more than 2 years following histological diagnosis of PXA is documented in 26 of 35 reported cases. The original description by Kepes, et al., of a favorable prognosis with PXA correct in 17 of the 26 patients followed for more than 2 years aliving a mean of 7.4 years(range 2 to 18 years). Nine (35%) of 26 patients with PXA in whom outcome details are available have died and in 5 cases, death has occured within 2 years of diagnosis, who received radiotherapy. Park, et al.(7) reported a recurrent intramedullary PXA arising from conus medullaris in a 41-year-old man who died 7 months after the second operation. He had received radiotherapy after the first operation. Their all 3 cases of supratentorial PXA survived without radiotherapy. The role of radiotherapy in the management of PXA is unclear(6, 8). Allegranza, et al.(10) suggested that PXA may in exceptional cases recur and display overt anaplastic change in their report of a giant cell glioblastoma evolving from PXA 8 years ago.

The diagnosis of PXA should be entertained in patients in whom a superficially placed, enhancing intracerebral tumor containing cystic portion that seems to be in contact with the meninges develops during the juvenile years.

REFERENCES