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M T Yoshino and R Lucio

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Pleomorphic Xanthoastrocytoma

Mark T. Yoshino¹ and Richard Lucio²

Summary: We report two cases of pleomorphic astrocytoma, a rare primary brain tumor with an unusually good prognosis. These lesions typically are peripherally located, partially cystic, temporal lobe masses. Dural attachment is frequent, calcifications are not. Angiography usually shows a hypovascular mass, although blood supply from the external carotid artery may be seen.

Index terms: Astrocytoma; Brain neoplasms, in infants and children

Pleomorphic xanthoastrocytoma is a rare tumor that has an unusually good prognosis, 10- and even 20-year survivals being reported after surgery (1, 2). Typically, this subtype of astrocytoma presents with seizures beginning in childhood and on computed tomography (CT) is found to be a superficial, partly cystic mass located in a temporal lobe (1-4). The purpose of this report is to describe the findings in two cases of pleomorphic xanthoastrocytoma and suggest imaging features that should place this tumor high in the radiographic differential diagnosis.

Case Reports

An 11-year-old boy (Case 1) presented with a grand mal seizure. Physical examination was remarkable only for papilledema. Head CT revealed a superficial, markedly enhancing right parietal mass with associated vasogenic edema (Fig. 1A). A large cystic component was present. The adjacent inner table of the skull was eroded. Right internal carotid angiography demonstrated a large hypovascular right parietal mass with only a faint blush in the capillary phase. Right external carotid arteriogram (Fig. 1B) also revealed a faint stain, the tumor being supplied by branches of the middle meningeal artery.

At surgery, the mass was found to be partly cystic and attached to the dura, but was easily removed from the adjacent normal brain. Histologic examination showed pleomorphic glial fibrillary acidic protein (GFAP) positive cells

with occasional atypical mitoses, a prominent reticulin network, and no necrosis.

A 9-year-old boy (Case 2) had a history of chronic headaches and recent onset of right-sided visual loss. Physical exam revealed bilateral papilledema and positive snout reflex, but no other focal findings. CT demonstrated a large, mostly cystic, right frontal lobe mass with densely enhancing peripheral nodules (Fig. 2A). Brain magnetic resonance (MR) imaging also revealed the mass that, as expected, had increased signal intensity on proton density and T2WI in both its cystic and solid compartments (Figs. 2B and 2C). No increased signal on T1WI was present in the tumor prior to contrast administration, but on post-Gd-DTPA study the nodular portion of the tumor markedly enhanced (Fig. 2D).

Craniotomy revealed a superficial, partly cystic, frontal lobe mass with adhesions to the dura. It was easily delineated from the adjacent normal brain except inferiorly, where macroscopic infiltration was apparent. Microscopic examination showed pleomorphic, GFAP positive cells surrounded by a prominent reticulin network. Scattered xanthoma cells were noted. Mitoses were present, but uncommon, and no necrotic areas were encountered.

Discussion

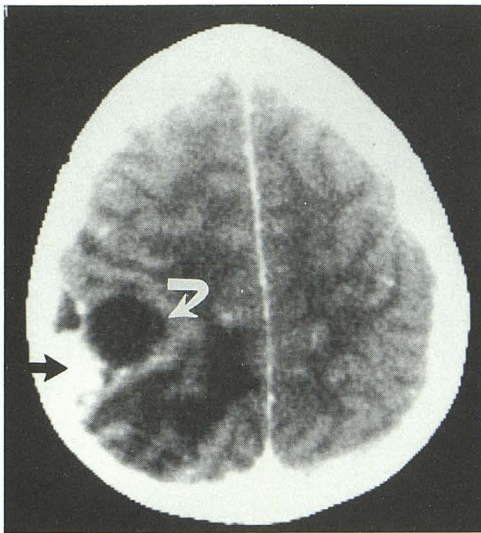
Pleomorphic xanthoastrocytoma was described in 1979 by John Kepes, a pathologist who reported a series of 12 patients (1). Since that time approximately 35 additional patients have been reported (3-12). Histologically, the tumors are markedly pleomorphic, but each cell contains a variable amount of lipid in its cytoplasm and is surrounded by reticulin fibers. The glial origin of these tumors is confirmed by positive GFAP staining. Because of the peripheral location of these lesions, they are believed to arise from subpial astrocytes (8). On gross examination, these tumors usually involve both the leptomeninges and underlying brain and frequently have a cystic component, although areas of frank necrosis are rare.

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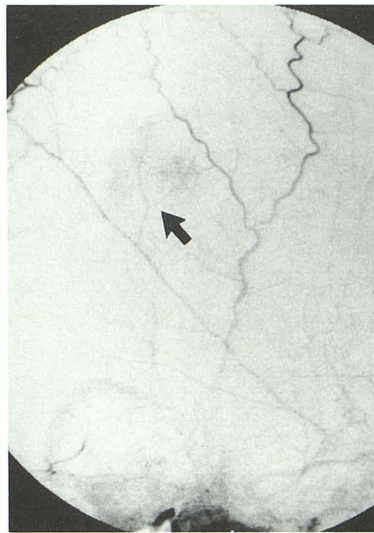
¹ Department of Radiology, University of Arizona Health Sciences Center, 1501 North Campbell Avenue, Tucson, AZ 85724. Address reprint requests to Mark T. Yoshino, MD.

² Richard Lucio, MD, 414 Windrift Drive, Belleville, IL 62221.

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A

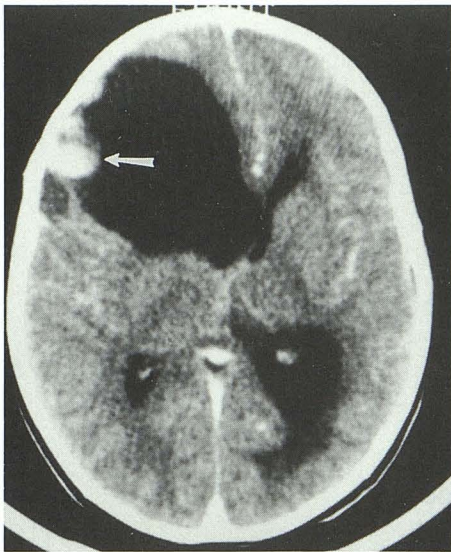


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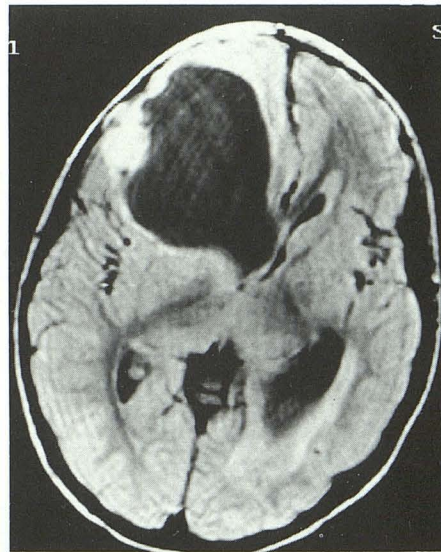
Fig. 1. Case 1.

A, Postcontrast CT scan shows the peripheral location of the enhancing portion of the mass (*straight arrow*) and its cystic component (*curved arrow*).

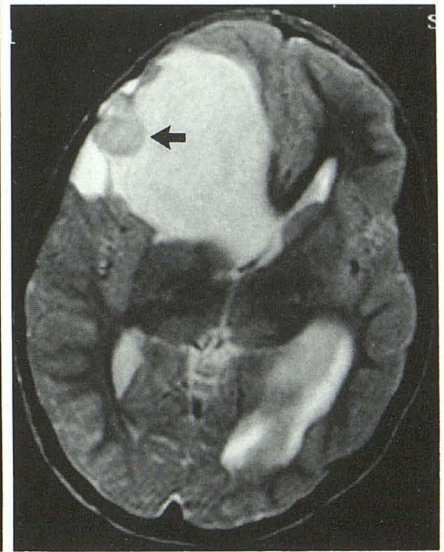
B, Selective right external carotid angiography demonstrates blood supply from the middle meningeal artery (*arrow*) and a faint "blush."



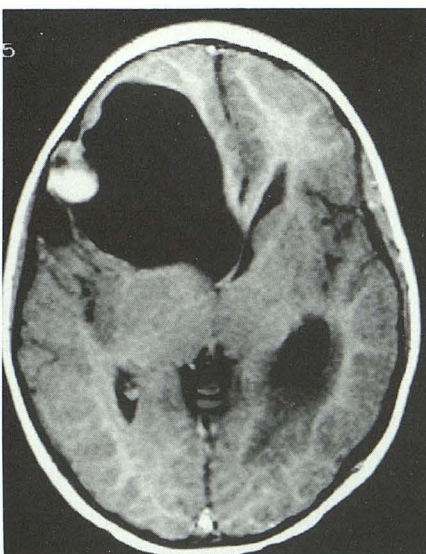
A



B



C



D

Fig. 2. Case 2.

A, Postcontrast CT scan demonstrates a large, predominantly cystic, right frontal lobe mass with peripherally located enhancing nodules (*arrow*).

B, Proton-density MR (2000/30/2) (TR/TE/excitations) also shows the nodular portion of the mass.

C, The nodular portion of the mass (*arrow*) is seen on this T2-weighted image (2000/120/2) in which the adjacent cystic portion of the mass has higher signal intensity.

D, T1-weighted (500/20/2) post-GD-DTPA image shows dense enhancement of the nodular portion of the mass.

Previously reported cases were peripherally located, with more than half being centered in the temporal lobes (3–12). Dural attachment was common at surgery. Half of those that had CT scans were cystic, but most had enhancing areas as well. Calcifications were rarely found. Both of our cases had typical findings on CT, being peripherally located, cystic masses with nodules of enhancement. As in previously reported cases, angiography in one of our patients showed his lesion to be hypovascular. In our patient, who underwent angiography, external carotid blood supply was demonstrated. This is unusual in glioblastoma and fungal/protozoan infection, meaning that less common elements in the differential diagnosis such as pleomorphic xanthoastrocytoma need to be considered. Additionally, although the angiographic findings in this one case cannot be used to extrapolate the general angiographic appearance of pleomorphic xanthoastrocytoma, they are not unexpected in light of the typically peripheral location of this tumor, its origin from subpial astrocytes, and frequent dural attachment.

The MR obtained in our second patient demonstrated the solid portion of the lesion to be of intermediate signal on T1WI and of increased signal intensity on proton density and T2WI, with marked post Gd-DTPA enhancement on T1WI. These signal intensities are nonspecific and the same as those expected for most other tumors. The clinical utility of the MR scan is twofold. First, multiplanar depiction of the lesion helps establish the peripheral origin of the tumor and, secondly, the cystic component of the mass is well visualized in orthogonal planes. This was useful information from the surgical standpoint because it facilitated entering the cyst first and decompressing it, which meant that there was less need for retraction of normal brain during subsequent excision of the solid portion of the lesion.

The differential diagnosis of a peripheral enhancing lesion in a child includes glioblastoma, ganglioglioma, gangliosarcoma, astrocytoma, meningioma, and meningiosarcoma. The latter two diagnoses are rare in children and, for that reason, as these two cases illustrate, pleomorphic xanthoastrocytoma needs to be considered in the differential diagnosis of a peripheral, cystic, enhancing lesion in a child, especially when there is external carotid blood supply to the tumor. This is important because aggressive surgery with attempt at complete removal is indicated given the favorable prognosis of these lesions.

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