Temporal pleomorphic xanthoastrocytoma with glycogen accumulation–case report.

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Pleomorphic xanthoastrocytoma (PXA) is a rare superficial glioma that predominates in the young and has good prognosis. A long history of repeated seizures is commonly associated with PXA, which is frequently observed in neuroimaging scans as a solid-cystic, contrast-enhancing lesion. We report a case in which PXA diagnosis was favored by its histological features, such as pleomorphic multinucleated giant cells, with disproportionately few mitoses and necrotic areas. An eye-catching feature was widespread, pale-staining, circumscribed deposits in the cytoplasm of tumor cells, which turned out to be glycogen upon histochemical and electron-microscopical examination. The stored material was strongly PAS-positive and digested by diastase, and had a finely granular ultrastructural appearance. No evidence of lipid droplets was found on oil-red-O staining. The tumor was immunoreactive for glial fibrillary acidic protein and vimentin. Many cells were positive for CD34 on the external membrane, a feature which has been described in chronic CNS lesions associated with epilepsy. Intracytoplasmic immunostaining for EGFR was observed in most tumor cells, which might have favored neoplastic proliferation. Nuclear immunolabeling for p53 protein was rare and does not support a major role for p53 mutation in PXA tumorigenesis. Intracellular accumulation of glycogen in glial tumors is uncommon and may originate from abnormalities in carbohydrate metabolic pathways.

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