CASE REPORT

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Pleomorphic xanthoastrocytoma as a component of a temporal lobe cystic ganglioglioma: a case report

Abstract We report a case of pleomorphic xanthoastrocytoma (PXA) as a component of a ganglioglioma in a 13-year-old Japanese boy. Magnetic resonance imaging showed a large cystic lesion with an enhanced mural nodule of the left temporal lobe. Microscopic examination of the tumor showed that it was composed of two distinct neoplastic components: dysplastic ganglion cells and a PXA. There were gradual transitions between the two neoplastic components, and the PXA constituted the gliomatous component of the ganglioglioma. The PXA component showed spindle-shaped and pleomorphic large cells with lipidized cytoplasm. The tumor cells were surrounded by numerous reticulin fibers. Immunohistochemically, the ganglion cells were negative for glial fibrillary acidic protein (GFAP), but showed positive staining for a 70-kDa neurofilament protein, synaptophysin, and NeuN. In contrast, PXA cells were positive for GFAP but negative for neuronal markers. Our case is therefore histologically classified as ganglioglioma with PXA as the glial component. These results suggested that PXA and ganglioglioma share a common origin and that the combination of PXA–ganglioglioma would be positioned along the spectrum between PXA and ganglioglioma. Alternatively, these results may support the hypothesis that PXA originates from glioneuronal progenitor cells capable of generating astrocytic and neuronal cell types.

Key words Pleomorphic xanthoastrocytoma · Ganglioglioma · Combined tumor · Glioneuronal tumor

Introduction

The pleomorphic xanthoastrocytoma (PXA) was first described by Kepes et al. in 1979. It is characterized as a distinctive meningocerebral glioma that occurs in young subjects and has a relatively favorable prognosis. Subsequently, many additional cases have been reported worldwide, and most patients have had a long, symptom-free postoperative survival. PXAs have also been shown to be immunoreactive for glial fibrillary acidic protein (GFAP), establishing their astrocytic lineage. However, over the past 20 years, there have been increasing reports of PXA with neuronal elements or composite PXA and ganglioglioma.

Recently, we treated a patient with a PXA as a component of a temporal lobe cystic ganglioglioma. In the present study, we discuss the clinicopathological features of this unusual composite PXA and ganglioglioma.

Materials and methods

Case report

A 13-year-old Japanese boy visited the Department of Neurosurgery of our hospital, complaining of head injury. The boy had no events during his peri- or postnatal periods and no previous neurological disease. He was right-handed and had no neurological deficits such as hemiparesis or visual abnormality. Magnetic resonance imaging (MRI) showed a large cystic lesion with a mural nodule of the left temporal lobe on T1-weighted images. On T1-weighted images with gadolinium enhancement, the mural nodule was markedly enhanced (Fig. 1A). On T2-weighted images, perifocal edema of the lesion was not apparent (Fig. 1B).

The patient was initially followed as an outpatient because he had no clinical symptoms. However, 1 month after the first medical examination, he showed an
acceleration of intracranial pressure, as evidenced by such symptoms as bilateral abducens nerve palsy, nausea, and vomiting. Accordingly, surgical removal of the tumor was scheduled.

At surgery, a yellowish tumor was found to be localized in the cystic wall as a mural nodule. An intraoperative frozen section showed neoplastic components of the tumor in the mural nodule but no neoplastic components in another cyst wall. Therefore, resection of the mural nodule was performed, except for the part of the tumor attached to the left middle cerebral artery and the other cyst wall. The postoperative course was uneventful. The patient showed no evidence of tumor recurrence in the 7-month follow-up period.

Histological and immunohistochemical studies

Tissue samples were fixed in 10% buffered formalin, embedded in paraffin, and processed conventionally for histology and immunohistochemistry. Sections (5 μm thick) were stained using hematoxylin and eosin (H&E) for histological evaluation. The remaining serial unstained sections were used for immunohistochemistry. Immunohistochemical studies were performed using peroxidase avidin–biotin methods (LSAB kit; DakoCytomation, Carpinteria, CA, USA) on paraffin sections following heat-induced antigen retrieval. Primary antibodies were directed toward glial fibrillary acidic protein (GFAP, prediluted; DakoCytomation), Neu N (dilution 1:50; DakoCytomation), synaptophysin (dilution 1:50; DakoCytomation), neurofilament protein (2F11, dilution 1:10; DakoCytomation), and MIB-1 (dilution 1:100; Immunotech, Marseille, France). The MIB-1 labeling index was determined by counting 1000 cells at 400× magnification in three different fields in the region of highest density of positively stained cells.

Results

Histological findings

Microscopic examination of the tumor showed moderate cellularity but great variation in the size and shape of the tumor cells with desmoplasia. Lymphocytic infiltration of the perivascular space was present at the periphery of the tumor (Fig. 2A). The dysplastic neurons showed large round nucleated cells with vesicular nuclei and prominent nucleoli (Fig. 2B). The high-power view showed binucleated neuronal cells (Fig. 3A). The tumor consisted of neuronal cells and astrocytic cells which, in several areas, blended with each other to create a transitional appearance. The tumor also contained nuclear pseudoinclusions in the spindle astrocytic components and numerous eosinophilic granular bodies (Fig. 3B,D). Pleomorphic astrocytic cells showed multiple lipid droplets within the cytoplasm (Fig. 3C,D).

No mitosis or necrosis was identified. Reticulin staining in the PXA-predominant area demonstrated staining around individual tumor cells (Fig. 4A). Immunohistochemically, the pleomorphic xanthic cells showed cytoplasmic immunoactivity with GFAP (Fig. 4B), whereas dysplastic ganglion cells were negative for GFAP (Fig. 4B). In contrast, the ganglionic tumor cells showed immunoreactivity with synaptophysin, neurofilaments (Fig. 4C), and Neu-N (Fig. 4D). The tumor showed a comparatively low MIB-1 labeling index (2%).

Discussion

PXAs are characterized as astrocytic neoplasms with relatively favorable prognosis, and are typically encountered in children and young adults, with superficial location in the cerebral hemispheres and involvement of the meninges.