Pleomorphic Xanthoastrocytoma Case Reports

2025

A 49-year-old woman with a mass in the left frontotemporal region. Microscopically, this mass is composed of the glial and rhabdoid elements, both of which have molecular features of PXA, and the rhabdoid elements assessed using immunohistochemistry for SMARCB1 (INI1) expression demonstrated expression loss. The DNA methylation profile showed a significant calibrated score of 0.81 for methylation class PXA. The tumor was eventually diagnosed as a PXA with SMARCB1 deficiency [1].

An 18-year-old left-handed African American male presented with a year-long history of seizures characterized by episodic palpitations, sweating, and agitation. Brain magnetic resonance imaging revealed an enhancing tumor in the right anterior entorhinal cortex with adjacent amygdalar enlargement. Interictal magnetoencephalography and video-electroencephalogram confirmed lesional right temporal lobe epilepsy. The patient underwent a partial right anterior temporal lobectomy, with histopathology revealing WHO Grade 2 pleomorphic xanthoastrocytoma with a BRAF V600E mutation. The amygdala showed no tumor infiltration, confirming reactive hyperplasia rather than neoplastic involvement. This case underscores the importance of distinguishing tumor infiltration from benign seizure-related amygdalar enlargement in long-term epilepsy-associated tumors, usefully informing surgical strategy [2]

A 10-year-old boy with clinical manifestations of recurrent epileptic seizures. Preoperative standardized antiepileptic drug treatment failed to control seizures. The patient's electroencephalogram (EEG) showed was the right temporal lobe is the main slow wave and spike slow wave emitting area, and magnetic resonance imaging (MRI) showed structural abnormalities in the right anterior temporal cortex. After multidisciplinary preoperative evaluation at the epilepsy center of Tianjin Children's Hospital, lesion enlargement resection was performed with the assistance of multimodal imaging and electrocorticography (ECoG) monitoring. There were no epileptic seizures during the 6-month follow-up after surgery.

For this patient with PXA accompanied by epilepsy, surgical resection can be the first line of treatment. Meanwhile, a comprehensive multidisciplinary preoperative evaluation should be conducted rather than solely relying on neurosurgery to determine surgical treatment. Additionally, imaging and intraoperative ECoG are crucial for the success of surgery, and appropriate enlargement and resection can effectively eliminate epileptic seizures [3].

2024

a 9-year-old girl with anaplastic PXA treated with PBT following incomplete surgical resection. A total

dose of 60 Gy (RBE) in 15 fractions was administered, leading to significant tumor reduction, no progression, and improved local control at the 1-year follow-up, with no observed adverse effects. Based on short-term follow-up results, our study highlights the potential of PBT in managing anaplastic PXA, demonstrating favorable local outcomes and a low incidence of radiation-induced complications. While long-term follow-up and evaluation are necessary to further support these findings, this case represents only the second reported instance of anaplastic PXA treated with PBT, contributing to the growing body of evidence supporting its efficacy in this rare tumor type [4].

A case of recurrent BRAF-mutant pleomorphic xanthoastrocytoma (central nervous system World Health Organization grade 3) treated with combination therapy with BRAF and MEK inhibitor. The patient received dabrafenib (BRAF inhibitor) and trametinib (MEK inhibitor); however, she developed resistance to the combination therapy. Remarkably, incidental drug discontinuation contributed to the disappearance of the resistant tumor. The same phenomenon was repeatedly observed after that. Genetic analysis demonstrated that the resistant tumor had BRAF V600E amplification; the resistant tumor remained BRAF→MEK→ERK pathway dependent, and drug resistance might be due to elevated BRAF V600E expression. We speculated that ERK1/2 signal extremes caused by the discontinuation of the combination therapy affected the resistant tumor survival.

This case study provides important insights into novel treatment strategies and their underlying mechanisms for gliomas with BRAF mutations [5]

2022

A 30-year-old woman with a previous history of unconfirmed resected lateral ventricle meningioma presented with severe headache for 1 day. Imaging examination revealed a mass in the right lateral ventricle with heterogeneous signal patterns, changes in the posterior fossa corresponding to a Dandy-Walker variant, and mild hydrocephalus.

Surgical complete resection of the mass was achieved. postoperative histopathological examination confirmed WHO grade II pleomorphic xanthoastrocytoma. Three years postsurgery, ventriculoperitoneal shunt was performed due to worsening of hydrocephalus. The patient has since remained symptom-free.

This is the first report of the concomitant occurrence of Dandy-Walker complex and pleomorphic xanthoastrocytoma. The association of neurological congenital malformation with intracranial neoplasms may be multifactorial, with underlying role of genetic mutations or chromosome alterations [6].

Finch et al., presented the case of a 16-year-old male who responded to vemurafenib monotherapy initially and had an additional response to vemurafenib following progression after a brief time off the medication [7].

2016

A case of PXA that was initially diagnosed as GBM is presented. A 42-year-old man visited the clinic because of right hemiparesis and total aphasia. Head magnetic resonance imaging demonstrated enhanced multiple cystic lesions in the left temporal lobe suggesting an intra-parenchymal brain tumor. The lesion was partially removed and GBM with a Ki-67 index of 20 % was diagnosed by pathological examination of the resected specimen. Despite receiving radiation and chemotherapy, the patient died 6 months after the first admission. At autopsy, the boundary between the tumor and normal brain tissue was clear. Large parts of the tumor demonstrated typical features of PXA, including pleomorphism, clear xanthomatous cells with foamy cytoplasm, positive silver staining, and a Ki-67 index of less than 1 %.

GBM should be diagnosed only when the majority of the tumor cells are undifferentiated. Although the operative specimen appeared typical GBM histologically, the diagnosis of GBM was subsequently excluded by the autopsy finding that much of the tumor had the characteristic features of a benign PXA. Therefore, the final diagnosis in this case was PXA with anaplastic features. PXA with anaplastic features should be carefully distinguished from GBM to facilitate appropriate decisions concerning treatment [8)].

Combined pleomorphic xanthoastrocytoma (PXA) and ganglioglioma (GG) is an extremely rare tumor, with fewer than 20 cases reported. Cicuendez et al report a case of combined PXA-GG in an 18-year-old man with a history of seizures. The tumor showed necrosis and the BRAF V600E mutation on histological examination, with no evidence of tumor recurrence 1 year after gross-total resection. The BRAF V600E mutation was present, which suggests that both cell lineages may share a common cellular origin [9)].

Yamada et al report the case of a 56-year-old man presenting with seizures and headaches. Magnetic resonance imaging revealed a large right temporal lobe mass with low T1 and high T2/FLAIR signal and a discrete contrast-enhancing focus. Histologically, the tumor showed 2 distinct components: an infiltrating astrocytoma harboring 5 mitoses/10 high-power fields and a relatively circumscribed focus, resembling PXA with, at most, 2 mitoses/10 high-power fields. No microvascular proliferation or necrosis was present in either component. The infiltrating astrocytoma component contained numerous axons, whereas the PXA-like component had sparse axons, as demonstrated by the neurofilament immunostain. Both components were positive for the mutant IDH1 R132H and showed loss of ATRX expression, whereas BRAF V600E was restricted to the PXA-like component. On sequencing of the 2 components separately after microdissection, both showed identical IDH1 R132H and TP53 R273C point mutations, whereas the BRAF V600E mutation was limited to the PXA-like component. These findings are consistent with clonal expansion of a morphologically distinct focus, harboring a private BRAF V600E mutation within an IDH1-mutant glioma. Intratumoral heterogeneity and clonal evolution, as seems to have occurred here, suggest reevaluation of “collision tumors” as a concept [10)].

2015

Cases of cerebellar PXA are rare, and those associated with neurofibromatosis type 1 (NF1) are even

less common, with only 2 cases reported to date. Takei et al present a third case of PXA-NF1 with unusual features. A 33-year-old woman presented with a history of headache. Her medical and family history was significant for NF1. Brain MRI revealed a 3.4 cm ill-defined lesion with a gyriform enhancing pattern in the left cerebellum, superficially mimicking Lhermitte-Duclos disease. The patient underwent a gross total resection of the lesion and had an unremarkable postoperative course. While the lesion had histological features typical of “pure” PXA (WHO grade II) it had an unusual growth pattern with thickening of the superficial cerebellar folia and predominant leptomeningeal involvement. No BRAF, IDH-1, or IDH-2 mutation was identified. Three months after surgery, local recurrence was detected, and the patient was treated with radiation therapy. One year after the first surgery, she underwent surgical resection of the recurrent/residual tumor. Histologically, the recurrent tumor showed very similar features to the initially resected tumor, with no anaplastic features. Most cerebellar PXAs have an indolent clinical behavior as do most cerebral PXAs. Whether co-existence of NF1 was a factor in altering the clinical course and biologic behavior of this patient's tumor is currently unknown [11)].

2012

An illustrative case of a PXA transforming to glioblastoma multiforme is presented [12)].

2010

A 16-year-old female, with unremarkable medical and family history, presented with a huge dural-based mass in the right frontotemporal fossae, manifesting as headache. The patient underwent subtotal tumor resection. Intraoperative findings revealed focal erosion in the temporal fossa dura mater and skull adjacent to the lesion. Most of the tumor was located extraaxially, but a part of the tumor had invaded the temporal lobe, and had tightly adhered to the middle cerebral artery and its perforating vessels. Histological examination revealed cellular pleomorphism with mitotic activity, focal necrosis, but lacking endothelial proliferation, consistent with anaplastic pleomorphic xanthoastrocytoma (PXA) with component of anaplastic astrocytoma. Postoperatively, the patient underwent local irradiation and temozolomide administration, but the tumor relapsed 13 months later. Second tumor resection was performed followed by gamma knife radiosurgery, but the residual tumor progressively grew, extending into the contralateral hemisphere, and formed an enormous mass in the left frontal lobe at 17 months. Magnetic resonance imaging performed at 18 months revealed extracranial infiltration of the frontal tumor, through the cribriform plate, with enormous extension into the paranasal sinuses, nasal cavity, and orbit during the next month. The patient died at 20 months after the initial surgery. PXA with anaplastic appearance may have a component of anaplastic astrocytoma with more aggressive behavior [13)].

2009

A 28-year-old man presented with a short history of headache, visual and aphasic speech disturbances. MR scans revealed a large, partly cystic, contrast-enhancing lesion of the left temporal lobe that upon microscopic examination was diagnosed as pleomorphic xanthoastrocytoma (PXA) with anaplastic features (WHO grade III). Remarkably, this tumor featured an unusual gliovascular, rosette-like histoarchitecture, which had previously been hypothesized to possibly indicate a greater

likelihood of PXA recurrence. Indeed, only 14 months later, the patient presented with a recurrent lesion, which contained the previous histology, but now also featured a distinct fibrosarcoma-like component replete with numerous osteoclast-type giant cells. In addition, whereas eosinophilic granular bodies were plentiful at the lesion's periphery, numerous CD34 - positive satellite cells were found in the adjacent non-infiltrated cortex. Regarding the origin of this recurrent tumor and in reflection of its composition of distinct PXA as well as sarcomatous components, the diagnosis of a pleomorphic xanthoastrosarcoma, to be conceptually considered as a gliosarcoma subtype, was made.To our knowledge, this is an unprecedented case of sarcomatous transformation of a PXA. Particular attention should be given to gliovascular pseudopapillary structures in PXAs, the presence of which may potentially herald a more aggressive clinical behavior [14)].

2006

A 59-year-old woman who presented with tumor bleeding onset and cerebrospinal fluid dissemination. The patient had sudden-onset right hemiparesis, aphasia, and consciousness disturbance and was admitted to a local area hospital. After emergency surgery had removed the hematoma, postoperative contrast-enhanced CT scan revealed a left temporal tumor. A second surgery was therefore performed for initial tumor removal 2 months later. Histopathological findings showed that the tumor was typical PXA with strong pleomorphism and xanthomatous changes and contained an ependymoma-like component in the center area. However, endothelial proliferation and mitosis were more remarkable compared to ordinary PXA. The MIB-1 labeling index was 9.8% high. From these findings, the histopathological diagnosis was anaplastic PXA. The patient underwent surgery to remove recurrent tumors 5 and 16 months later. The patient died 36 months after the first onset, and CT revealed glioblastoma-like findings and cerebrospinal fluid dissemination. This case report is the first case in which PXA presented with tumor bleeding onset. Histopathological findings suggested anaplastic PXA from the first surgical specimens, and PXA recurred many times. We thus believe that the patient displayed primary anaplastic PXA rather than secondary anaplastic PXA that results in malignant transformation [15)].

2003

Lipidized glioblastoma multiformis (LGB) and pleomorphic xanthoastrocytoma (PXA) are often supratentorial in location and occur in the second to fourth decade. This report presents two young patients, one having LGB and the other having PXA in the cerebellum. Histological differentiation between LGB and PXA is discussed [16)].

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