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Unusual location of pleomorphic xanthoastrocytoma: a case report

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Abstract

Background Pleomorphic xanthoastrocytoma is a rare astrocytic tumor often diagnosed at a young age. Typically, they appear as supratentorial cortical tumors, frequently involving the temporal lobe with few reported rare locations. The prognosis is favorable following surgical excision; however, recurrence, dissemination, and anaplastic transformation occurred in some cases.

Case presentation A 50-year-old female presented with convulsions and an altered consciousness. Imaging showed a periventricular mixed solid and cystic lesion. Histopathological examination revealed features of pleomorphic xanthoastrocytoma WHO grade 2 without necrosis or mitotic activity.

Conclusions This report highlights the classic imaging findings of pleomorphic xanthoastrocytoma but in an atypical periventricular location. Although rare, pleomorphic xanthoastrocytoma should be considered in the differential diagnosis of mixed solid and cystic periventricular lesions.

Keywords Pleomorphic xanthoastrocytoma, Periventricular, Magnetic resonance imaging, Neurosurgery, Case report

Background

Pleomorphic xanthoastrocytoma (PXA) is a rare astrocytic tumor that originates from subpial astrocytes and comprises less than 1% of astrocytomas. It was first described by Kepes and colleagues in 1979 [1]. It is included in the World Health Organization (WHO) classification of central nervous system tumors under the group of circumscribed astrocytic gliomas and can be grade 2 or 3 depending on the mitotic counts [2]. Histologically, as the name implies, it is characterized by pronounced cellular pleomorphism, cell lipidization, perivascular inflammatory cells, and an abundant reticulin network. Tumor cells are either spindle cells with elongated nuclei or giant round cells with multilobulated nucleus, single nucleus, or multiple nuclei. Necrosis and mitosis are rare [3].

Commonly, PXA is diagnosed in children and young adults without a gender predilection. They are typically supratentorial tumors with the temporal lobe being the most frequent location, followed by the frontal and parietal lobes. Characteristically, they are superficial cortical-based tumors, and hence longstanding epilepsy is the most frequent clinical presentation [1, 3]. Following gross total resection, the prognosis is usually favorable, with a 5-year survival rate up to 81% and a 10-year survival rate up to 70%. There are several factors that are associated with poor prognosis including leptomeningeal spread, anaplastic features, subtotal resection, and advanced age [3–5].

It has been reported that 70% of these tumors appear on imaging as cystic masses with an enhancing mural nodule and 30% are solid masses that may show internal cystic changes. Peritumoral edema is usually minimal.

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On computed tomography (CT), the density of the solid component is variable and usually iso- or hypodense and rarely hyperdense. Calcifications can be seen but they are uncommon. On magnetic resonance imaging (MRI), the cystic component shows signal intensity similar to cerebrospinal fluid (CSF). The solid component is typically iso- or slightly hyperintense compared to the cortex on T1-weighted images, and slightly hyperintense on T2-weighted images. Heterogenous enhancement of the solid component and peripheral enhancement of the cyst wall is generally seen [1, 3].

Rare cases of PXA have been reported to have occurred sporadically in the suprasellar region, pineal region, cerebellum, and spine [4, 6–8]. In a few publications, this rare tumor has been described in rare intraventricular and periventricular locations [9–17]. We report a case of PXA in an atypical periventricular location.

Case presentation

A 50-year-old previously healthy female presented to the emergency department with seizures and an altered consciousness level. She was not known to have epilepsy. On clinical examination, she was afebrile with normal and stable vital signs. Neurologic examination revealed left hemianopsia. The other cranial nerves were intact. Sensory, motor, and cerebellar exams were normal. Respiratory, cardiovascular, and abdominal examinations revealed no abnormalities. Routine laboratory tests including complete blood count, random blood sugar, renal function test, and liver function test were all within normal limits.

She was referred to our radiology department for non-enhanced computed tomography (NECT) to exclude acute hemorrhage. Non-enhanced axial cuts were obtained through the head with 5 mm slice thickness and 0.5 mm thin reconstructions using a Canon Aquilion PRIME SP scanner. A right temporal-parietal periventricular isodense mass was identified. It was associated with cystic changes and a dominant large cyst. No frank calcifications or gross intratumoral hemorrhage were seen. Mild supratentorial ventricular dilatation was also noted without periventricular interstitial edema (Fig. 1).

Then, MRI was done for lesion characterization. Multiple sequences on multiple planes were performed through the head pre- and post-intravenous gadolinium administration using a Philips Ingenia 3.0 T MRI machine. The solid component was deep, centered in the periventricular region, and protruded into the right lateral ventricle. The cystic component was peripheral and reached the subcortical region (Fig. 2).

The patient was prepared and taken electively to the operating room for transcortical gross total tumor resection via right parieto-occipital craniotomy. Histopathological examination revealed PXA WHO grade 2 features without necrosis or mitotic activity (Fig. 3). Postoperatively, the patient's neurological symptoms have improved apart from persistent left hemianopsia.

One-year postoperative MRI demonstrated the expected old postoperative changes and focal dilatation of the temporal horn of the right lateral ventricle without periventricular interstitial edema. No features of residual or recurring tumors were identified (Fig. 4). The patient has been followed now for two years, and she is in good condition. Antiepileptic medication is prescribed once daily, and no seizures have been reported. Another follow-up MRI two years after surgery showed reduced temporal horn dilatation, unchanged old postoperative changes, and no evidence of recurrence.

Discussion

Classicly, PXA is a supratentorial cortical-based astrocytic tumor [1, 3]. The intraventricular and periventricular locations of PXA are extremely rare. To date, only nine cases of intraventricular and periventricular PXAs (grades 2 and 3) have been found described in the English literature, and only two of these cases are located in the periventricular region (Table 1) [9–17]. We report a rare atypical periventricular location of PXA.

Preoperative diagnosis of PXA is generally suggested on imaging if a cortical-based complex solid and cystic lesion is discovered in a young patient with a history of longstanding epilepsy [1, 3]. In our case, the periventricular lesion's location and the patient's age were atypical. Therefore, the preoperative differential diagnosis was wide and included ependymoma, PXA, and ganglioglioma, yet, these tumors commonly occur in young adults. An atypical appearance of metastasis and diffuse glioma were also included as they are more common in older adults [18].

In light of previously reported cases of intraventricular and periventricular PXA, it is possible to postulate that their clinical presentation is more variable and diverse. Including our case, there were three cases of females and seven cases of males among different age groups. It is, however, difficult to conclude whether there is an age or gender predilection based on the few PXA cases described in the literature in these rare locations. Similar to our case, five of the reported cases were grade 2 [10, 12–14, 17], while four were grade 3 PXAs [9, 11, 15, 16]. An association with neurofibromatosis 1 was reported with one case of periventricular PXA grade 2 [12].

It is important to note that despite being in an atypical location, our case and eight reported intraventricular and periventricular PXAs demonstrated the classic imaging pattern of PXA, a solid mass with a cystic component

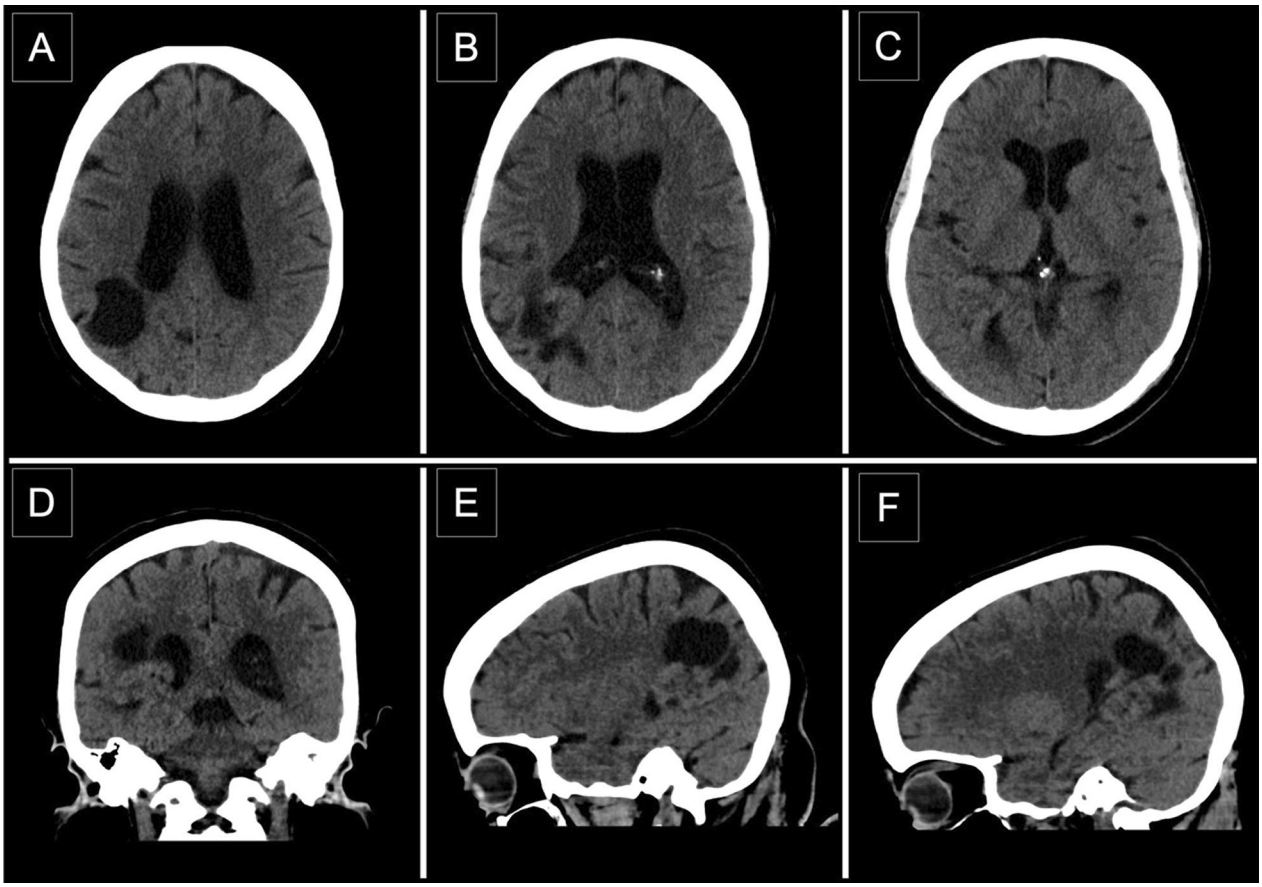

Fig. 1 Pre-operative NECT Brain. Axial (**A**–**B**–**C**) cuts and coronal (**D**) and sagittal (**E**–**F**) reformates show right parietotemporal periventricular complex mass. The solid component is isodense to the grey matter without frank calcifications or gross intratumoral hemorrhage. The cystic component shows a density similar to CSF. Mild supratentorial ventricular dilatation is also noted without periventricular interstitial edema

[9–16]. Similar to our case, Liu et al. [14] described an intraventricular PXA grade 2 that has a dominant cyst. On the contrary, Menéndez et al. [17] reported an intraventricular PXA grade 2 within the third ventricle that appears as homogenously enhancing solid mass without cystic changes.

In our case, the solid component was isodense on NECT in contrast to three of the reported intraventricular and periventricular PXAs that had NECT and all were hyperdense [11, 12, 17].

Bettencourt et al. [11] described an intraventricular PXA grade 3 associated with calcifications and intratumoral hemorrhage.

On MRI, the solid component in our case showed an isointense signal on T1-weighted images, a slightly hyperintense signal on T2-weighted images, and a heterogenous enhancement similar to what has been described in the literature [9, 10, 13–16]. However, the enhancement appears to be less intense in our case. These findings support that PXA in intraventricular and periventricular locations generally has imaging characteristics similar to PXA in typical locations. It should be noted, however, that they have a higher risk of obstructive hydrocephalus [9, 11, 12, 15, 16].

Comparing the follow-up results of intraventricular and periventricular PXA with what has been found in the typical locations of PXA, it is possible to suggest that the predictors of poor prognosis are similar including anaplastic features and leptomeningeal dissemination. There is, however, a need for further investigation with a larger group of patients and longer follow-up periods [3–5, 9–17].

Conclusions

This report illustrates the classical imaging findings associated with PXA but at an atypical periventricular location. It is noteworthy that although this is a rare location, our case highlights the importance of including PXA in the differential diagnosis of intraventricular and periventricular complex solid and cystic masses.

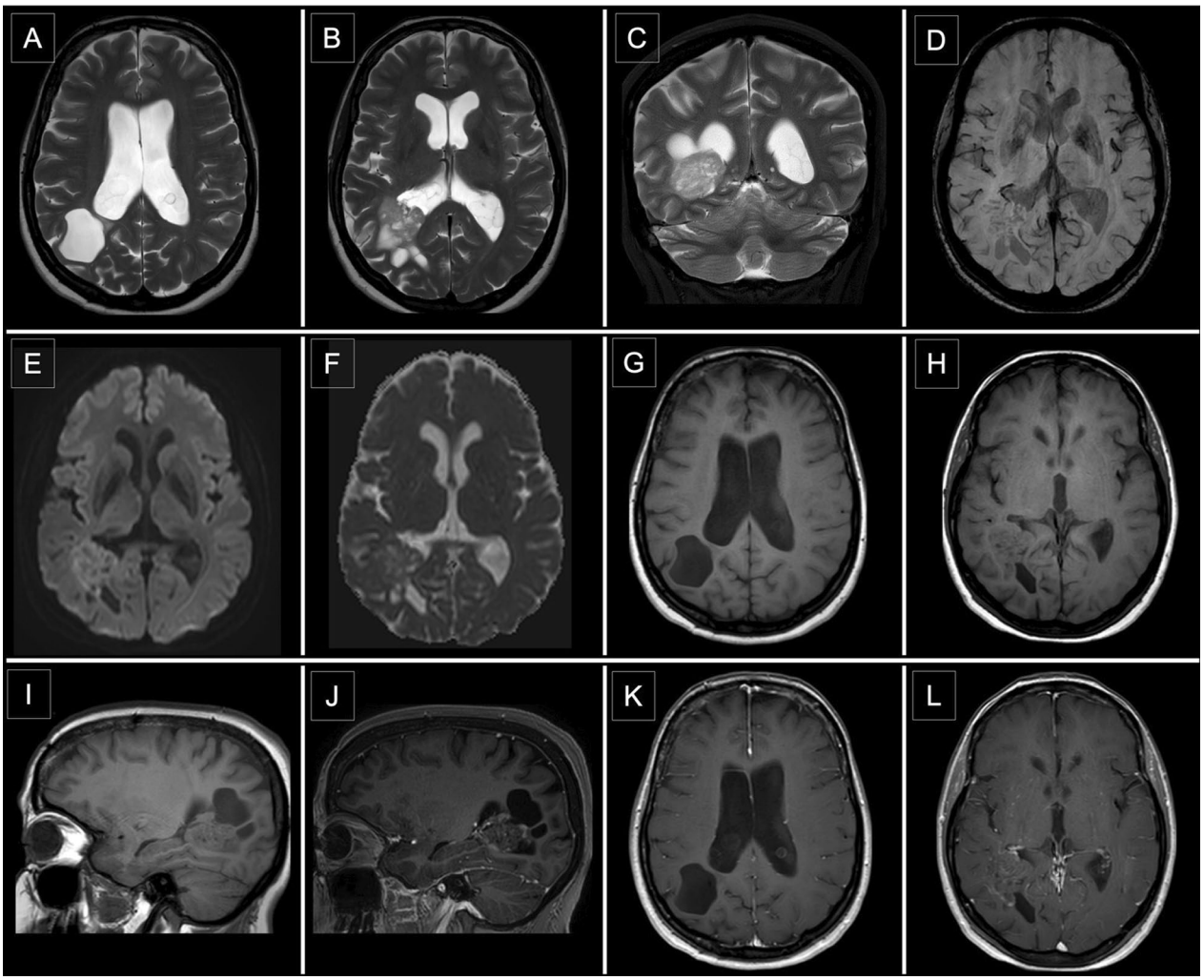

Fig. 2 Pre-operative MRI brain. Axial (**A**–**B**) and coronal (**C**) T2-weighted images show the right parietotemporal periventricular complex mass. Its solid component is centered at the periventricular region, protrudes into the ventricle, and shows a slightly hyperintense signal. The cystic component is peripheral reaching the subcortical region and shows a signal similar to CSF. Axial susceptibility-weighted image (**D**) shows no significant hemorrhage or calcification. Axial diffusion-weighted image (**E**) and apparent diffusion coefficient map (**F**) demonstrate no diffusion restriction. Axial (**G**–**H**) and sagittal (**I**) T1-weighted images show an isointense solid component and CSF signal intensity of the cystic component. Sagittal (**K**) and axial (**K**–**L**) T1-weighted images post-contrast show mild heterogenous enhancement of solid component and non-enhancing cystic component

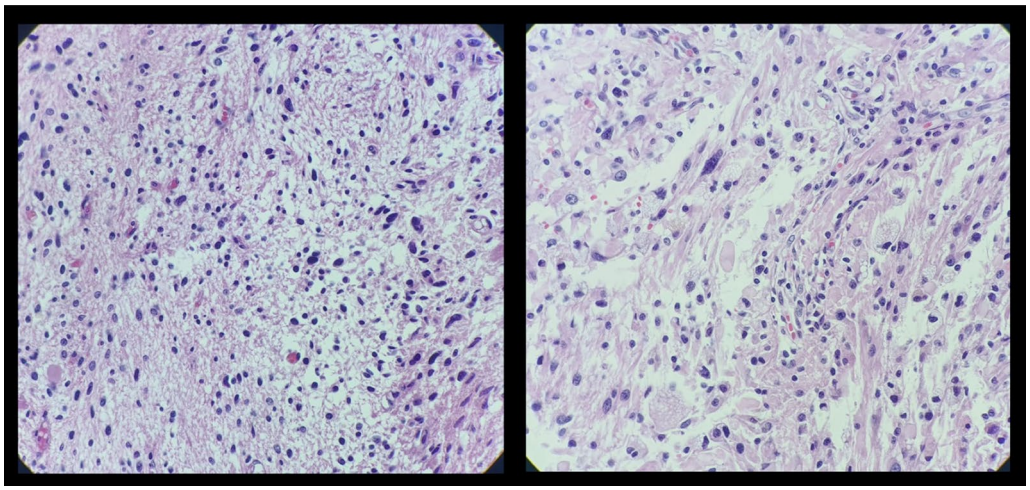

Fig. 3 Pathology Slides. Hematoxylin and eosin-stained sections show pleomorphic spindle cell astrocytes with lipidized cells and eosinophilic granular bodies

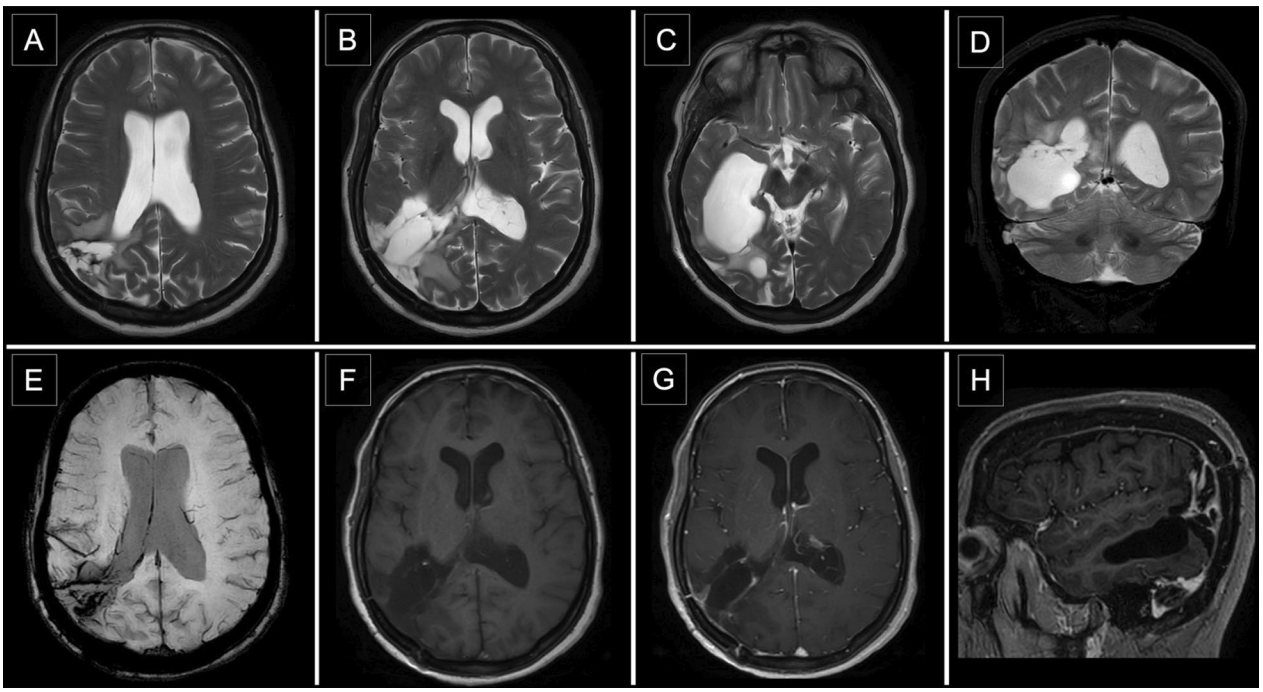

Fig. 4 Postoperative MRI brain. Axial (**A**–**B**–**C**) and coronal (**D**) T2-weighted images show a right parietotemporal surgical cavity extending from the ventricular wall to the cortex with surrounding hyperintensity representing gliosis and dark T2-signal related to postoperative hemosiderin deposition. Dilatation of the temporal horn of the right lateral ventricle is noted as well. Axial susceptibility-weighted image (**E**) shows a blooming artifact at the surgical region related to hemosiderin deposition. Axial T1-weighted image pre-contrast (**F**) and axial (**G**) and sagittal T1-weighted images post-contrast show meningeal enhancement and enhancing bands at the surgical site indicating granulation tissue. There is no evidence of residual tumor or recurrence

Table 1 Summary of intraventricular and periventricular PXAs reported in the English literature

Author/year of publication	Age and gender	Associated neurological conditions	Presentation	Location	Imaging pattern	Grade	Treatment	Follow-up
Wu et al. [15]	45-year-old male	Cerebral palsy, Corpus callosum agenesis	Difficulty walking and altered consciousness	Intraventricular; third ventricle	Solid mass with cystic changes	3	Subtotal resection, radiotherapy and chemotherapy	1.5 years; No lesion growth
Bettencourt et al. [11]	33-year-old male	None	Dizziness and memory impairment	Intraventricular; right lateral ventricle	Solid mass with cystic changes, hemorrhage and calcification	3	Gross total resection, radiotherapy and chemotherapy	7 months; Recurrence
Liu et al. [14]	28-year-old male	None	Incidental after minor traffic accident	Intraventricular; left lateral ventricle	Solid mass with cystic changes and dominant cyst	2	Total Gross total resection	8 months; No recurrence
Roberti et al. [16]	65-year-old female	None	Memory impairment, urinary incontinence and ataxia	Intraventricular; left lateral ventricle	Solid mass with cystic changes	3	Gross total resection	3 months; Recurrence
Menéndez et al. [17]	24-year-old male	None	Progressive headache, hypersomnia and behavioral change	Intraventricular; third ventricle	Solid mass	2	Gross total resection	48 months; No recurrence
Adeleye et al. [12]	10-year-old male	Neurofibromatosis 1	Progressive painless right-sided visual impairment	Periventricular; left thalamic and lateral ventricle	Solid mass with cystic changes	2	Gross total resection	10 months; No recurrence
Yang et al. [10]	42-year-old female	None	Headaches and poor concentration	Intraventricular; right lateral ventricle	Solid mass with cystic changes and extensive subarachnoid dissemination	2	Subtotal resection	Died three days after surgery
Gonçalves et al. [13]	15-year-old male	None	Seizures	Periventricular; left temporo-occipital	Solid mass with cystic changes	2	Gross total resection	5 years; Recurrence 10 years; Free of disease
Fu et al. [9]	52-year-old male	None	Headache and forgetfulness	Intraventricular; right lateral ventricle	Solid mass with cystic changes	3	Subtotal resection, radiotherapy and chemotherapy	6 months; No recurrence

Abbreviations

PXA	Pleomorphic xanthoastrocytoma
WHO	World Health Organization
CT	Computed tomography
MRI	Magnetic resonance imaging
CSF	Cerebrospinal fluid
NECT	Non-enhanced computed tomography

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STA and MA made a substantial contribution to writing the manuscript. STA, MA, and AbA contributed to preparing the radiological figures. AhA wrote the pathology part and prepared the pathology figure. AAA performed the surgery and contributed to writing the case presentation. AbA wrote the imaging technical details. All authors contributed to data collection, reviewed the final manuscript, and approved it.

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References

1. Shaikh N, Brahmbhatt N, Kruser TJ et al (2019) Pleomorphic xanthoastrocytoma: a brief review. CNS Oncol 8:CNS39. https://doi.org/10.2217/cns-2019-0009
2. Torp SH, Solheim O, Skjulsvik AJ (2022) The WHO 2021 classification of central nervous system tumours: a practical update on what neurosurgeons need to know: a minireview. Acta Neurochir (Wien) 164:2453–2464. https://doi.org/10.1007/s00701-022-05301-y
3. Mahajan S, Dandapath I, Garg A et al (2022) The evolution of pleomorphic xanthoastrocytoma: from genesis to molecular alterations and mimics. Lab Invest 102:670–681. https://doi.org/10.1038/s41374-021-00708-0
4. Tan D, Lai LT, Daly CD et al (2020) Spinal pleomorphic xanthoastrocytoma: case report and literature review. World Neurosurg 141:25–32. https://doi.org/10.1016/j.wneu.2020.05.117
5. Mallick S, Benson R, Melgandi W et al (2018) Grade II Pleomorphic Xanthoastrocytoma; a meta-analysis of data from previously reported 167 cases. J Clin Neurosci 54:57–62. https://doi.org/10.1016/j.jocn.2018.05.003
6. Telemi E, Martirosyan NL, Avila MJ et al (2019) Suprasellar pleomorphic xanthoastrocytoma: a case report. Surg Neurol Int 10:1–5. https://doi.org/10.25259/SNI-83-2019
7. Hanna JA, Mathkour M, Gouveia EE et al (2020) Pleomorphic xanthoastrocytoma of the pineal region in a pediatric patient with neurofibromatosis type 1. Ochsner J 20:226–231. https://doi.org/10.31486/toj.18.0156
8. Kim SH, Hwang K, Lee KS et al (2020) Cerebellar pleomorphic xanthoastrocytoma with BRAF V600E mutation. World Neurosurg 139:577–581. https://doi.org/10.1016/j.wneu.2020.04.113
9. Fu YJ, Miyahara H, Uzuka T et al (2010) Intraventricular pleomorphic xanthoastrocytoma with anaplastic features. Neuropathology 30:443–448. https://doi.org/10.1111/j.1440-1789.2009.01080.x
10. Yang WQ, Huang B, Liang CH (2012) Pleomorphic xanthoastrocytoma in the lateral ventricle with extensive subarachnoid dissemination: report of a case and review of the literature. Chin Med J (Engl) 125:396–399. https://doi.org/10.3760/cma.j.issn.0366-6999.2012.02.042
11. Bettencourt S, Almeida G, Maia T (2023) A rare case report of intraventricular anaplastic pleomorphic xanthoastrocytoma. Cureus 15:e35975. https://doi.org/10.7759/cureus.35975
12. Adeleye AO, Okolo CA, Akang EE, Adesina AM (2012) Cerebral pleomorphic xanthoastrocytoma associated with NF1: an updated review with a rare atypical case from Africa. Neurosurg Rev 35:313–319. https://doi.org/10.1007/s10143-011-0362-1
13. Gonçalves VT, Fabiano R, Queiroz LDS, Zanardi VDA (2011) Periventricular pleomorphic xanthoastrocytoma (PXA): an uncommon tumor at an atypical site. Arq Neuropsiquiatr 69:570–570. https://doi.org/10.1590/S0004-282X2011000400034
14. Liu X-F, Du X, Zhang X-T et al (2019) Pleomorphic xanthoastrocytoma inside lateral ventricle: a rare case report and literature review. Int J Clin Exp Pathol 12:1118–1123
15. Wu X, Yokoyama K, Sumita K et al (2024) Intraventricular pleomorphic xanthoastrocytoma: a case report and systemic review. Cureus 16(1):e52510. https://doi.org/10.7759/cureus.52510
16. Roberti F, Baggenstos M (2018) Intraventricular anaplastic pleomorphic xanthoastrocytoma: very rare localization and early recurrence of a rare tumor. Cureus 10(5):e2665. https://doi.org/10.7759/cureus.2665
17. Menéndez R, Fernández J, Monti A, Sevlever G (2014) Intraventricular pleomorphic xanthoastrocytoma: a case report. Turk Neurosurg 24:987–991. https://doi.org/10.5137/1019-5149.JTN.10063-13.1
18. Smirniotopoulos JG, Jäger HR (2020) Differential diagnosis of intracranial masses. In: Hodler J, Kubik-Huch RA, von Schulthess GK (eds) Diseases of the brain, head and neck, spine 2020–2023: diagnostic imaging. Cham (CH): Springer; 2020. Chapter 8. https://doi.org/10.1007/978-3-030-38490-6_8

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