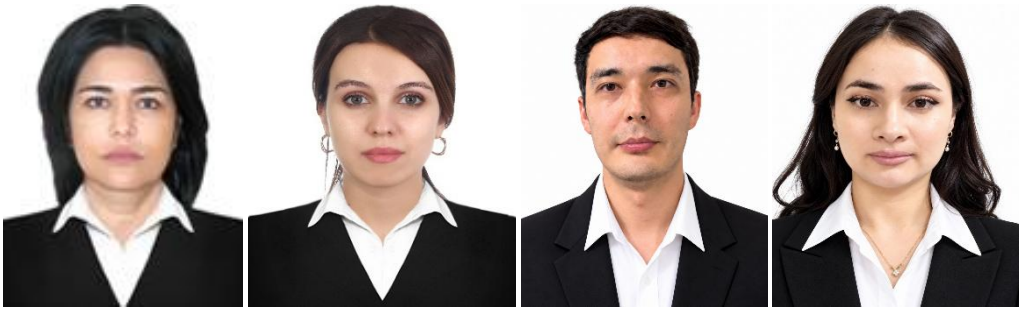
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PLEOMORPHIC XANTHOASTROCYTOMA, CNS WHO GRADE 3, IN A FOUR-YEAR-OLD CHILD: RADIOLOGIC–PATHOLOGIC CORRELATION OF A RARE CORTICAL TUMOR



Djuraeva Nigora Mukhsumovna, Abdukhalimova Khanum Valentinovna, Kholmirzaev Murodjon Ali ugli, Abduvakhobova Komila Mirsaid kizi
Republican Specialized Scientific and Practical Medical Center of Surgery named after Academician V. Vakhidov, Republic of Uzbekistan, Tashkent

ТЎРТ ЁШЛИ БОЛАДА ПЛЕОМОРФ КСАНТОАСТРОЦИТОМА, CNS WHO GRADE 3: КАМ УЧРАЙДИГАН КОРТИКАЛ ЎСМАНИНГ РАДИОЛОГИК-ПАТОЛОГИК КОРРЕЛЯЦИЯСИ

Джураева Нигора Мухсумовна, Абдухалимова Ханум Валентиновна, Холмирзаев Муроджон Али ўғли, Абдувахобова Комила Мирсаид қизи
Академик В. Воҳидов номидаги Республика ихтисослаштирилган илмий-амалий тиббиёт маркази давлат муассасаси, Ўзбекистон Республикаси, Тошкент ш.

ПЛЕОМОРФНАЯ КСАНТОАСТРОЦИТОМА, CNS WHO GRADE 3, У ЧЕТЫРЁХЛЕТНЕГО РЕБЁНКА: РАДИОЛОГО-ПАТОЛОГИЧЕСКАЯ КОРРЕЛЯЦИЯ РЕДКОЙ КОРТИКАЛЬНОЙ ОПУХОЛИ

Джураева Нигора Мухсумовна, Абдухалимова Ханум Валентиновна, Холмирзаев Муроджон Али угли, Абдувахобова Комила Мирсаид кизи
ГУ «Республиканский специализированный научно-практический медицинский центр хирургии имени академика В. Вахидова», Республика Узбекистан, г. Ташкент

e-mail: komilakhonabdukodirova190117@gmail.com

Резюме. *Мақсад: бола ёшидаги кам учрайдиган кортикал ўсма – плеоморф ксантоастроцитоманинг (ПКА) МРТ белгилари ва радиологик-патологик корреляциясини клиник кузатув мисолида тавсифлаш. Материал ва усуллар: бош оғриғи, кўнгил айниши, қусиш, бош айланиши ва юришда беқарорлик билан мурожаат қилган 4 ёшли қиз болага 3,0 Т МРТ (DWI/ADC, SWI, контрастли Т1, диффузион-тензор трактография), микрожарроҳлик амалиёти, гистологик ва иммуногистокимёвий текширувлар ўтказилди. Натижалар: МРТда ўнг пешона-тепа соҳасининг латерал қисмларида 5,8×5,7×6,4 см ўлчамли, асосан кистоз, периферик солид компонентли, нотекис периферик контрастланувчи, яққол вазоген шиш ва масс-эффект билан кечувчи кортико-субкортикал ҳосила аниқланди. Ўсма субтотал олиб ташланди. Гистологик ва иммуногистокимёвий текширувда GFAP, Olig2 ва CD34 мусбат, IDH1 ва BRAF V600E манфий, Ki-67 тахминан 35% бўлган ўсма аниқланиб, ПКА, CNS WHO grade 3 (ICD-O 9424/3) ташхиси қўйилди; операциядан кейин адъювант нур терапияси ўтказилди. Хулоса: болаларда бош миянинг йирик юза жойлашган кистоз ҳосилаларида ПКАни дифференциал ташхисга киритиш лозим; якуний ташхис МРТ маълумотларини гистологик ва иммуногистокимёвий натижалар билан биргаликда талқин қилишни талаб этади.*

Калит сўзлар: *плеоморф ксантоастроцитома, кўп параметрли нейровизуализация, кистоз-солид ҳосила, иммуногистокимёвий текширув, радиологик-патологик корреляция.*

Abstract. *Aim: to describe the MRI features and radiologic–pathologic correlation of a rare paediatric cortical tumour, pleomorphic xanthoastrocytoma (PXA), using a clinical case. Materials and methods: a 4-year-old girl presenting with headache, nausea, vomiting, dizziness and gait instability underwent 3.0 T MRI (DWI/ADC, SWI, contrast-enhanced T1, diffusion tensor tractography), microsurgical resection, and histological and immunohistochemical examination. Results: MRI showed a 5.8×5.7×6.4 cm predominantly cystic cortico-subcortical mass in the lateral right frontoparietal region with a peripheral solid component, irregular peripheral enhancement, marked vasogenic oedema and mass effect. Subtotal resection was performed. The tumour was positive for GFAP, Olig2 and CD34 and negative for IDH1 and BRAF V600E, with a Ki-67 index of approximately 35%; PXA, CNS WHO grade 3 (ICD-O 9424/3), was diagnosed, and adjuvant radiotherapy was given. Conclusion: PXA should be included in the differential diagnosis of large*

superficial cystic cerebral masses in children; the final diagnosis requires interpretation of MRI findings together with histological and immunohistochemical data.

Keywords: *pleomorphic xanthoastrocytoma, multiparametric neuroimaging, cystic-solid mass, immunohistochemistry, radiologic–pathologic correlation.*

Pleomorphic xanthoastrocytoma (PXA) is a rare circumscribed astrocytic tumour that occurs in young people and corresponds to CNS WHO grade 2 or 3 [2]. It accounts for less than 1% of all astrocytic tumours, and its frequency is roughly one-tenth that of pilocytic astrocytoma [3]. PXA typically affects children and young adults, especially in the second decade of life; most tumours are superficial and involve the cortex and meninges [3]. PXA is most commonly found in the temporal lobe, and seizures are the most frequent presenting symptom. Histologically, PXA and anaplastic PXA are distinguished, differing in grade, malignancy and prognosis. Surgery is the treatment of choice, and total resection is usually associated with a favourable prognosis.

On imaging, PXA typically appears as a cortical tumour with a cystic component and strong contrast enhancement, particularly in the temporal lobe. Slow growth may be suggested by the absence of surrounding oedema and by scalloping of the adjacent bone; a dural tail indicates reactive dural involvement, whereas calcifications are infrequent [7]. We report a case of PXA, CNS WHO grade 3, in a 4-year-old girl, focusing on the MRI findings, the main imaging differential diagnoses and the correlation with the pathological diagnosis.

Aim of the study: to describe the MRI features and radiologic–pathologic correlation of pleomorphic xanthoastrocytoma, CNS WHO grade 3, in a young child.

Case presentation. A 4-year-old girl presented with a several-month history of headaches, nausea, vomiting, dizziness and gait instability. The neurological symptoms progressed despite initial conservative outpatient treatment. Brain MRI revealed a large right hemispheric lesion, and the patient was referred to a neurosurgical centre.

Imaging findings. MRI of the brain was performed on a 3.0 T Magnetom Vida system (Siemens Healthineers, Erlangen, Germany). The protocol included sagittal three-dimensional magnetisation-prepared rapid gradient echo, dark-fluid T2-weighted imaging, axial and coronal T2-weighted turbo spin-echo imaging, diffusion-weighted imaging with apparent diffusion coefficient maps, susceptibility-weighted imaging, three-dimensional time-of-flight angiography, post-contrast T1-weighted imaging after intravenous administration of 20.0 mL of Magnilek, and diffusion tensor tractography. Imaging (24.09.2025) demonstrated an irregularly rounded cystic-solid lesion with relatively clear, uneven contours, centred in the lateral parts of the right frontoparietal region and compressing the posterior horn of the right lateral ventricle, measuring approximately 5.8×5.7×6.4 cm (Fig. 1). Marked mass effect was present: the adjacent regions of the right hemisphere were compressed, with zones of perifocal vasogenic oedema and a leftward midline shift of approximately 7 mm.

The solid portions of the lesion were predominantly hypo- to isointense on T1-weighted images and heterogeneously intermediate to hyperintense on T2-weighted images, whereas the cystic component had low signal characteristics. Diffusion-weighted imaging showed a slightly hyperintense signal in the solid component with mild peripheral oedema. After contrast administration, moderate enhancement of the solid component and of the walls of the cystic component was noted. In the subcortical and periventricular white matter of both hemispheres, asymmetric single round foci (about 3 mm in diameter) with indistinct margins, homogeneously hyperintense on T2-weighted and T2 dark-fluid images, were identified. Diffusion tensor tractography demonstrated displacement and compression of the superior fronto-occipital fasciculus, corticopontine tract, anterior thalamic radiation, superior longitudinal fasciculus, posterior limb and retrolenticular part of the internal capsule, corticospinal tract, superior thalamic radiation and stria terminalis (Fig. 2).

Surgical treatment and follow-up. On 7 October 2025 the patient underwent right fronto-temporal craniotomy with subtotal resection of the cystic-solid mass. The postoperative course was uneventful, and early postoperative imaging showed no acute complications. Follow-up MRI on 5 November 2025 revealed postoperative changes with residual enhancing tumour tissue in the surgical bed: an irregularly shaped postoperative cystic area measuring 2.8×1.9×2.9 cm with a small zone of peripheral oedema, heterogeneous signal and intense uneven peripheral enhancement (Fig. 4). Areas of linear signal loss spreading in a fan-like pattern from the deep structures to the cortex, measuring 2.7×1.7 cm and 1.4×0.6 cm, with peripheral gliosis and moderate peripheral enhancement, were also detected. Spinal MRI did not reveal leptomeningeal dissemination or metastases. Given the diagnosis of PXA, CNS WHO grade 3, and residual disease, adjuvant radiotherapy was administered.

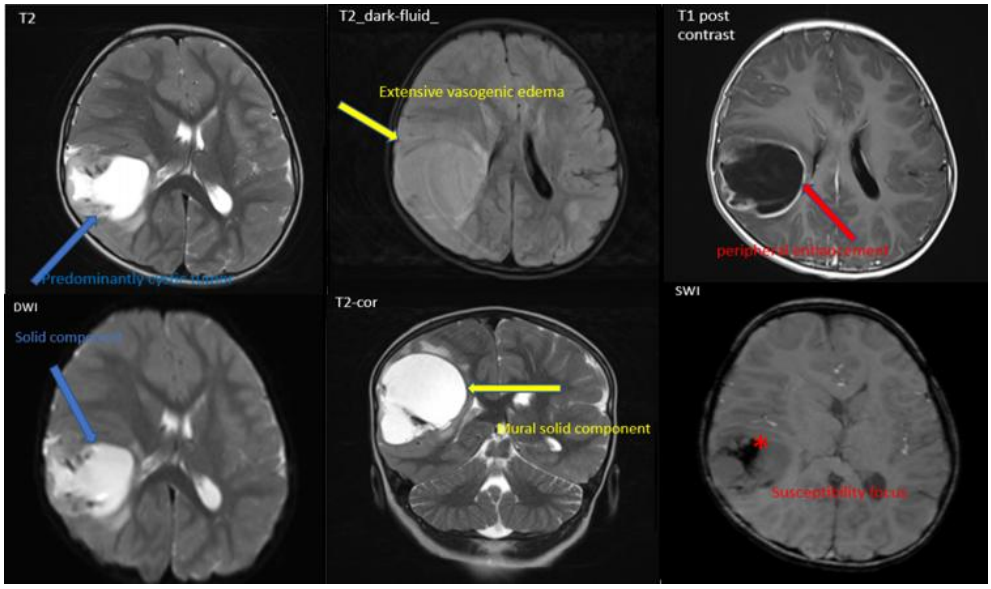


Fig. 1. Preoperative MRI: predominantly cystic cortico-subcortical mass with a solid component, peripheral enhancement and extensive vasogenic oedema

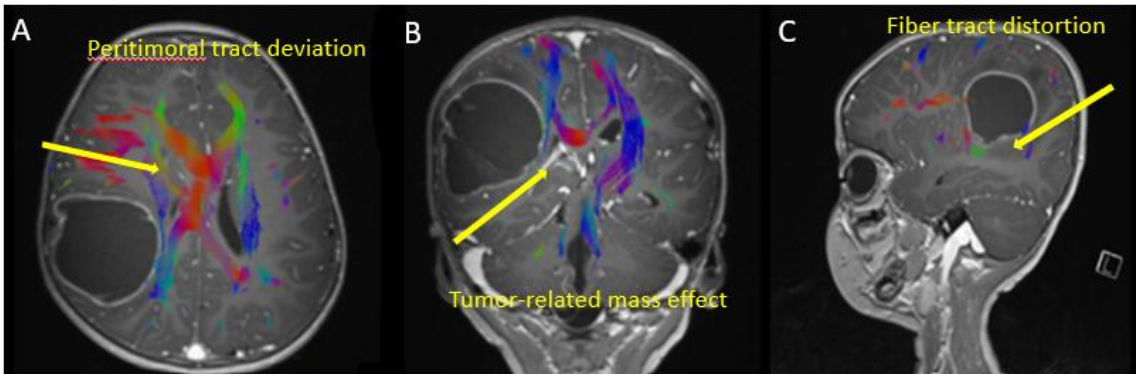


Fig. 2. Preoperative MR tractography: displacement and distortion of the adjacent white-matter tracts (A–C)

Histopathological findings. The tumour tissue contained numerous bizarre giant xanthomatous cells with one or more large nuclei, intracellular vacuoles and well-defined light cytoplasm. The neoplasm formed solid sheets; the nuclei were predominantly hyperchromatic, oval and irregular, with pronounced pleomorphism, multinucleated giant cells and increased mitotic activity. Extensive necrosis, moderate endothelial proliferation and focal lymphoid infiltration were noted (Fig. 3). Immunohistochemically, the tumour was positive for GFAP, Olig2 and CD34 and negative for IDH1 and BRAF V600E; the Ki-67 proliferation index was approximately 35%. The final diagnosis was anaplastic PXA, CNS WHO grade 3 (ICD-O 9424/3).

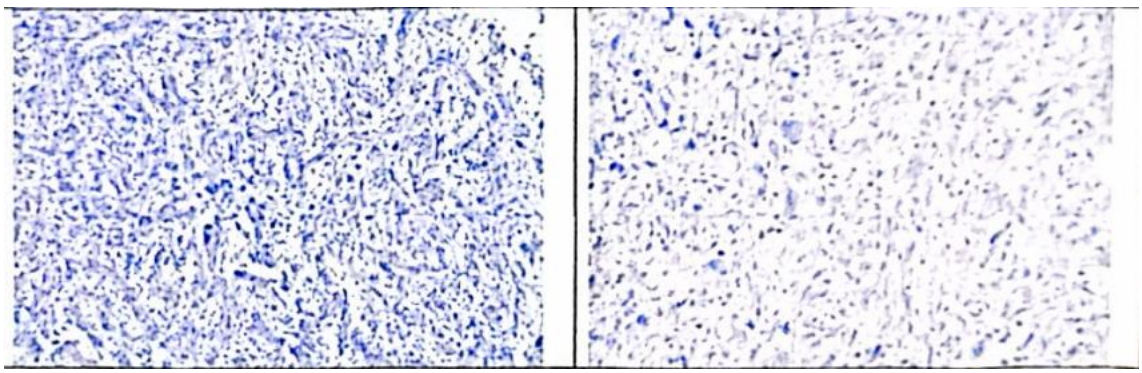


Fig. 3. Histopathological findings: pleomorphic tumour cells with hyperchromatic irregular nuclei and multinucleated giant cells

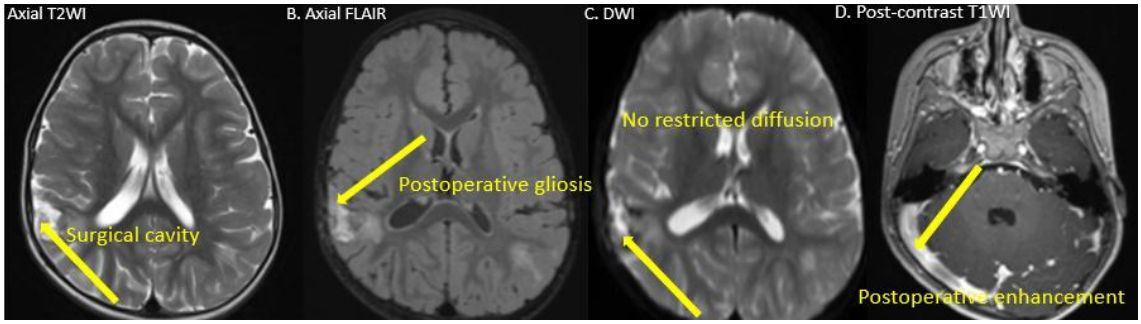


Fig. 4. Postoperative MRI after subtotal resection: A – axial T2WI, B – axial FLAIR, C – DWI, D – post-contrast T1WI

Discussion. PXA accounts for less than 1% of primary intracranial tumours. Histologically, it is characterised by numerous pleomorphic giant cells, spindle cells and foamy (xanthomatous) cells expressing glial fibrillary acidic protein, a reticulin network and eosinophilic granular bodies [6, 8]. The tumours are located superficially, involve the meninges and may originate from subpial astrocytes; rarely they arise in the cerebellum, spinal cord or retina, but most are found in the cerebral hemispheres. In the current WHO CNS classification, PXA corresponds to CNS WHO grade 2 or 3; up to 20% of tumours progress to a higher grade, and the main feature of anaplasia is high mitotic activity (5 or more mitoses per 10 high-power fields).

PXA is a tumour of the young (about 60% of cases occur before the age of 18), without sex predominance. These slow-growing tumours typically cause long-standing seizures, headaches and dizziness. More than 70% of patients survive 10 years after diagnosis, but progression to less differentiated tumours is possible. Surgery is the treatment of choice: total resection gives the best results, and repeat resection is required for recurrence [5]. Chemotherapy and radiotherapy may be used for unresectable tumours and for primary or recurrent anaplastic PXA, but their efficacy has not been proven; the extent of initial resection and mitotic activity are the main predictors of survival.

On imaging, PXA is a supratentorial cystic tumour at the periphery of the cerebral hemisphere (60%) with a solid component adjacent to the meninges; the solid nodule enhances after contrast administration, and a dural tail may be present (70%). An arc-shaped defect of the adjacent inner table of the skull may be seen. The tumours are most often located in the temporal, frontal and parietal lobes but may occur anywhere in the cerebral hemispheres, including the ventricles. Tumour size and the degree of enhancement vary; calcifications and haemorrhages are infrequent. The signal of the solid component varies, whereas the cyst usually follows cerebrospinal fluid on all sequences. Parenchymal invasion, metastases and necrosis occur rarely.

The tumour is removed as completely as possible; after complete removal, only serial MRI is required to monitor for regrowth, and repeat surgery may be performed if a residual tumour grows or recurs [1]. Gross total resection usually eliminates seizures; radiotherapy may be considered but does not influence long-term outcomes. In our patient, the Ki-67 index of approximately 35% indicated high proliferative activity and an unfavourable prognosis, and the absence of the BRAF V600E mutation may limit the potential response to targeted therapy.

The main radiological differential diagnosis includes other cortical tumours. Ganglioglioma is among the closest differentials: both lesions occur at a young age and may appear as a superficial cystic-solid mass; a partially cystic mass with an enhancing mural nodule is seen in about 45% of cases, but enhancement is often less prominent, calcification is present in about 50%, and the dural tail sign is absent. Dysembryoplastic neuroepithelial tumours are predominantly cortical, well circumscribed and have a characteristic «bubbly» appearance without enhancement. Desmoplastic infantile ganglioglioma manifests as a very large cystic-solid hemispheric mass (usually more cystic, with a mural nodule) involving both the cortex and meninges, most often in the frontal and parietal lobes. Pilocytic astrocytoma (CNS WHO grade 1) typically appears as a cyst with an enhancing mural nodule, often adjacent to the ventricular system [4].

Conclusions. Pleomorphic xanthoastrocytoma, CNS WHO grade 3, should be included in the differential diagnosis of large superficial cystic cerebral masses in children.

Superficial cortico-subcortical location, a predominant cystic component, peripheral enhancement and extensive vasogenic oedema without marked diffusion restriction on MRI may suggest the diagnosis; definitive diagnosis requires histopathological and molecular analysis according to the current WHO CNS criteria. This case illustrates the importance of radiologic–pathologic correlation in the evaluation of rare paediatric brain tumours.

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ПЛЕОМОРФНАЯ КСАНТОАСТРОЦИТОМА, CNS WHO GRADE 3, У ЧЕТЫРЁХЛЕТНЕГО РЕБЁНКА: РАДИОЛОГО-ПАТОЛОГИЧЕСКАЯ КОРРЕЛЯЦИЯ РЕДКОЙ КОРТИКАЛЬНОЙ ОПУХОЛИ

Джураева Н.М., Абдухалимова Х.В., Холмирзаев М.А., Абдувахобова К.М.

Резюме. *Цель: описать МРТ-признаки и радиолого-патологическую корреляцию редкой кортикальной опухоли детского возраста – плеоморфной ксантоастроцитомы (ПКА) – на примере клинического наблюдения. Материал и методы: девочке 4 лет, поступившей с головной болью, тошнотой, рвотой, головокружением и неустойчивостью походки, выполнены МРТ 3,0 Тл (DWI/ADC, SWI, T1 с контрастированием, диффузионно-тензорная трактография), микрохирургическое удаление опухоли, гистологическое и иммуногистохимическое исследования. Результаты: по данным МРТ в латеральных отделах правой лобно-теменной области выявлено преимущественно кистозное кортико-субкортикальное образование размером 5,8×5,7×6,4 см с периферическим солидным компонентом, неравномерным периферическим контрастированием, выраженным вазогенным отёком и масс-эффектом. Выполнено субтотальное удаление опухоли. Опухоль была GFAP-, Olig2- и CD34-положительной, IDH1- и BRAF V600E-отрицательной, индекс Ki-67 – около 35%; установлен диагноз ПКА, CNS WHO grade 3 (ICD-O 9424/3), проведена адъювантная лучевая терапия. Выводы: ПКА следует включать в дифференциальный ряд крупных поверхностно расположенных кистозных образований головного мозга у детей; окончательный диагноз требует совместной интерпретации данных МРТ, гистологического и иммуногистохимического исследований.*

Ключевые слова: *плеоморфная ксантоастроцитома, мультипараметрическая нейровизуализация, кистозно-солидное образование, иммуногистохимическое исследование, радиолого-патологическая корреляция.*