Pleomorphic Xanthoastrocytoma mimicking meningioma in an adult.

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Published on: Apr 15, 2023
DOI: https://doi.org/10.21428/077ce266.10bd496d
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History
A 48-year-old woman presented with a headache with neck spasms on her right side.

Keywords:
Pleomorphic Xantho-astrocytoma (PXA), Meningioma, Hemangiopericytoma, Extra-axial lesions

DICOM images
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Imaging Findings:

![Image of MRI head showing a heterogenous intensity lesion in the right frontal region.]

Figure 1
Right frontal region shows a heterogenous intensity lesion.

MRI head
The right frontal region shows a heterogenous intensity lesion with the perilesional cystic component. The cystic component has a blood fluid level within (T2 hypointense blood has gravitated inferiorly, and T2 hyperintense water is noted superiorly). The solid component has a heterogenous signal.
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Figure 2
Pre-contrast T1 and post-contrast fat-suppressed T1 images. The solid component shows avid contrast enhancement.

Post-contrast scan

There is avid enhancement noted in the solid component of the lesion.

Figure 3
Flair and T1 coronal images demonstrate the cortical buckling and dural-based enhancing component.

Flair and T1 coronal images (Figure 3) demonstrate the cortical buckling and dural-based enhancing component. A small vessel flow void is noted on FLAIR seems displaced away from the lesion. T1 post-contrast sagittal image shows a dural tail sign in its posterior aspect. These findings are suspicious of an extra-axial lesion, likely a meningioma.
However, signal intensity on T2 is not as hypointense as it would be suspected in meningioma.

Diffusion, unfortunately, cannot add more value because of the surrounding hemorrhage.
Inference:
Given the heterogeneous signal on T2, with signs suggestive of the extra-axial lesion. We were thinking about extra-axial lesions like meningioma. Meningiomas are known to be associated with peritumoral intraparenchymal cysts. However, it is rare for those to bleed.

Because the lesion is not predominantly hypointense, we considered other entities like hemangiopericytoma, but there are no large flow voids to suggest high vascularity. Microcystic meningioma can look T2 hyperintense.

We also considered differentials of cysts with mural nodules, pleomorphic xantho-astrocytoma (PXA), and pilocytic astrocytoma. Both are more common in the pediatric population. Pleomorphic astrocytomas can be closely abutting the meninges, making it harder to differentiate from extra-axial lesions. Therefore PXA became the second likely differential after meningioma.

Lymphoma was not in the differential as the tumor was not hypointense on T2 and did not show diffusion restriction. Metastasis can mimic any dural-based lesion; therefore, it is always a differential. However, because of the lack of prior tumor history was considered a less probable differential.

Differential diagnosis
Meningioma

Pleomorphic xantho-astrocytoma (PXA)

Pilocytic astrocytoma

Metastasis
Pathology correlation

The tissue sample showed findings suggestive of anaplastic pleomorphic xanthoastrocytoma, WHO grade 3 likely. Showing microvascular proliferation. GFAP staining was positive. BRAF and IDH mutation studies were not performed.

Diagnosis

Pleomorphic Xantho-astrocytoma.

Discussion:

Our case demonstrates atypical looking at extra-axial lesions can have an unusual, rare diagnosis. And not all dural tails are meningiomas. Along with the enhancement pattern and traditional signs of intra v/s extra-axial lesions should not be given more weightage over the signal intensity of the lesion. Kawano et al. described PXA can cause a desmoplastic reaction as they arise on the brain surface with strong adhesions to the dura mater [1]. This can cause a dural tail sign [2]. Case series by Schmidt et al. have shown pleomorphic xanthoastrocytomas can be present in adult patients [3]. Differentials of the cyst with mural nodules include other entities like haemangioblastoma (peak age 40–60 years), which was not considered due to lack of flow voids, gangliogliomas (peak age 10–20 years) which are more temporal based and in younger populations, however, can not be totally excluded, DIG (desmoplastic infantile ganglioglioma) is as the name suggest happen in infants (<2 years) of age [4]. Pilocytic astrocytoma (peak age 5–15 years) is also a valid differential; however, they do not attach to meninges compared to PXAs. A study by She et al. have demonstrated the utility of diffusion-weighted imaging in differentiating pilocytic astrocytoma from PXAs, PXAs showing more diffusion restriction compared to pilocytic astrocytoma [5]. According to WHO 2021 classification of brain tumors, pilocytic astrocytomas, and pleomorphic xantho-astrocytomas are classified under ‘Circumscribed astrocytic gliomas’ [6]. Our case also showed a hemorrhage in the cystic component of the tumor. Few case reports in English literature have described PXA as a hemorrhagic lesion [2] [7].

Supratentorial ependymoma can have meningeal involvement and, therefore, can show a dural tail, mimicking meningioma [8][9][10]. It would be difficult to differentiate PXAs from such meningeal involving supratentorial ependymoma. Rarely supratentorial ependymoma can present as a cyst with a mural nodule [11][12]. More studies to differentiate them based on diffusion and perfusion parameters are needed.

The histologic classification of PXAs with anaplastic characteristics is controversial since some patients, despite anaplastic features, may have a very long survival while others show relatively rapid progression. High mitotic rates, necrosis, and rhabdoid cells are histologic characteristics related to poor survival.[13] It becomes important to distinguish PXA from IDH wild glioblastoma in adult patients as most PXAs are IDH wild. BRAF V600E mutation may guide in such cases. However, one case report suggests that epithelioid glioblastoma can arise from previously known PXA tumor even after a decade [14]. This suggests there is a lot we do not yet know about the pathogenesis of IDH wild glioblastomas.
Some of these lesions and PXA are considered part of low-grade, developmental, epilepsy-associated brain tumors (LEAT), which are not related pathologically but more to clinical presentation [15]. However, our case is not low-grade PXA, and the patient did not present with epilepsy. Lyndon et al. list many dural-based lesions, neoplastic and non-neoplastic, which can meningioma but do not list PXAs or Supratentorial ependymoma, likely because they are rare [16].

Genetic mutations are important in differentiating PXA from other tumors and planning treatment strategies [17]. Unfortunately, our case sample did not undergo genetic testing. PXAs, gangliogliomas, and extracerebellar pilocytic astrocytomas can show BRAF V600E mutation, while this mutation is absent in diffuse astrocytic tumors [18][19]. BRAF V600E mutation should not be confused with BRAF gene fusions [20]. CDKN2A/B loss and TERT promoter mutations have been described in both anaplastic PXAs and anaplastic meningiomas [21][22]. Thus, brain tumor classification is more complicated not only for radiologists but also for histopathology colleagues who now need to access various genetic markers. It is critical that we are aware of these genetic breakthroughs in order to have discussions with our pathology colleagues in tumor board meetings for the benefit of our patients.

References


