Pleomorphic xanthoastrocytoma
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Section: Neuroradiology
Imaging Technique: CT
Imaging Technique: MR
Case Type: Clinical Cases
Authors: Chourmouzi D, Drevelegas A, Boulogianni G
Patient: 36 years, male

Clinical History:

The patient had a 2-month history of epilepsy. His electroencephalogram showed localised epileptic discharge.

Imaging Findings:

The patient had a 2-month history of epilepsy. His electroencephalogram showed localised epileptic discharge. His past medical history was clear. CT and MR were performed.

MR was performed on a 1.5T MRI scanner, with SE T1-weighted, FSE T2-weighted, and post-contrast SE T1-weighted sequences in the sagittal, axial and coronal planes. A large right cortical parietal cystic lesion with solid mural nodule was found, suggestive of a low-grade tumour (Figs 1 and 2). A complete resection of the tumour was made on operation.

Pathological study showed a cellular pleomorphic tumour with perivascular lymphocytes, compatible with pleomorphic xanthoastrocytoma.

Discussion:

Pleomorphic xanthoastrocytoma (PXA) was first described by Kepes et al. in 1979. It is a circumscribed astrocytoma frequently characterised by a large cystic component with a superficial mural nodule, which is usually attached to the leptomeninges. As this type of tumour is generally benign, especially when compared with diffuse astrocytomas, its identification is of therapeutic and prognostic importance.

Pleomorphic xanthoastrocytomas (PXA) are rare tumours of children and young adults, comprising less than 1% of all gliomas. They are generally classified as WHO classification grade II neoplasms, but have been known to undergo malignant transformation. Like pilocytic astrocytomas, PXA occur more frequently in the first two decades of life. Unlike pilocytic astrocytomas they occur more commonly in the cerebral hemispheres with a predilection for the temporal lobes followed by the parietal, occipital and frontal lobes.

PXA is considered to be a relatively benign variant of astrocytoma and to arise from the subpial astrocytes of the superficial cortex. It is histologically characterised by pleomorphic, lipid-laden neoplastic cells with surrounding reticulin layers, and exhibits immunoperoxidase staining for GFAP.

Microscopically pleomorphism is the hallmark of PXAs in which spindle and round cells are the main population. If only a small amount of tumour is provided to the pathologist for analysis, PXA may be confused with glioblastoma.

The MRI and CT appearance of the tumour may assist pathological interpretation of this tumour.

PXA presents either as a cyst with mural nodule or, less commonly, as a completely solid tumour. The mural nodule
is usually attached to the leptomeninges. On unenhanced CT the mural nodule or the solid portion of the tumour appears hypo- or hyperdense. After the administration of contrast medium PXAs enhance markedly. MRI reveals that, relative to grey matter, the solid component of these tumours is of similar signal intensity on T1-weighted images and increased signal intensity on T2-weighted sequences. Imaging features of low grade, such as lack of peritumoral oedema and calvarial scalloping, frequently accompany these tumours. Post-contrast T1-weighted imaging shows intense enhancement of the mural nodule or of the solid tumour. The wall of the cyst may or may not be enhanced. Dural leptomeningeal, or gyriform enhancement may be present.

Differential diagnosis includes pilocytic astrocytoma, gaglioglioma, and oligodendroglioma. Because of their peripheral location, they can be confused with meningioma.

Unlike malignant gliomas, PXA is not treated with radiotherapy after partial removal. Therefore, it is important to recognise and identify this type of glioma as a distinct entity; however, both recurrence and anaplastic transformation to more aggressive histological properties have been described. If PXA has a high probability of recurrence, it may be necessary to perform total removal or radiotherapy.

**Differential Diagnosis List:** Pleomorphic xanthoastrocytoma

**Final Diagnosis:** Pleomorphic xanthoastrocytoma

**References:**

Miyagi Y, Suzuki SO, Iwaki T, Shima F, Ishido K, Araki T, Kamikaseda K.

Tien RD, Cardenas CA, Rajagopalan S.

Description: Post-contrast CT shows a hypodense right parietal lesion with hyperdense mural nodule.
 Origin:
Description: Axial T1-weighted image (TR/TE: 570/15) shows a well-circumscribed, parietal lobe cortex, low signal lesion with an isointense mural nodule (arrow). Origin:
Description: On axial T2-weighted image (TR/TE: 4000/90) the lesion shows high signal intensity while the mural nodule is isointense to grey matter. Note the absence of peritumoral oedema. Origin:
Description: Coronal post-contrast T1-weighted image shows intense enhancement of the mural nodule. Note the enhancement of the wall as well. Origin: