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PLEOMORPHIC XANTHOASTROCYTOMA IN A 40-YEAR-OLD PATIENT: A CASE REPORT

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INTRODUCTION

Pleomorphic xanthoastrocytoma (PXA) is a low-grade brain tumor (WHO II) that was first reported by Kepes et al. in 1979 [1]. PXA accounts for less than 1% of all brain tumors [2] and the majority of tumors are located in the supratentorial compartment, mainly in the temporal lobe, followed by the frontal lobe and parietal lobe (1). diagnosed in the second decade of life. The average age at diagnosis is 29 ± 16 years, with no predominance in terms of gender (2). The onset symptoms are consistent with mass effect, predominantly headache and neurological disorders that vary from focal symptoms, depending on the location, to epileptic seizures, which is the most common initial symptom(3).

On imaging exams, it has characteristics that resemble high-grade gliomas. They most commonly occur as well-circumscribed superficial masses with leptomeningeal contact. These tumors usually consist of solid and cystic components (2).

On contrast-enhanced Computed Tomography (CECT), the appearance of the tumor is variable, with cystic components generally appearing hypodense and solid components appearing hypo- to isodense and rarely hyperdense. The solid component and wall of the cyst usually show moderate to intense enhancement. Focal calcification may also be seen in the solid component. There may be a cutout of the overlying skull (4).

In the Magnetic Resonance Imaging (MRI) examination, the signal is isointense on T1, hyperintense on T2 and in the post-gadolinium T1 sequence, its solid component presents heterogeneous contrast enhancement, with peripheral enhancement of the edge of the cyst (2).

This tumor is most often located in the temporal lobe. Its histopathology was first described after microscopic review of 12 cases of almost identical appearance. It is a moderately cellular tumor, predominantly pleomorphic, with foci of lymphoplasmacytic infiltration, without necrosis and rare mitoses. Tumor cells are spindle-shaped with elongated nuclei or large round cells with a single or multilobed nucleus or multiple nuclei (bizarre multinucleated giant cells) that invade the brain parenchyma directly or into perivascular spaces. There is also evidence of an external basal lamina on electron microscopy, which consists of reticulin fibers produced and present around tumor cells.

The overall survival of PXA is favorable, with 3- and 5-year survival rates of 80% and 75%, respectively. Progression-free survival rates at 3 and 5 years are greater than 60%. This decreases with increasing grade/anaplasia. It should be kept in mind that a subset of patients with PXA are considered cured of the disease even with surgical resection alone. This distinguishes PXA from other infiltrative gliomas (2).

A total resection correlates with 10-year overall survival (OS) of 82%. As routine practice, total resection (RT), if feasible and safe, should be the surgeon's goal. Some reports have noted an association with the use of RT and improved progression-free survival (PFS).

CASE REPORT

A 40-year-old female patient with a history of cerebral neoplasia sought the General Hospital of Fortaleza (HGF) due to her first convulsive episode, without sphincter release, lasting 10-15 minutes.

The head CT revealed an expansive brain lesion with a neoplastic appearance with an epicenter in the third ventricle, paramedian to the right, extending towards the lateral ventricles, extending inferiorly to the right nucleus-capsular region, contrast-enhancing, with cystic areas in between.

The patient had another episode of seizures without sphincter release one day after admission.

The patient underwent complete resection of the intracranial tumor, with the apposition of an external ventricular shunt (EVD) and sending the material for histopathological examination.

Histopathological examination confirmed the hypothesis of PXA.

One day after surgery, a head CT was performed for follow-up, which demonstrated the usual postoperative findings, such as bilateral frontal pneumocephalus, heterogeneous frontal and periventricular hypodensity with hemorrhagic foci, with effacement of adjacent cortical sulci. Seven days after surgery, another CT scan was performed, which revealed a gradual decline in the findings, with a frontal subdural hematoma.

An MRI was performed 13 days after surgery, with additional findings of microangiopathy, unrelated to the surgery.

The patient's postoperative course was also marked by significant clinical complications, including hyponatremia, indicative of a salt wasting syndrome, and a hospital infection in the form of pneumonia, requiring the administration of antimicrobials.

DISCUSSION

The case described shows remarkable agreement with the current literature since the clinical presentation was an epileptic seizure, which is the most common initial symptom of PXA, however there are reports that mental confusion and headache are also possible initial symptoms of this pathology. A peculiarity observed is that the patient is 40 years old, which is an unusual age at which PXA is affected, given that the average age at diagnosis is between 16-29 years old. However, the literature reports rare cases of PXA in patients aged 2 to 68 years (2).

These tumors have a predilection for the temporal lobe, therefore, it normally presents, as a clinical aspect, seizures in most cases. However, there are other less specific manifestations, such as dizziness, headache and in some cases the patient may present asymptomatically. PXA can be classified as grade II or III in the WHO classification. This classification is based on their histological characteristics. Type III have an increased mitotic rate of more than 5 mitoses per 10 high-power fields, but are histologically very similar to grade 2 tumors, although necrosis and vascular proliferation are more common.

They can be difficult to distinguish from epithelioid glioblastomas. PXA appears on radiological examinations as a solid-cystic lesion, with a solid component that presents with isointensity or hypointensity on T1 sequences, with a cystic component with hypointensity on T1 and hypersignal on T2. In the FLAIR sequence, it appears more hyperintense than CSF, as it has a high protein content. Furthermore, because it is slow growing, it may not have surrounding vasogenic edema, which is found in faster growing masses. There may be indentation of the overlying internal bone plate, due to its peripheral location. Furthermore, there may be reactive dural involvement, expressed as a dural tail sign and rarely presents with calcifications.

Histological features are variable, with spindle cells, polygonal cells, multinucleated cells, and lipid-laden xanthomatous astrocytes are also identified. Furthermore, it presents nuclei with a pleomorphic appearance, with common nuclear inclusions and highly variable nuclear size.

Immunohistochemistry shows positivity for Glial fibrillary acid protein (GFAP), which is positive when there is the presence of glial cells, tumors of glial origin. Furthermore, it

presents variable nuclear and cytoplasmic positivity for S-100, which is a family of cytoplasmic calcium-binding proteins expressed in numerous cell lines that can be targeted by immunohistochemistry. Furthermore, it is positive in about 1% of cells in Ki-67.

PXA do not have epithelioid cells, but often exhibit eosinophilic granular bodies and xanthomatous change. Furthermore, they are characterized by the abundant presence of reticulin, and the frequent presence of intracellular inclusion and lymphocytic infiltration. Glioblastomas are distinguished from xanthocrocytomas by having epithelioid cells. They rarely exhibit eosinophilic granular bodies, xanthomatous change and reticulin. On the other hand, glioblastomas are characterized by microvascular proliferation and the presence of necrosis (4,5).

CONCLUSION

PXA is a rare low-grade brain tumor that mainly presents in patients in their 2nd decade of life with clinical symptoms such as seizures and headache. From this perspective, exams such as MRI, CT, immunohistochemistry are extremely important for the differential diagnosis of the disease.

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