Clinical, histological and imaging aspects of pleomorphic xanthoastrocytomas: the key is in the name

Aspectos clínicos, histológicos e imagenológicos dos xantoastrocitomas pleomórficos: a chave está no nome

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Pleomorphic xanthoastrocytoma (PXA) is a rare tumor of the central nervous system, first described in 1979[1]. It was added to the World Health Organization (WHO) classification of tumors in 1993[2]. First reports emphasized that despite their highly pleomorphic and bizarre cytology, which suggested a malignant behavior, these tumors appeared to have a relatively favorable prognosis in most of the cases[1], with a 75% overall survival at 5 years[3].

PXA were probably some of the first tumors in whose characterization immunohistochemical markers were essential. Namely, the demonstration of glial fibrillary acidic protein (GFAP) led to the definition of PXA as a new clinicohistopathologic tumor entity of astrocytic nature[1]. Prior to its initial description, PXA was often classified among giant cell glioblastomas or mesenchymal tumors such as fibrous xanthomas or sarcomas (due to its rich reticulin network)[4].

In the following years, clinical and surgical series were reported[5]; patients with PXA typically present as children or young adults with almost 70% under 30 years of age, and equal frequency of occurrence in both males and females[4,6]. Presenting symptoms consist primarily of seizures, often of long duration[3,4]; followed by dizziness, headache and focal neurological deficit[6].

Studies focusing on imaging aspects of PXA demonstrated that these tumors are often superficial with leptomeningeal extension and involve the temporal lobe most frequently (up to almost 50% in a large series)[4]; other common locations include the parietal, frontal, and occipital lobes. Patterns of presentation can be purely cystic (uni or multilocular), mixed cystic-solid and purely solid; the solid portions usually enhance intensely after the use of contrast material[6,7]. Remodeling of the adjacent calvarium can be appreciated, related to the slow growth of this superficial lesion.

Surgery is clearly the treatment of choice, and the extent of primary resection has great impact on prevention of recurrence and survival rates. Cases in which a gross total removal is achieved, a "wait and see" approach can be recommended followed, if necessary, by additional surgery for residual and/or recurrent tumor[4]. At present, the role of adjuvant radiotherapy and/or chemotherapy remains uncertain, but it seems that patterns of care are following the treatment patterns of gliomas[3].

More recently, unusual aspects pertaining PXA started to be reported, such as atypical locations (especially in the posterior fossa and spine[4,8]) and an association with neurofibromatosis type I[9].

Uncommon histological aspects include descriptions of PXA in conjunction with ganglioglioma, dysembryoplastic neuroepithelial tumor, oligodendroglioma and even cortical dysplasia[10,11]. This finding led some authors to speculate about the histogenesis of PXA as deriving from bipotential precursor cells, in opposition to previous analyses that proposed mesenchymal precursors or a subclass of astrocytes as the tumor origin[12].

Prognosis seems to be worse than initial reports in a subset of patients. Although PXA is considered a WHO grade II tumor, anaplastic transformation can occur in up to 15 to 20% of

patients[4]. Histological signs of anaplasia can be observed in this regard, as manifested by increased mitotic activity, acquisition of necrosis and/or endothelial proliferation; however, sometimes recurrence is associated with tumors that remain histologically unchanged[4]. Leptomeningeal dissemination is usually related to malignant features of PXAs, although this is not always the case[13].

Gonçalves et al.[14], in the present issue of *Arquivos de Neuro-Psiquiatria*, shed light on some interesting topics related to histology and imaging aspects of PXA, in agreement with the literature and also adding local flavor to the study of these tumors.

First of all, in their sample they corroborate what literature states about the bad prognosis of PXA when detected in elderly patients, as their oldest patient deceased only one month after surgery. In this age range PXA usually show a more severe clinical picture, and more ominous histological features[15].

Another interesting topic was the association found between peritumoral edema and a short time interval (less than one year) between the beginning of the clinical picture and the diagnosis. This finding contrasts with most of the published data, as PXAs are commonly associated with long-standing symptoms. Although further studies are essential, as the authors underscore, this association deserves distinction.

It is also extraordinary that in four out of the nine presented cases there was an association between PXA and other tumors/cortical dysplasia; reinforcing what was already mentioned above and demonstrating the need of a detailed histological analysis.

Finally, when dealing with clinical, histological and imaging aspects of PXA, if there is a central characteristic common to all of them, it is pleomorphism, and therefore, one can really conclude: the key is in the name.

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