

A CASE REPORT: PLEOMORPHIC XANTHOASTROCYTOMA

OLGU SUNUMU: PLEOMORFİK KSANTOASTROSİTOM

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Abstract

Pleomorphic xanthoastrocytoma accounts for less than 1% of all astrocytic neoplasms and is typically encountered in children and young adults, in a superficial location in the cerebral hemispheres with involvement of the meninges. The authors reported a 9 year- old child who had long term seizures and showed a cystic mass with a mural nodule in the right temporal lobe on MRI scans. Microscopically the tumor consisted of highly pleomorphic bizarre astrocytic cells and ganglion cells with abundant amphophilic cytoplasm. Finally this case was reported as an instance of a pleomorphic xanthoastrocytoma (Pam Med J 2008;2(2):104-106).

Key words: Brain, tumor, Pleomorphic Xanthoastrocytoma, ganglion, child

Özet

Pleomorfik ksantoastrositom (PXA) tüm astrositik tümörlerin % 1'den azını oluşturan, tipik olarak çocuk ve genç erişkinlerde görülen, daha çok temporal lobda olmak üzere serebral hemisferlerde yerleşen ve meninkleri de tutan bir tümördür. Yazarlar, MR görüntülemeye temporal lob yerleşimli, mural nodülü olan kistik kitleye sahip, uzun süredir nöbetleri olan 9 yaşında bir çocuk sundu. Mikroskopik olarak; tümör oldukça pleomorfik, atipik astrositik hücreler ve geniş amfofilik sitoplazmaya sahip ganglion hücrelerinden oluşmaktaydı. Sonuç olarak bu olgu pleomorfik ksantoastrositom olarak rapor edildi (Pam Tıp Derg 2009;2(2):104-106).

Anahtar Kelimeler: Beyin, tümör, Pleomorfik Ksantoastrositom, ganglion, çocuk

Introduction

Pleomorphic xanthoastrocytoma (PXA) is an astrocytic neoplasm, typically encountered in children and young adults, with a superficial location in the cerebral hemispheres and involvement of the meninges [1]. Although there have been case reports on PXA of other sites (e.g. uncus, cerebellum) and some reports have an older age distribution [2,3]. Many patients have a long history of seizures (epilepsy is a common clinical feature) because of the superficial, cortical location. Sometimes PXA associates with cortical dysplasia. Cortical dysplasia associated tumor has a greater tendency to occur in older age [3]. Computed tomography (CT) and magnetic resonance imaging (MRI) scans usually outline the tumor mass and/ or its cyst [1,3]. Macroscopically, the tumor appears as a cyst, which has a mural nodule within the cyst wall. Microscopically, it is characterized by neoplastic astrocytes that show marked nuclear pleomorphism, rare to absent mitotic figures, nuclear pseudoinclusions, and variable lipidization [4]. Diagnostic hallmarks are lipid-laden pleomorphic cells, with frequent

bizarre multinucleated cells that express GFAP, a dense intercellular reticulin network, perivascular lymphocytic infiltrates and eosinophilic globular bodies. Recent studies have focused on the neuronal differentiation of PXAs [5]. Histologically typical PXA cells exhibit immunohistochemical markers of neuronal lineage, namely synaptophysin, neurofilament and MAP2 [5]. The complete excision of the tumor mass without adjuvant therapy is the preferred treatment and the majority of patients have a favourable prognosis [6].

In this report, we presented a new case of PXA that occurred in a 9-year-old boy, as a cystic mass with a mural nodule in the right temporal lobe.

Case Report

A 9-year- old boy presented with a history of headache and seizures. There were no specific findings on his medical and neurologic examination or laboratory data. On CT scan, he had a cystic mass, measuring 4x4 cm on right temporal lobe. MR imaging demonstrated a

cystic mass (5x5 cm) with a mural nodule (1x1) cm, that showed hypo-intensity on T1-weighted images and hyper-intensity on T2 weighted images (Figure 1). Minimal perifocal edema with mass effect was noted.

Macroscopically, the specimen consisted of gray-white soft tissue, measuring 4.5x2.5x1 cm which had focal hemorrhagic areas. Touch preparations showed eosinophilic granular bodies, pleomorphic multinucleated cells and spindle cells. Formalin-fixed, paraffin-embedded tissue blocks were sectioned at 5 mm for histochemical and immunohistochemical analysis. Sections were stained with hematoxylin-eosin, reticulin, PAS and diastase-PAS. Immunohistochemical staining using primary antisera against GFAP, synaptophysin, neurofilament protein, Ki-67, p-53 applied.

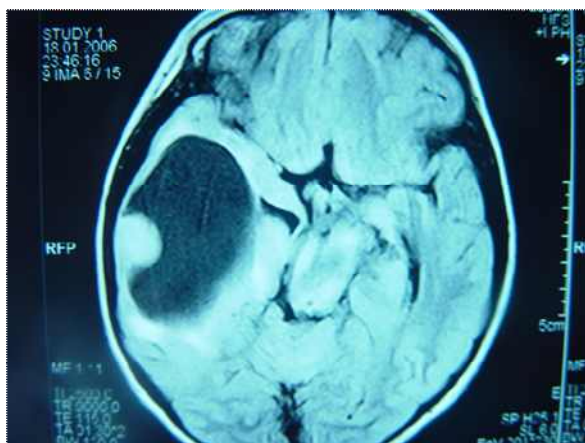


Figure 1. MR imaging demonstrates a cystic mass, measuring 5x5 cm with a mural nodule, measuring 1x1 cm

Haematoxylin-eosin sections showed neuroectodermal neoplasm, comprising an admixture of markedly pleomorphic, bizarre, hyperchromatic spindled glial cells and ganglion cells with abundant amphophilic cytoplasm. The lesion also contained many eosinophilic granular bodies. There was a prominent perivascular lymphocytic infiltration (Figure 2). Cortical brain tissue, which was not infiltrated by the tumor, was seen in only one area. There was no necrosis.

Mitotic activity was confined to the glial element, 2 per 10 high-power fields. Reticulin staining, surrounding individual tumor cells and cell groups, was prominent in focal areas (Figure 3). Immunohistochemically, the cells with ganglionic features showed staining for synaptophysin, whereas many of the spindled, pleomorphic cells were immunoreactive for GFAP (Figure 4a, b). Immunoreactivity for Ki-67 was 0.5 %, whereas

that for p53 was 1 %. PAS, d-PAS staining was positive in eosinophilic granular bodies.

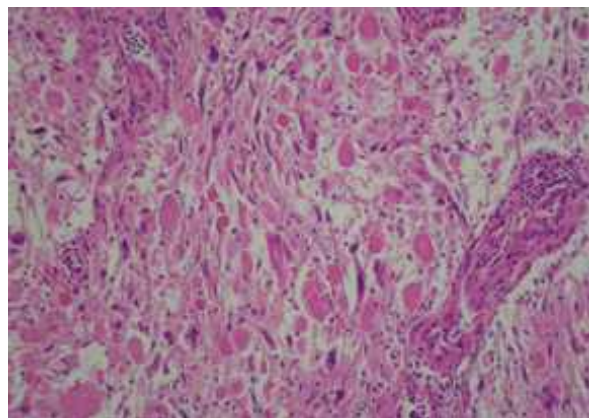


Figure 2. Pleomorphic cells, eosinophilic granular bodies and perivascular lymphocytic infiltration (H&E, X200)

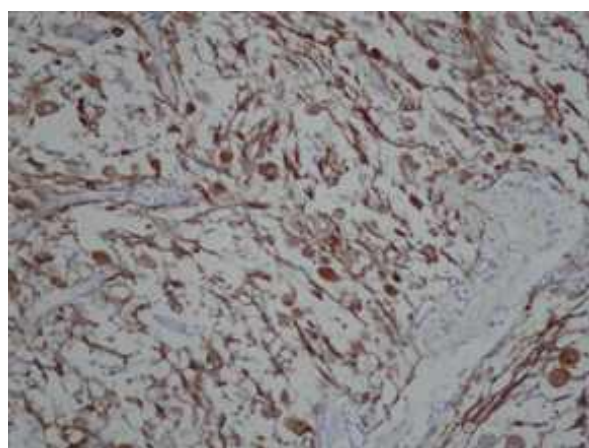


Figure 3. Dense reticulin staining, reticulin surrounds each cell or cell groups (Reticulin, X200)

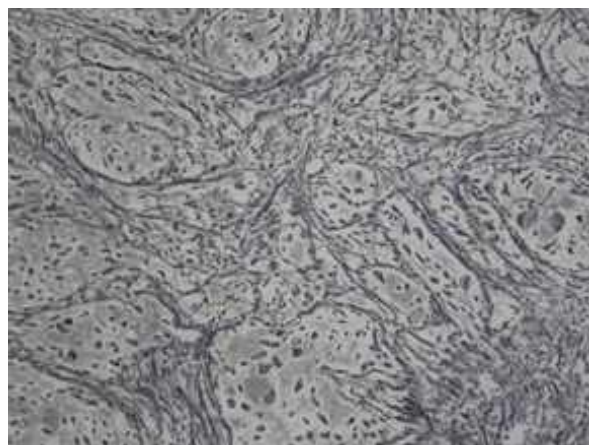


Figure 4. a: GFAP (+) pleomorphic astrocytes (X 100), b: Cytoplasmic staining of synaptophysin in ganglion cells (X100)

Discussion

PXA is a primary neoplasm of the central nervous system, originally described in 1979 by Kepes *et al* [6-9]. It is characterized by marked cellular pleomorphism, bizarre multi-nucleated

giant cells, prominent lipid droplets in the cytoplasm, frequent perivascular lymphocytic infiltration, a rich reticulin fiber network, and positive immunoreactivity for GFAP [5]. In many cases, its superficial location, relative circumscription, and cyst/mural nodule architecture facilitate a gross total removal [8]. Mitotic index emerged as the only histologic finding that, together with extent of surgical resection, could independently predict tumor recurrence and overall survival [8].

There is a relationship between PXA and neuronal/glioneuronal lesions. The association may take several forms. The first form includes the occurrence of a composite PXA-ganglioglioma in which the two neoplastic elements coexist with minimal intermingling. A second form consists of the finding of dysmorphic mono- or binucleate ganglion cells distributed individually within the substance of a PXA. The third form, is that of histologically typical PXA, the cells of which express synaptophysin or neurofilament immunoreactivity [1,9]. Coexpression of neuronal and glial markers by the same cell, although a rare event, is considered evidence of a hybrid phenotype, presumably a manifestation of truly divergent differentiation [7,10]. So, these data suggest glioneuronal differentiation of this tumor [4,7,10,11].

In the present case, the tumor was an admixture of markedly pleomorphic, bizarre, hyperchromatic spindled glial cells and ganglion cells with abundant amphophilic cytoplasm, which also contains many eosinophilic granular bodies and a prominent perivascular lymphocytic infiltration. There was no necrosis. Mitotic activity was confined to the glial element, 2 per 10 high-power fields. Reticulin staining, surrounding individual tumor cells and cell groups, was prominent in focal areas. Immunohistochemically, the cells with ganglionic features stained for synaptophysin, where as many of the spindled, pleomorphic cells were immunoreactive for GFAP. Finally we reported this case as a pleomorphic xanthoastrocytoma with ganglion cell differentiation.

PXA is a relatively indolent tumour with a prolonged clinical course, being designated as a WHO grade II tumour [1]. However, some cases may show more anaplastic features and aggressive clinical behaviour [8,11]. In the present case, during the 36 months follow up period, there was no recurrence.

If there is a cystic superficially located tumor on the cerebral hemispheres, we must consider PXA, although it is a rare entity. Typical histopathologic features of PXA help to accurate diagnosis of this tumor with detailed clinical data.

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