

Total removal of a rolandic pleomorphic xanthoastrocytoma in a child: Case report

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Introduction

The pleomorphic xanthoastrocytoma (PXA), described for the first time in 1979, is a rare tumor of the Central Nervous System, accounting for less than 1% of all astrocytic tumors^{1,2}. It mainly affects children and young adults, with peak of incidence in the second decade of life^{1,2}. The typical clinical presentation includes seizures, occasionally accompanied by headache and focal deficits^{1,2}. It presents a relative good prognosis, classified by the World Health Organization (WHO) as grade II astrocytic tumor, with median survival of 70% at 10 years after complete resection². In this paper we report a case of a 10-year-old girl, with clinical of Bravais-Jacksonian seizures due to PXA localization in primary motor area, which made resection a challenge.

Case report

A previously healthy 10-year-old girl was admitted to the Pediatric Emergency Care Service, after two episodes of Bravais-Jacksonian seizures with clonic activity in the left hemibody, lasting about 5 minutes, with spontaneous remission, and without loss of consciousness. Computed tomography (CT) was unconvincing. Magnetic resonance imaging (MRI) demonstrated a cystic lesion of 1.7 cm x 1.7 cm x 1.4 cm in the

right motor cortex, corresponding to rolandic region (Brodmann's area 4). The lesion showed a small perilesional vasogenic edema, subtle skull remodeling, hyposignal on T1 and hypersignal on T2 with contrast enhancement in the cyst wall (Figure 1). Surgical treatment was proposed. Under general anesthesia, a right parietal craniotomy was performed. After dural opening and cere-

bral exposition, infusion of propofol and neuromuscular blockade was suspended. Cortical stimulation mapping was performed to localize the motor area. An Ojemann Cortical Stimulator (Integra LifeSciences) was used, with 1-mm electrode tips 5 mm, delivering 60 Hz / 1 msec biphasic square wave pulses, with progressive increase of stimulation intensity from 2 mA to a maximal 10

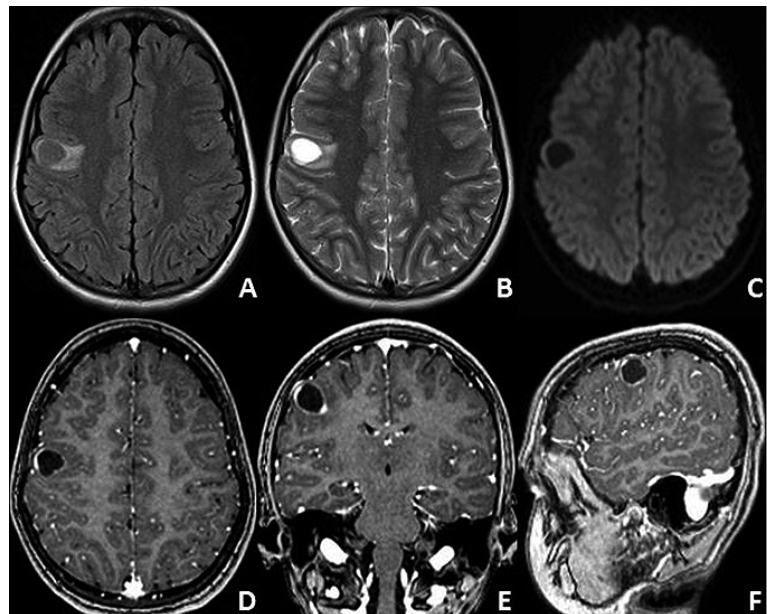


Figure 1. Preoperative MRI. **A:** Axial T2-Flair showing small perilesional edema; **B:** Axial T2 revealing cystic portion of the tumor; **C:** Axial Diffusion, with no water diffusion restriction; **D, E and F:** Axial, Coronal and Sagittal T1-contrast sequences.

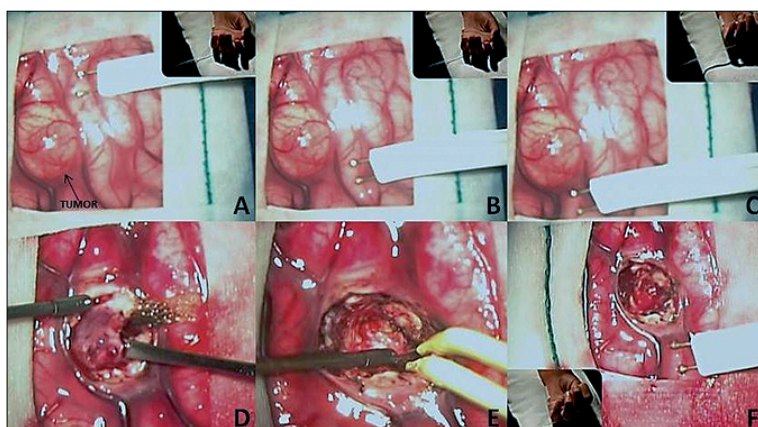


Figure 2. Intraoperative images. **A** and **B**: tumor localization in rolandic area, with no hand contraction under cortical stimulation; **C**: hand contraction under cortical stimulator, revealing precentral gyrus; **D** and **E**: total removal; **F**: preservation of hand contraction after resection.

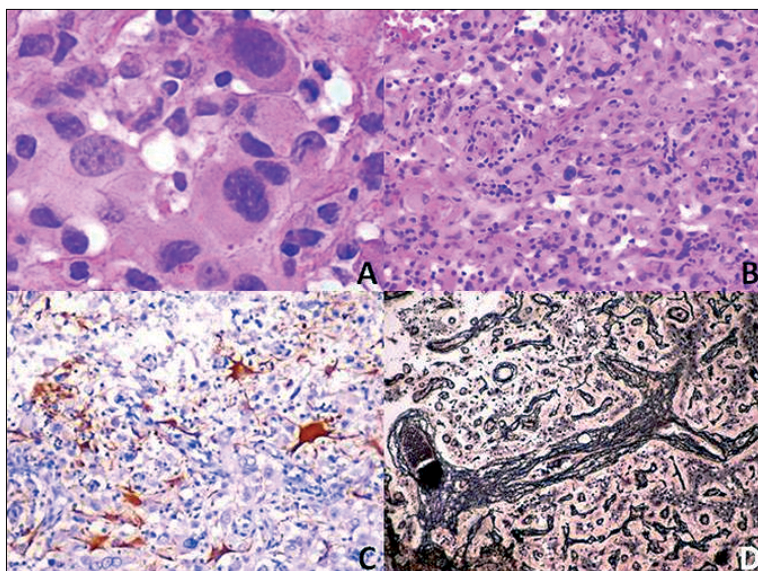


Figure 3. Histopathological images. The tumor revealed typical histological and immunohistochemical findings of PXA. **A**, **B** and **C**: presence of giant pleomorphic and spindle GFAP positive cells with intracytoplasmic fat; **D**: amid reticulin fibers accompanied by granular bodies.

or recurrence, with gradual removal of anticonvulsant medications, and MRI without image of recurrence (Figure 4).

Discussion

The PXA usually has a favorable prognosis, justifying the accurate diagnosis of this entity^{2,3}. Although the majority of the cases has a benign clinical course, there is the possibility of recurrence with malignant transformation^{3,4}. Typical histological characteristics were found in this case, such as spindle cells with ample eosinophilic cytoplasm and eccentric, large and pleomorphic nuclei, with intracytoplasmic fat (Figures 3A, 3B and 3C). The absence of necrosis and mitotic index less than 5 mitoses per 10 high power fields confirm the indolent nature of the PXA, as well as the negativity for the Ki-67 in immunohistochemistry. The presence of inflammatory infiltrates, as well as dense reticular network can be seen indicating significant desmoplastic reaction (Figure 3D). The neuroimage is characterized by solid-cystic pattern with contrast enhancement, by solid component, and cyst walls. On MRI, it presents hypointense on T1 and hyperintense on T2^{3,4}. It is proposed that the presence of perilesional edema, infratentorial location and diameter larger than 3 cm are markers of poor prognosis⁴. Calcification and extension to the skull are rare and suggest chronicity. This case presented a cystic lesion without signs of poor prognosis: 1.7 cm in diameter, little perilesional edema (< 1 cm) and chronicity signs as bone remodeling of the cranial vault (Figure 1). The complete tumor resection, including cystic wall, remains the treatment of choice^{3,4}. Adjuvant therapy with radiotherapy and chemotherapy still have not shown consistent data⁴. However, recent studies have presented reports of good response, especially for PXA with anaplastic features³. Also, *in vitro* sensitivity to some chemotherapeutic agents has been documented^{3,4}. Some studies suggest that adjuvant therapy may act to reduce the recurrence rate and may therefore be an option for recurrent, metastatic or residual tumors⁴. Still, the risk of inducing malignant transformation by radiation must be considered and faced with the possible recurrence protective effect^{2,3,4}. Concerning cortical stimulation in the anesthetized patient,

At this intensity it could realize left hand contraction (Figures 2A, 2B and 2C). After stimulation, the patient was submitted to microsurgery with total removal of the tumor (Figures 2D and 2E). At the end of microsurgery, a new cortical stimulation was performed, with hand contraction preserved (Figure 2F). The patient waked up from anesthesia without neurological deficits. A control CT was performed on the fifth postoperative day, without bleeding or abscess signs. The patient was discharged with anticonvulsant drugs

and general recommendations. Histopathological analysis revealed typical histological and immunohistochemical findings, such as the presence of giant pleomorphic and spindle GFAP positive cells and amid reticulin fibers, accompanied by granular bodies, allowed the diagnosis of pleomorphic xanthoastrocytoma without anaplastic features (Figure 3). Outpatient follow-up is being conducted with six monthly clinical examination and neuroimaging. After three years from the surgery, the patient remains without sequelae

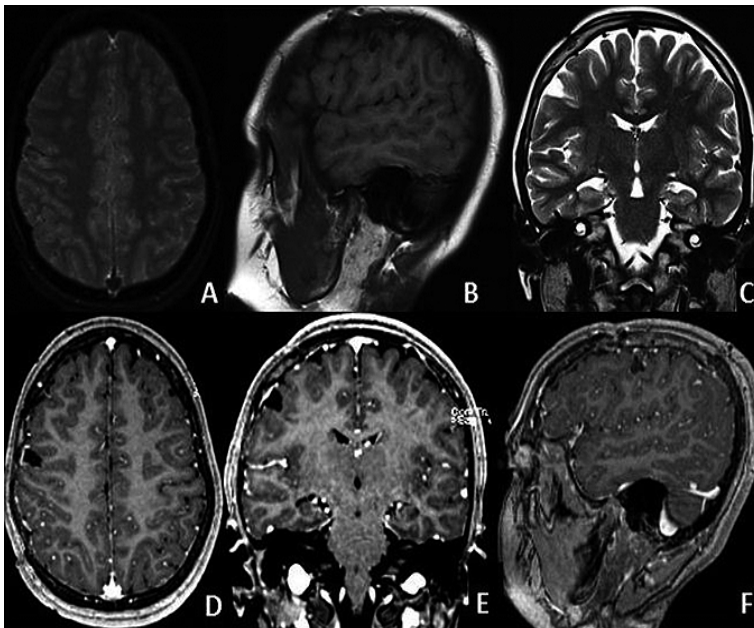


Figure 4. Postoperative MRI. Absence of recurrence after three years. **A** and **B**: Axial and Sagittal T1-no contrast; **C**: Coronal T2; **D**, **E** and **F**: Axial, Coronal and Sagittal T1-contrast.

the starting current is usually 4 mA and is increased at 2 mA intervals until an EMG or a motor response is visually identified in the left arm^{4,5}. It was stated that is necessary higher intensity stimulations, more than 12 mA, in

that patients with vasogenic edema and severe neurological deficit to obtain positive stimulation⁵. In this case the response has occurred with 10 mA. The preservation of neurological function is due to existence of multiple cortical

representations for hand and forearm movements in rolandic area⁵. There are two theories for this: or a secondary accessory eloquent cluster masked by the lesion, without any repercussion on the normal functioning of the primary cluster, which was detected by stimulation before resection; or the primary functional cluster was compensated by the secondary clusters recruited during the tumour growth, illustrating a local brain plasticity phenomenon⁵.

Conclusion

The PXA is usually a tumor of good prognosis. However, due to the possibility of malignant transformation, the detailed examination of histological and immunohistochemical characteristics is essential. Characteristics such as the presence of necrosis, cell proliferation index (Ki-67) and inflammatory infiltration should be insistently investigated. The importance of the extent of surgical resection, with complete removal of the lesion, should be highlighted.

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