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Medical Case Studies

Case Report

A case report of medulloblastoma in the cerebellopontine angle in 1-year old child

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Medulloblastomas (MB) are malignant embryonic neuroepithelial tumors of the cerebellum, accounting for 25% of primary tumors of the Central Nervous System of childhood, which predominantly grow in the cerebellar vermis and hardly in the cerebellopontine angle (CPA). The study aims to report the surgical management of a childhood medulloblastoma in CPA. The author reports the case of a 1-year-old male child who initially presented with right hemifacial weakness for 40 days. Magnetic resonance imaging (MRI) of the skull showed lesion in the cerebellar cistern. Subjected to surgical excision of the lesion with histopathological and immunohistochemical results of medulloblastoma. This neuroectodermal tumor localization is rare and aggressive, been an unusual differential diagnosis of lymphomas, schwannoma and meningioma. Thus, histopathology is the most reliable method for diagnosing the tumor.

Key words: Medulloblastoma, cerebellopontine angle, tumor.

INTRODUCTION

Medulloblastomas are common childhood tumors, especially in the first decade of life, representing 25% of all tumors of the Central Nervous System in this age group. Embryologically related with external granular layers of the cerebellar hemispheres, either with germinal cells remnants located at posterior medullary velum (Millard and De Braganca, 2016; Valtz et al., 1991). The proper tumor classification is due to embryonic and undifferentiated neuroepithelial cells from the cerebellum, especially the cerebellar vermis (Rutka and Hoffman, 1991; Jakacki, 2005; Gajjar et al., 2004).

In turn, medulloblastoma in CPA are uncommon, with rare reports in the literature, these mainly related to the adult population, representing up to 75% of the population group reaching the disease (Noiphithak et al., 2016; Faried et al., 2016). In adults, medulloblastomas is predominantly lateral located in the cerebellar hemispheres, in comparison with the axial position, with cystic formation and ill-defined margins, extending to the fourth ventricular foramen (Taylor et al., 2012; Helseth et al., 1999; Savardekar et al., 2012).

The diagnosis is made by neuroimaging exams and

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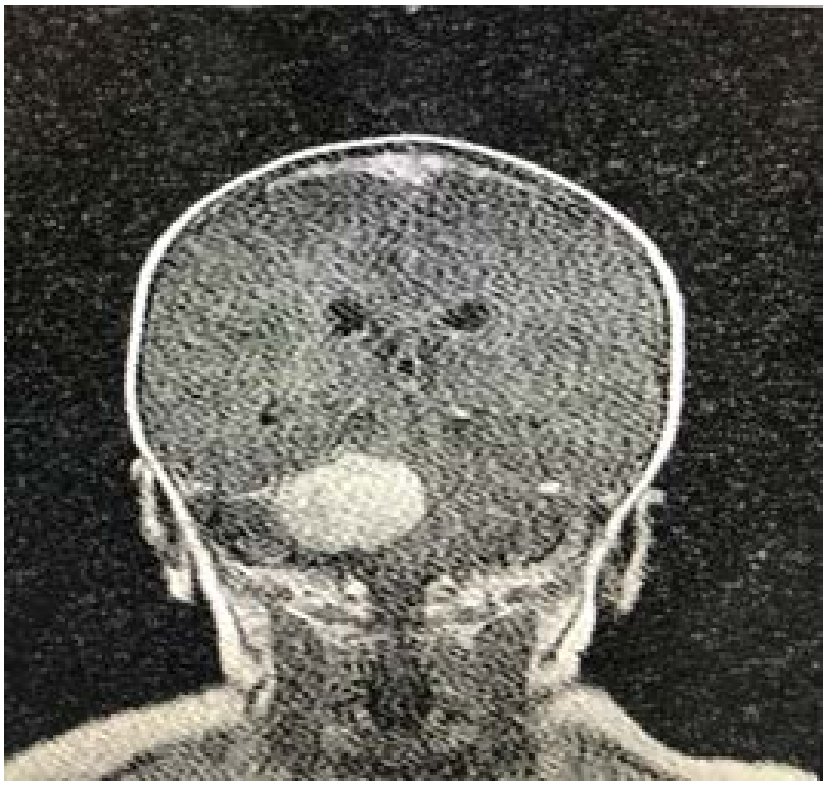

Figure 1. T1-weighted coronal section of magnetic resonance imaging showing a lesion with sharp limits, located in the right cerebellopontine angle that displaces and deforms the brain stem and the right cerebellar hemisphere.

confirmed by means of histopathology and immunohistochemistry techniques, presenting great difficulty in the neuroradiological differential diagnosis when compared with vestibular meningiomas and schwannomas. Treatment consists of surgical resection, when possible, followed by chemotherapy, however an early diagnosis is necessary for a better prognosis (Faried et al., 2016; Spina et al., 2013; Bhaskar et al., 2017). The present study aims to report a clinical case of medulloblastoma in the pontocerebellar angle and thus contribute to scientific knowledge on this topic.

MATERIALS AND METHODS

Report of a single case of a patient seen at the Regional Hospital of Baixo Amazonas Dr. Waldemar Penna, Santarém, Pará. The data were collected through the analysis of the patient's medical record, having access to the data contained therein such as medical developments, laboratory tests, exams imaging and histopathological.

CASE REPORT

Clinical history

A one-year-old male patient attended in a pediatric service with a history of facial weakness. After the initial analysis, he was referred to the oncology and neurosurgery services, being admitted for clinical follow-up. The patient evolved with progressive neurological disfunction, presenting Glasgow 15 with right hemifacial paresis, followed by cyanosis, mydriatic pupil and associated dysphagia.

Imaging, myelogram, histopathological and immunohistochemical exams

The initial approach involved the screening for neurological tumors and after the confirmation, was realized the tumor staging. Cerebral magnetic resonance imaging (Figures 1 and 2) was performed, showing a lesion located in the right cerebellar cistern, with displacement and deformation of the brain stem and the right cerebellar hemisphere. Figure 3 shows a penetrating lesion in the internal auditory canal, with full canal filling and dilation of the internal auditory canal. Magnetic resonance imaging of vertebral column showed no abnormality. The myelogram exam showed 37% of mature lymphocytes, 1% of monocytes and 6% of medium to large cells, with fine chromatin, Grumpetch's nuclear shadow, absent granules and high

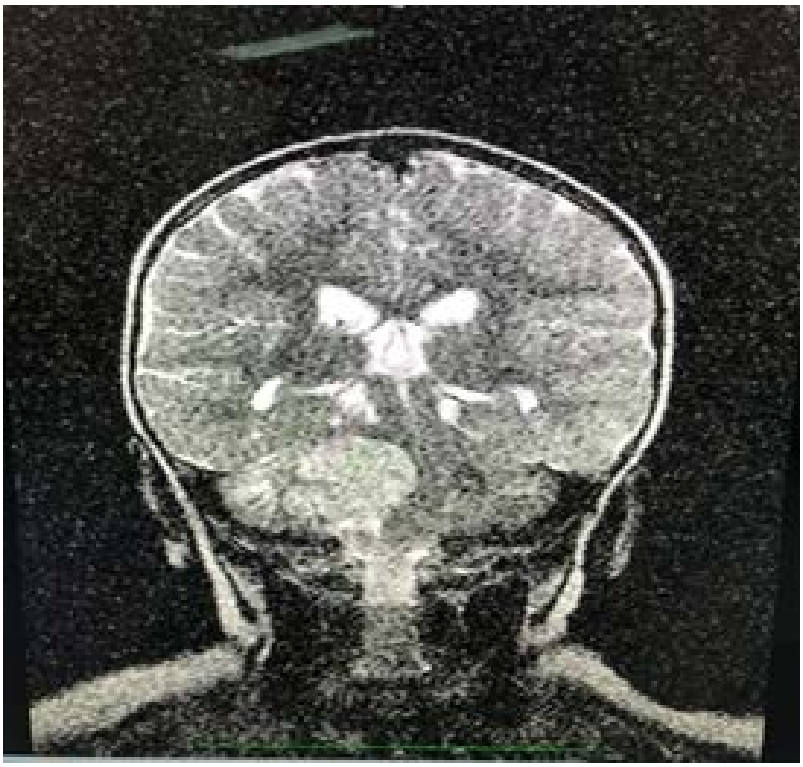

Figure 2. T2-weighted coronal section of magnetic resonance imaging showing a hypertense lesion in the region of the right cerebellopontine angle with a mass effect on the brainstem.

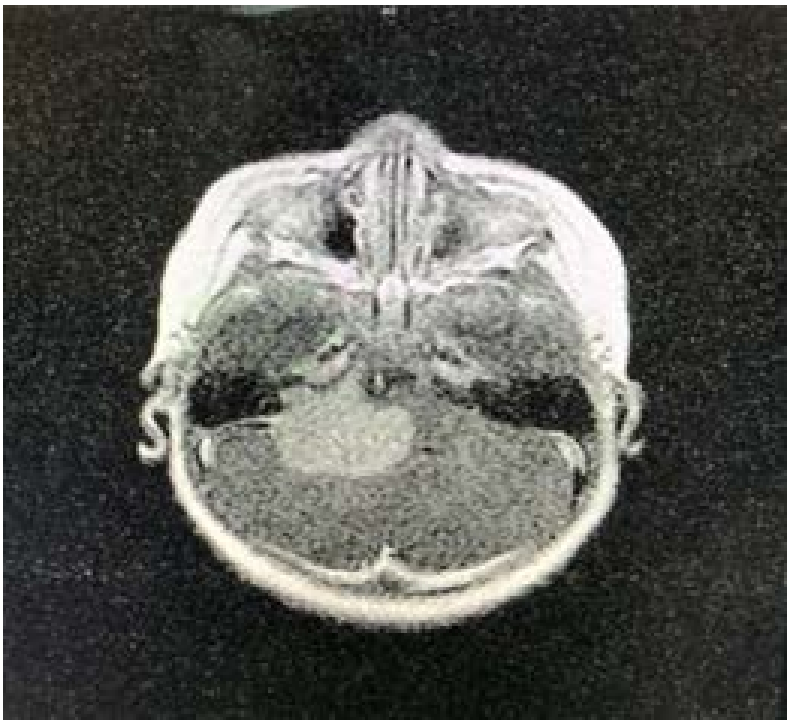

Figure 3. Axial magnetic resonance imaging, T1-weighted hypertense homogenous extra-axial image. The lesion penetrates the internal ear canal, filling it completely and dilating the internal auditory canal.

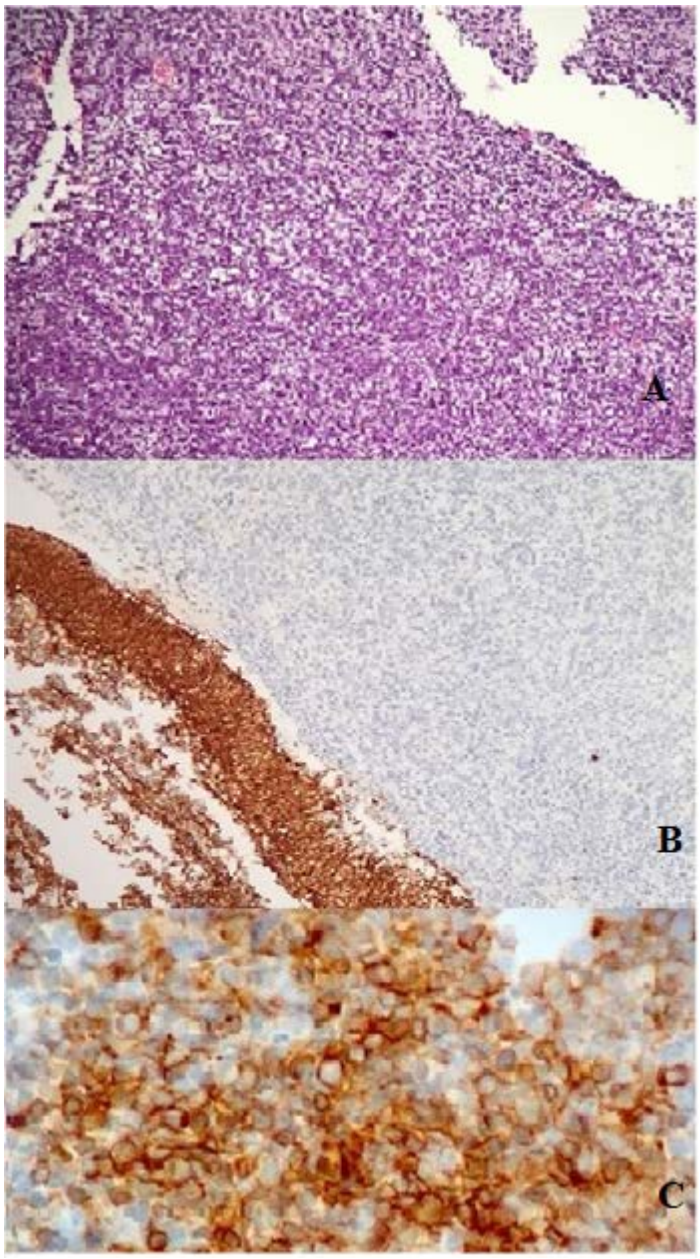

Figure 4. (A) Histopathological analysis - Increased parallel alignment of neoplastic nuclei in rows. (A) Histopathological analysis - Increased parallel alignment of neoplastic nuclei in rows "or rhythmic palisades". There are also Homer Wright's pseudorosettes and fat cells. (B) Immunohistochemistry analysis - GFAP positivity in tissue suggests malignant lesion. (C) Immunohistochemistry analysis - CD99 staining is predominantly noticed on cell membranes and here we see positivity in different sections of the tumor suggesting medulloblastoma.

nuclecytoplasmatic ratio.

The histopathological result (Figure 4A) showed tumor cells arranged in Homer Wright's pseudorosettes, morphologically classified as classic medulloblastoma. Which are dense cellular neoplasms, consisting of small, oval cells with a high nucleus/cytoplasm ratio. Immunohistochemistry (Figure 4B and C) showed positivity for glial fibrillary acid protein (GFAP) and CD99. In addition to positivity for synaptophysin, beta catenin, and KI-67, genetically classified as WNT-activated

medulloblastoma. Using histopathology and immunohistochemistry information, the diagnosis of medulloblastoma was established, based on guidelines (Pietsch and Haberler, 2016).

Therapeutic conduct and outcome

After diagnosis, the patient underwent tumor resection at a right cerebellar point angle, by retrosigmoid approach, without complications and no need of ventricular shunt technique. Surgical procedure was subsequently accompanied by an oncological therapeutic plan based on three cycles of chemotherapy, with methotrexate, vincristine and cyclophosphamide. However, after 21 days of the surgical procedure, the patient progressed to a coma, followed by multiple organ failure related with neurogenic shock, until the diagnosis of brain death was confirmed.

DISCUSSION

MB is a highly malignant and aggressive neuroectodermal tumor of cerebellar origin, densely cellular and distinct from sarcomas and other gliomas. The current literature considers it as a common pediatric tumor of the posterior fossa representing 20-25% of all pediatric tumors, which develop in the cerebellar vermis and in the fourth ventricle. It can be found in the midline, in the cerebellar hemisphere or in the CPA. In adults and children, the most common location is the cerebellar hemisphere, with CPA being an extremely rare variant (Spina et al., 2013; Batista et al., 2017).

The epidemiological profile of the tumor, considering the higher prevalence in the first decade of life, is explained by its embryonic characteristic. Clusters of neuroepithelial cells from the roof of the fourth ventricle can migrate to make the composition of the outer granular layer of the cerebellum. Migratory cells can remain resident until the first year of age, being related to the appearance of medulloblastomas in any part of the migratory path of these cells (Rubinstein, 1972; Zimmerman et al., 1978). The rare location in CPA makes MB an unusual differential diagnosis of tumors that affect the appropriate area, with the most frequent differentials in relation to MB: Lymphomas, schwannoma, meningioma, primary cholesteatomas and epidermoid tumors. Vestibular schwannomas and meningiomas are the two most frequent lesions and represent 90% of all tumors in CPA (Batista et al., 2017)

Imaging findings have variable and non-specific characteristics, making it difficult to differentiate, giving preference to histopathological and immunohistochemical diagnosis. The appearance of medulloblastoma on magnetic resonance imaging usually manifests itself with hypo-intensity on T1-weighted images and heterogeneous hyperintensity on T2-weighted images and improves with contrasts (Faried et al., 2016; Bhaskar et al., 2017; Batista et al., 2017; Nyanaveelan et al., 2007). Histopathologically, classic medulloblastoma contains small individual cells with scarce cytoplasm and hyperchromatic nuclei that are often elongated. The tumor may express neurosecretory granules or Homer Wright rosettes. The characteristic immunohistochemistry reveals positivity for synaptophysin with variable reactivity to GFAP, β-catenin, membrane epithelial antigen and cytokeratin (Pietsch and Haberler, 2016).

In our report, the results showed tumor reactivity to synaptophysin and GFAP. Synaptophysin is an integral membrane glycoprotein from presynaptic vesicles of neuronal and neuroendocrine cells that has been identified as a reliable marker for neuronal differentiation, which is commonly seen in medulloblastoma. GFAP is an intermediate astroglial cytoskeletal protein that is commonly expressed in neural stem cells (Noiphithak et al., 2016; Faried et al., 2016; Bhaskar et al., 2017). The preoperative clinical history showed an atypical symptom of right hemifacial weakness and absence of classical symptoms. The literature shows classic symptoms of MB as irritability, lethargy, nausea and vomiting, morning headaches, anorexia, behavioral changes and seizures. The finding is important, since demonstrates the atypical presentation of the clinical case reported (Millard and De Braganca, 2016).

As for treatment, total removal with adjuvant radiotherapy is commonly performed as therapy, when age and other prerequisites permits. Chemotherapy has also been combined in some cases, as the residual tumor is associated with worse progression. However, Pant et al. (2016) states that aggressive brain tumor removal is not recommended, since a high rate of postoperative complications have been reported after tumor resection. Thus, the outcome of MB treatment in the CPA is not well established due to limited reports, because despite the 60 to 70% increase in survival, attributed to improvements in surgical techniques and adjuvant therapies, more cases are still needed to confirm these discoveries (Noiphithak et al., 2016; Spina et al., 2013).

CONFLICT OF INTERESTS

The authors have not declared any conflict of interests.

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