Medulloblastoma in adults, regarding a case

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ABSTRACT

Posterior fossa tumors are uncommon in adults: they represent 15% or 20% of all brain tumors. Medulloblastoma, which corresponds to this classification, occurs in less than 1% of cases, unlike pediatric population, in which it is the most common malignant brain tumor. The most frequent medulloblastoma sub-type in adults is the desmoplastic, whose characteristics are correlated to the findings in the imaging studies. We present the case of a female patient with a posterior fossa tumor whose anatomopathological result was medulloblastoma.

Key words: Acute intermittent porphyria, Porphobilinogen.

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INTRODUCTION

Medulloblastoma is the most common malignant brain tumor in pediatric patients[1,2] and accounts for 35-40% of the types of posterior fossa tumors in children between 3 and 9 years of age[3,4], peaking at 4 years old, although some cases have also been reported since birth[5]. Medulloblastomas may also appear in adulthood. However, at this age they only account for 0.4-3% of primary tumors in the central nervous system (CNS)[4,6]. The cases described have been diagnosed between 20 and 24 years old or in the third and fourth decade of life[2].

Medulloblastomas have neuroectodermal origin, and are considered by the World Health Organization (WHO) as malignant embryonal tumors and invasive of the cerebellum, with high histology grade (aggressive nature, malignant cytology, rapid evolution and high mortality if not treated properly)[1], with fast dissemination through the cerebrospinal fluid (CSF) and, unlike low-grade tumors, they have a high heterogeneous component, which explains why the ADC diffusion analysis does not always values them adequately (it is described that they reach low values)[3,7]. Even then, in magnetic resonance (MR) studies the heterogeneous appearance of the tumor is pronounced. Next, we present a case of medulloblastoma in an adult patient.

CLINICAL CASE

A 23-year-old woman with approximately one month of disease, during which she had a pulsating headache in the occipital region, of moderate intensity and progressive course, associated to occasional dizziness and nausea, was admitted in emergency because of the increased intensity of her headache and nausea, in addition to vomits up to three times. In the physical examination, there was slight instability when walking and, in the Sensitized Romberg Test, a slight pulsation to the right. The rest of the physical examination was normal.

Finding in images:

The brain scan (BS) (Figure 1) reported an expansive lesion in the posterior fossa, of 4 cm, located in right paravermal area, with bilobed aspect of well defined borders, heterogeneous structure and central areas of cystic or necrotic appearance. There was also a slight dilation of the third ventricle and of the lateral ventricles with hypertensive signs due to a compressive effect on the fourth ventricle and the Sylvian aqueduct. No transthoracic extension was observed and the angiographic study was normal.

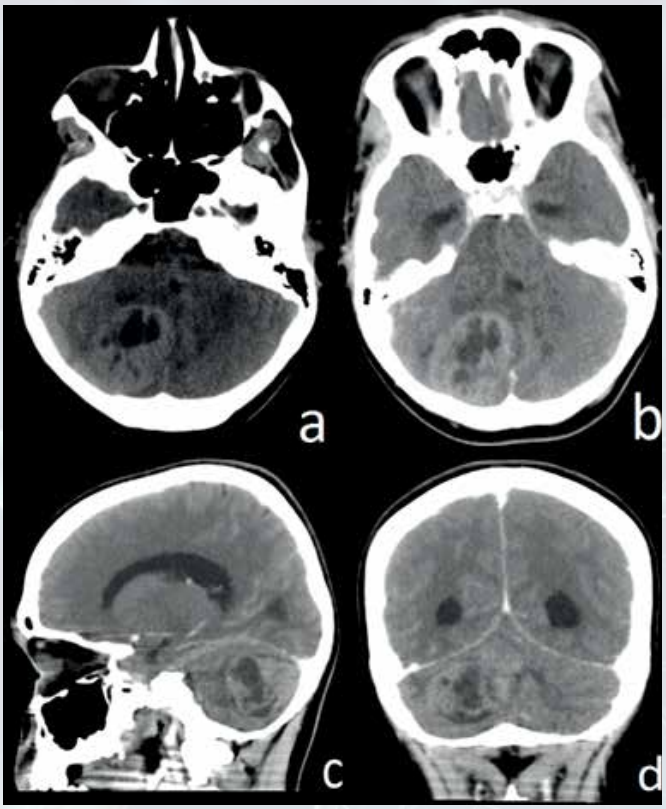

Figure 1. Axial brain CT without crontrast (a) and with contrast (b), sagittal (c) and coronal (d) with contrast. Intra-axial tumor in the right cerebellum hemisphere, with well-defined borders, heterogeneous appearance, with central cystic area (necrotic) and contrast capture.

In the brain magnetic resonance imaging (Figure 2), there was a neoformative process, located in the cerebellar hemisphere on the right side at the paravermal level, with a size of 4.7 cm. It was found that the lesion presented bilobed configuration with well defined borders, a solid nodule in cephalic projection and a basal component with a vast areas of necrosis. On T1-weighted images, it was hypo-intense, but hyper-intense in T2-weighted sequences.

The capture of the contrast substance was tenuous and heterogeneous. It was associated to limited surrounding edema of vasogenic type, with mass effect and midline displacement, causing deformation of the fourth ventricle and mild dilatation of the third ventricle and lateral ventricles accompanied by mild subependymal edema. No supratentorial extension was detected.

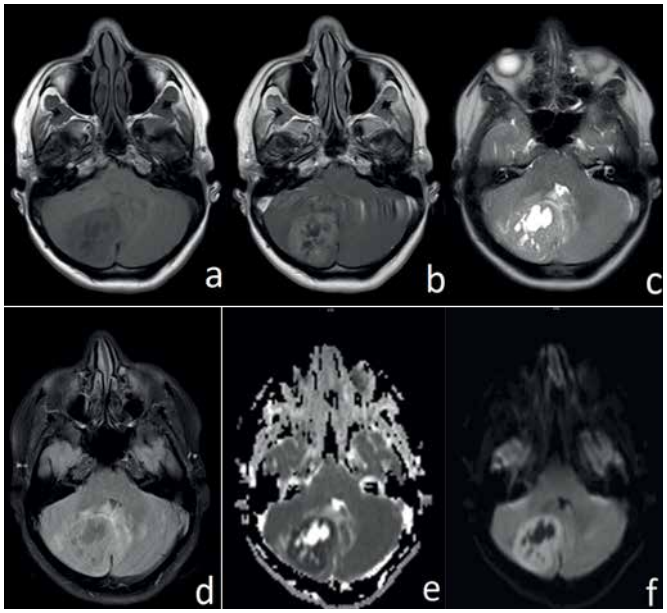

Figure 2. Brain RM. Neo-formative mass of well-defined borders in the right cerebellar hemisphere with midline movement, hypo-intense in sequence T1 (a), which enhances with the contrast substance (b). Slightly hyper-intense in T2-sequence (c), showing vasogenic peritumoral edema (FLAIR) (d) and central hyper-intensity in relation to tumor necrosis. It presents hypo-intensity in ADC (e) and restricts the dissemination (f).

Diffusion restriction was found. In the spectroscopy (Figure 3), inversion of peaks and values of Choline (CHO) and N-acetyl aspartate (NAA) were identified, indicating high cellularity of the lesion. The anatomopathological diagnosis was nodular desmoplastic medulloblastoma and is detailed in Table 1.

The patient was subjected to a surgical procedure, initially for the placement of a peritoneal ventricle bypass system and, then, for removing the lesion, which was achieved after two surgeries. After the treatment, 32 sessions of radiotherapy and neo-adjuvant chemotherapy were performed. The last control, 18 months after the surgery, showed no recurrence or distant metastases, and, clinically, there is no focalization or residual neurological alteration.

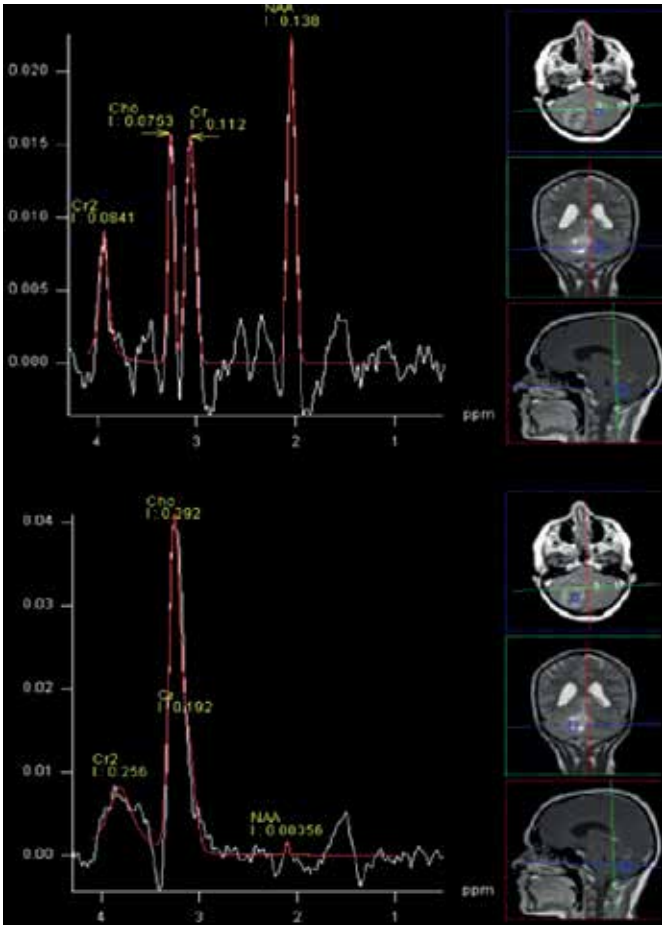

Figure 3. Curve of spectroscopy. Curve of spectroscopy in normal brain tissue (left picture) and curve of spectroscopy on tumor lesion of the right brain hemisphere (right picture), which displays the cholina peak (CHO), and a decrease in the N-acetyl aspartate and creatine.

Tabla 1. Reporte de anatomía patológica

Diagnóstico anatomopatológico
Macroscopía:
Formación nodular, de tejido de color blanquecino, de consistencia blanda, de 4 cm × 3 cm de diámetro. Al corte tejido sólido.
Meduloblastoma desmoplásico nodular. Grado histológico (OMS): IV
- Inmunohistoquímica:
Sinaptofisina: Positivo
PGAF : Negativo
Ki67 : 70%
- Histoquímica:
Reticulina : Patrón nodular

DISCUSSION

Posterior fossa tumors are less frequent in adults than in children and represent only 15 to 20% of all brain tumors[8].

There are many types of tumors that can affect the posterior fossa, but this list of differential diagnoses can be reduced by determining the anatomical location and the imaging characteristics of these injuries[9]. The initial fundamental analysis involves establishing the extra-axial location (more frequent in posterior fossa tumors) or intra-axial location. In this clinical case, with the images obtained by BS (Figure 1), we can establish the intra-axial origin of the mass which affects the right cerebellar hemisphere. The differential diagnosis, from this point on, includes some lesions such as metastases representing the most frequent cerebellar masses in adults. Hemangioblastoma is the most common primary cerebellar tumor in adults (occurs more frequently as a cystic lesion with a highly vascular mural nodule), as well as the cerebellar astrocytoma.

The symptoms among posterior fossa tumors are similar, since they occur as a consequence of the expansive effect of the tumor, moving the structures of this region (brain stem, cerebellum, fourth ventricle) and giving rise to dysfunction of the brain stem or cerebellum, which is often associated to hydrocephalus due to obstruction of cerebrospinal fluid flow in the fourth ventricle[10]. These characteristics associated to rapid growth typically lead patients suffering from a medulloblastoma to present headache, nausea, vomits -manifestations that reflect the increased intracranial pressure-and, in some cases, ataxia.

In this case, the symptoms presented a progressive course of a month of evolution approximately, which is compatible with those previously reported[11].

Other symptoms and signs reported and that the patient didn´t present were seizures (many times associated to metastatic spread), ataxia associated to spasticity, papilla edema, nystagmus, disdiadococinesia, ataxia of a limb and acute neurological deterioration due to secondary hemorrhage for medulloblastoma.

Medulloblastoma in children has been associated to some syndromes such as familial adenomatous polyposism, Turcot syndrome, Li-Fraumeni syndrome, neurofibromatosis type 1 and 2, Rubinstein-Taybi syndrome, Fanconi anemia, and Nijmegen syndrome[12].

Occasionally, it has been reported in children with other genetic disorders, such as Gorlin syndrome. There are many genes involved, including the gene PTCH1 and SMO. It has also been reported its association to second malignancies induced by treatment with radiotherapy[13].

Histologically, the medulloblastoma is classified into four sub-types: classic, desmoplastic/nodular, with extensive nodularity and large cells/anaplastic[1,12]. It has been found that the prognosis of medulloblastoma is associated to its histological type. The anaplastic one and that one of large cells have been associated to high risk of relapse and poor response to treatment[1]. The sub-types of better prognosis are the extensive nodular and the desmoplastic/nodular. Likewise, the histological subtype has been correlated to some features in IRM, for example, the ring enhancement has been associated to the anaplastic and large cell sub-type[1].

Medulloblastoma appears, in imaging studies, as a rounded or ovoid tumor, with circumscribed margins. In adults, it is mostly located in the cerebellar hemisphere (in children, it frequently affects the cerebellar vermis)[4,14]. Both characteristics are identified in the case of our patient (Figures 1 and 2). It has been previously reported that findings in imaging studies (BS and RM) are more variable in adult cases than in children[6]. The BS allows an initial evaluation by establishing the presence of the lesion in 95% of the cases[10]. However, given that the posterior fossa generates a lot of "noise" by the *beam hardening* artifact[15], in some cases its use is limited. As it was found in our clinical case, medulloblastoma in BS is presented as a hyperdense lesion in relation to the adjacent gray substance, which is related to the histological composition of the tumor that is characterized by densely grouped round cells. Medulloblastoma presents enhancement after the injection of contrast substance, although in lower degree in adults compared to children. This has been linked to high amounts of reticulin fibers in the stroma of the tumor, especially in the desmoplastic sub-type, which usually occurs in adults. It can also be evaluated the presence of peritumoral edema.

In a high percentage of cases, lesions present low-density areas, related to cysts or intra-tumor necrosis, as observed in our case[14,1,6].

In MR, it has been found that medulloblastoma is somewhat variable[6]. On T1 and T2-weighted images, we can find intensity and variable capture of the contrast (usually hypo-intense in T1 and iso or hyper-intense in T2). Cystic or necrotic areas are easily detected in T2 sequences because of its hyper-intensity[6]. Areas of hemorrhage or calcification and leptomeningeal infiltration could be found1. They restrict dissemination in MR studies (differentiating medulloblastomas from other posterior fossa tumors)[1,3].

The prognosis is related to the percentage of tumor removed (the higher tumor tissue, the better prognosis), the age of the patient (the lower age, the worse prognosis) and the dissemination at the time of diagnosis[16]. Survival in previous reports is around 81% at 5 years old and 62% at 10 years old[17].

GRANTS OR FINANCIAL SOURCES
Nothing.

CONFLICTS OF INTEREST
The authors do not report conflicts of interest regarding the present manuscript.

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