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Original Research Paper

Medulloblastoma in Children About A Series of 70 Cases.

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ABSTRACT:
Medulloblastoma is the most common malignant tumor in children. This is a retrospective study of 70 cases of medulloblastoma operated on at the EHS Ali Ait Idir between 2011 and 2017, the study parameters of which relate to the age group, sex, clinical manifestations and radiological explorations. and the therapeutic management as well as the results of the anatomopathological examination.

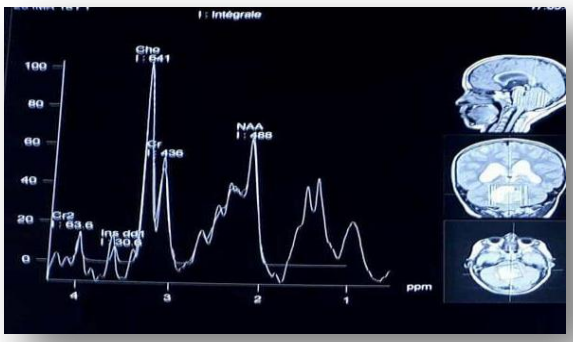
Keywords: Medulloblastoma, malignant tumor, pathological examination.

INTRODUCTION:

Medulloblastoma (MB), a malignant embryonal tumor of the cerebellum and the most common malignant brain tumor in children. The term "Medulloblastoma was proposed by Bailey and Cushing in 1925, it occurs at all ages, It presents a bimodal peak between 3 years and 4 years and between 8 years and 10 years, but it can occur throughout childhood., representing 20 to 25% of pediatric tumors and 30% of cranial tumors of the posterior fossa. John J. Gregory, Jr., MD, Rutgers, New Jersey Medical School. Last fully revised August 20, but medulloblastoma can occur as part of certain syndromes (eg, Gorlin syndrome, Turcot syndrome

MATERIALS AND METHODS:

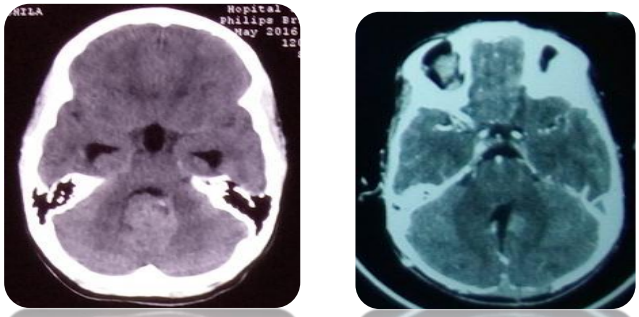
We report a series of 70 cases of medulloblastoma operated on in our department at E.H. S Ali Ait Idir between 2011 and 2017: this is a retrospective study of 21 infant cases between (0-3 years); 6 cases between (3-5 years); 43 cases between (5-15 years old) including 43 boys and 27 girls. Clinically, the syndrome of intracranial hypertension secondary to hydrocephalus is found in all cases. The explorations by CT () and brain and spinal MRI which made it possible to clarify the location of the tumor, its site and its relationships, Brain CT and preoperative brain and spinal MRI were used in a complementary manner in all our patients. Medulloblastoma has a high power of dissemination. Which makes exploration of the spinal axis obligatory. It is preferable to perform a spinal MRI preoperatively to avoid artifacts that may result from surgery. Spectroscopy, a spectroscopic assessment was carried out in 04 patients showing an increase in choline N-acetylaspartate but providing no additional benefit in the management of medulloblastoma.



Spectroscopy a spectroscopic assessment in a patient showing an increase in choline N-acetylaspartate

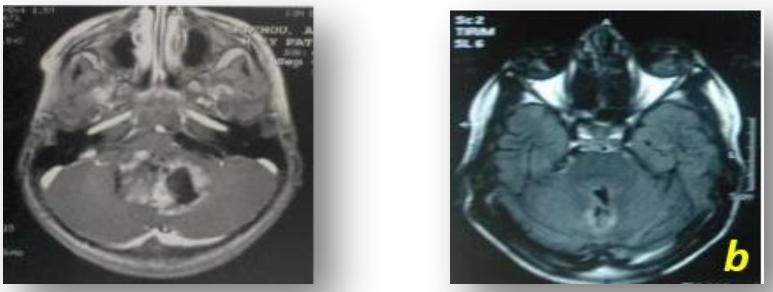
It is important to improve intracranial hypertension, the comfort of surgical excision and the immediate postoperative period.
All our patients benefited from a ventriculoperitoneal diversion. And operated via the median suboccipital route. There are two most common surgical approaches: the Transvermal and Telovelar approaches. The choice of one or the other will depend

on the tumor volume, the extension and above all the habits of the surgeon. The goal of this Telo velar approach [170,157] is to access the fourth ventricle by opening the tela choroid and the lower medullary velum, the two thin plates of tissue which form the lower half of the roof of the fourth ventricle are incised. We also found that opening the tela choroid alone would provide adequate ventricular exposure, in most cases, without incising the vermis.



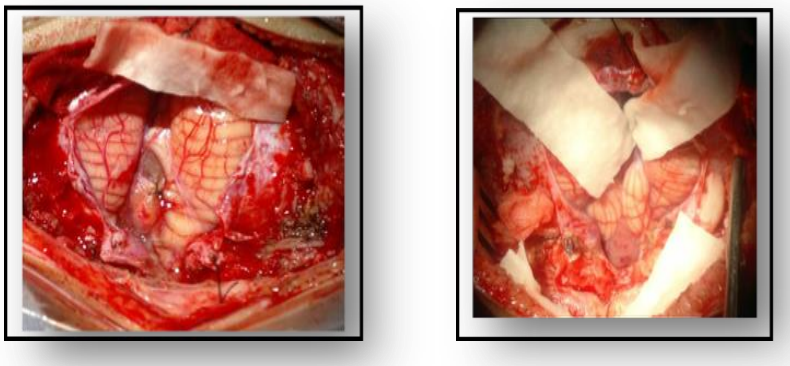
(a) (b)

Ct scanner: Vermien medulloblastoma (a) pre-operatoire (b) post-operatoire



A B

T1 brain MRI with Gado (a) preoperative (b) postoperative Remnant <1.5 cm



A B

(a) Trans-vermian approach (b) telovelar approach.

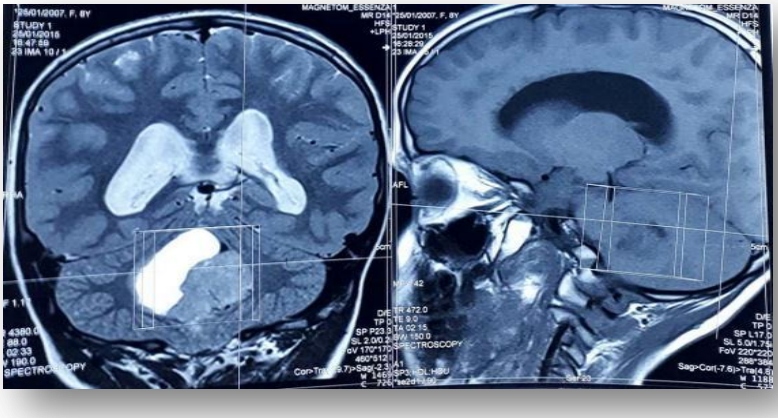
RESULT:

The anatomo-pathological study was conclusive in all operated cases and provided the following results;

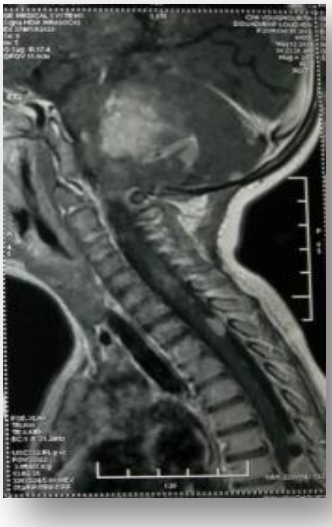
- Classic medulloblastomas 42 cases - Desmoplastic medulloblastomas (extensive nodularity): 22 cases
- Anaplastic form 4 cases
- Large cell form 2 cases.

Overall, standard risk medulloblastomas are classic and/or desmoplastic medulloblastomas - total surgical excision (absence of visible remnant on early postoperative imaging) or subtotal (incomplete excision (for example a tumor partially inserted at the level of the floor of the IVth ventricle) or a surface area of the tumor residue less than 1.5 cm2). and not metastatic; those at high risk are anaplastic/large cell medulloblastomas or those with incomplete excision or with metastases: presence of a tumor remnant whose

surface area is greater than 1.5 cm2 or metastatic medulloblastoma, from M1 to M4.



Pre and post-operative brain MRI, presence of tumor remnant.



Spinal Cord MRI:

Spinal cord metastasis. Infants are generally defined as high risk group up to age 3 years. [59]

This classification was applied for the 70 patients. The 06 patients who died postoperatively could not be classified.

the search for tumor cells in the cerebrospinal fluid (CSF) was carried out by lumbar puncture (LP) in the second postoperative week in 22 cases, the PL could not be carried out due to postoperative deaths in 6 cases, by elsewhere in 11 cases due to the absence of reagents and these patients were classified as high grade. The initial extension of the disease, local and metastatic, was classified according to the T stage of the Chang classification and the Histological type according to the classification into risk groups recognized by the International Society of Pediatric Oncology: standard and high risk. risk . (non-metastatic disease and incomplete excision, histological type: large cell or/anaplastic), and high metastatic risk (metastatic disease, excision of the primary tumor complete or not), the quality of the excision being defined according to the account operative rendering and early postoperative imaging.

Standard risk 30 cases, High risk 34 cases.

The frequency of the combined LC/A form varies from 10% to 22% when the anaplastic MB is (more than 50%). Nodular/desmoplastic medulloblastoma and MBEN account for 7% and 3% of all medulloblastomas. Classic tumors constitute the remaining large cell/anaplastic LC/A. Mortality The operative mortality rate (D0 – D 30) remains high in our series compared to the literature series Bartelet 2013 1%, Motollese 2011 1 percent in our series 8 percent

but with the precarious neurological state of certain patients preoperatively.

In our 2011-2017 series a period of 7 years is obtained. A median survival at 3 years is 78% and 62% at 5 years.

Until February 2018, we observed a median survival of around 51% at around 8 years. The treatment of recurrences is not standardized. Their prognosis remains severe. Recurrence in high-risk patients is more likely during the first 2 years (85%) than in

average-risk patients (65%) (Packer et al., 2008). The rate of surgical revision at the level of the posterior cerebral fossa represents 7% (i.e. 5 patients). We note that recidivism is more significant in the age group of 3 to 4 years.

DISCUSSION:

The clinical diagnosis is most often suggestive but not specific for medulloblastoma but for all tumors of the posterior cerebral fossa. On the other hand, the pre- and post-operative radiological assessment allows the classification of medulloblastoma into T and M; T representing the size of the tumor and local extension (T1 to T4) and M determining the presence of fluid metastasis (classification in 5 stages). On the radiological level there is a Radiology-anapath correlation Prediction of the molecular subgroup of imaging the vast majority (94%) of medulloblastomas appear in the cerebellum and the majority of these, from the vermis (75%).

This pattern is particularly common in group 3 and group 4 and in some tumors of the SHH subgroup.

- cerebellar peduncle; most likely WNT subgroup and therefore better prognosis.
- cerebellar hemisphere; most likely a SHH subgroup and therefore an intermediate prognosis Protoplastic/nodular/medulloblastoma with extensive nodularity (MBEN). Medulloblastomas are classified according to the WHO 2016 Classification into: Classic, desmoplastic (extensive nodularity), anaplastic and large cell medulloblastomas

In recent years, several teams have proposed convergent biological classifications, which has led them to a consensus allowing them to be divided into 4 groups:

- Sonic hedgehog (SHH) signaling. (25% of MB)
- WNT group The prognosis of tumors in this group is excellent (EFS of 90% at 5 years).[69,89,59] 10% of MB) • Group 3: represents approximately 30% of MB. • Group 4: which represents 30%.

Furthermore, whether in our series or the different series in the literature, the indication for biopsy remains very low because total excision is the only guarantee of a better median survival with preserved quality of life. In the prospective study our median survival at 3 years is 78% and 62% at 5 years, it remains lower than that of Western series which is around 85% at 3 years and 70 to 75%. at 5.

CONCLUSION:

The treatment of medulloblastoma has continued to advance. This progression or this evolution has occurred with a better knowledge of the disease on the one hand and on the other hand thanks to progress in imaging, surgical technique, anatomo-biological study and complementary treatment (chemotherapy, radiotherapy). The prognostic criteria for medulloblastomas are those obtained at the end of the clinical, radiological and anatomo-biological assessment.

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