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Desmoplastic Infantil Ganglioglioma: A Case Report

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Abstract **Case Report**

Infantile GGDs are a group of rare neuroepithelial tumors that are benign to the small child, most often occurring in the first two years of life. In magnetic resonance imaging (MRI), it appears as a large supratentorial tumor with a double component: solid, with a broad base of dural; and central cystic implantation, whose wall, typically, does not improve after injection of the contrast product. Edema is usually absent, and when it exists, it is often unimportant with a low mass effect compared to tumor volume. The histological study finds a mixed cell population, with neuronal differentiation. Total resection, if possible, is the treatment of choice, and adjuvant treatment may not be necessary beyond surgery. Despite morphological and radiological similarity to aggressive tumors, GGDs have a good prognosis. We present an observation of a child with GGD and we review the anatomo-clinical, radiological and therapeutic characteristics of this group of very rare tumors.

Keywords: Brain tumor; neuroepithelial tumors; ganglioglioma; clinic, radiology; treatment.

INTRODUCTION

Desmoplastic ganglioglioma (GGD) is a benign neuroepithelial tumor of the central nervous system (CNS), corresponding to WHO grade I, occurring mainly in children, before the age of two years [1].Typically characterized by bulky size; glial and neuroganglionic differentiation. The involvement of several lobes is frequent, with a predilection for the frontal and parietal lobes [1-3]. Its metastatic power is extremely rare; but some cases of malignant transformation are reported in the literature. The revealing symptomatology differs according to the age of the child and the cerebral localization the radiological diagnosis is based on cerebral MRI and the spectro sequence, objectifying a double solid-cystic component, and a slight peri-lesional edema generally without mass effect [1, 5]. The GGD have a good prognosis and complete surgical excision is sufficient as treatment [6]. The use of adjuvant therapy is still controversial, Because of the small number of patients.

OBSERVATION

A 7 years old boy without particular antecedent, presents with a 2 years history of intermittent headache; complicated 5 months ago with a Intra cranial hypertension syndrome.

Clinical exam found a left hemi paresis (facial and corporal). The clinical picture (HTIC syndrome, sign of focus) evokes an expansive cerebral process.

A Computed tomography (Fig-1) confirmed the existence of a right heterogeneous parietal tumoral process, measuring 41 mm, with a fluid component evoking a glioblastoma, responsible for a mass effect and generates a discreet perilesional edema

Magnetic resonance imaging showed a 43 x 41 x 41 mm right parietal process with double component (solido-kystic), in hypointense T1, hyperintense T2, enhanced in its septums and bourgeoning parts after injection of contrast product, with poorly perilesional edema responsible for a mass effect on homolateral ventricular system and median line, the spectro sequence shows an decrease in the choline / creat ratio and lipid peak (Figure 2 & 3).

The young boy underwent a partial excision-biopsy

Pathological analysis showed a malignant proliferation of cerebral parenchyma, made of cells of variable size, a mitotic index of 3 mitosis par 10 fields, organized and supported by a desmoplastic fibrillar network, suggesting an infantile desmoplastic ganglioma.

Immuno-chemical analysis supports the diagnosis; positive staining for: anti-GFAP antibodies, anti-vimentine, anti-PS 100, and anti-CD68.

Post-operative MRI showed a right fronto-temporo-parietal solido-kystic process of 27x 15 mm with a mass effect on ventricular systeme and cranial flap, with drop of all metabolites and lipid peak. Evoking malignant transformation **(**Figure 4-6).

The young patient underwent a second resection; witch pathological and immune-chemical analysis concluded to an anaplasic glial tumor.

He received an adjuvant radiation therapy, at a dose of 54 Gy in 30 sessions of 1,8 Gy.

The evolution was marked by a clinical improvement, an MRI is planned in 3 month for evaluation.

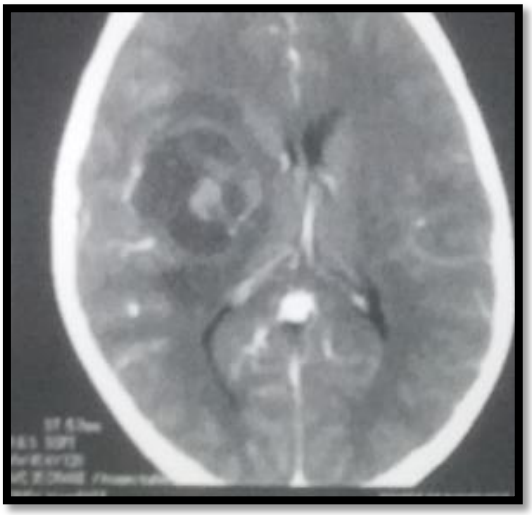

Fig-1: Brain CT axial section: right heterogenous parietal tumoral process, responsible for a mass effect and generates a discreet perilesional edema

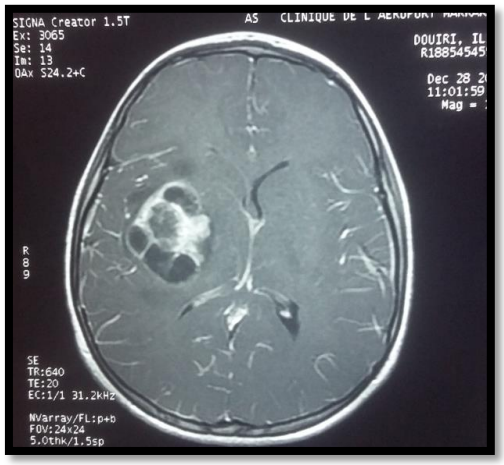

Fig-2: T1-weighted axial section: right parietal process with double component (solido-kystic), cysts appear hypointense on T1-weighted, generates a discreet perilesional edema.

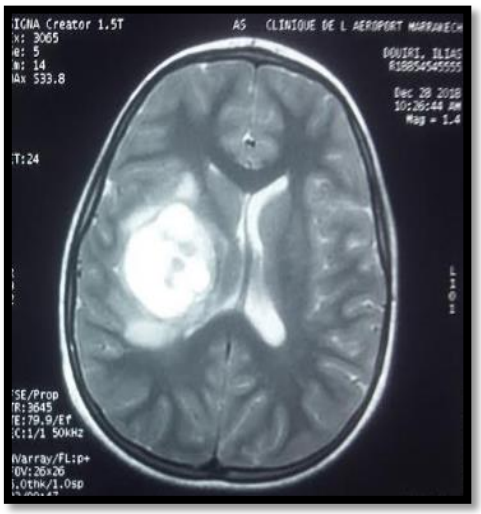

Fig-3: Brain MRI. T2-weighted axial section: right parietal process Lesion predominantly cystic tumor with a fleshy nodule peripheral and very discreet edema

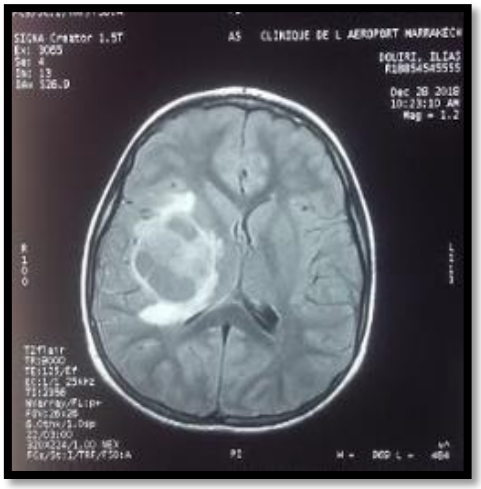

Fig-4: BRAin MRI, FLAIR section: pcs with double componenet in intermediate intensity

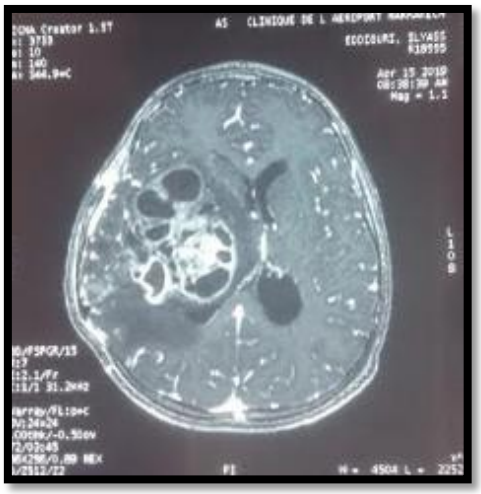

Fig-5: Right fronto-temporo-parietal solido-kystic process with a mass effect on ventricular systeme and cranial flap. The component fleshy intensifies after injection of contrast medium

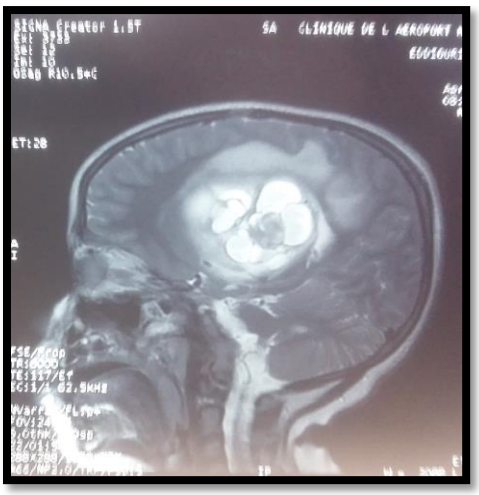

Fig-6: Bain MRI; T2 weighted frontal section;fronto-temporopariatal solido-kystic pcs

DISCUSSION

Described for the first time by Vandenberg [7], GGd is a generally well-differentiated neuroepithelial tumor, are classically considered World Health Organization (WHO) grade I or II, rarly anaplastic features or necrosis in the glial component can result in upgrading to WHO grade III or IV. However it constitutes a rare tumor entity which represents only 0.3% of brain tumors at any age [8], 1.25% of childhood brain tumors [3] and 1.3 to 15.8% of brain tumors in infants [9]. There is a slight male predominance [4].

DIG is a tumor that affects both cerebral hemispheres; predominantly at the temporal lobe, followed by the frontal and parietal lobes as our observation. Atypical localizations are described in the literature, citing the suprasellar region [10-13], the brainstem [14] or the thalamus [15] as well as multiple locations [4].

The revealing symptomatology depends on the age of the child and the seat of the tumor, often progressive and slow, it generally includes an increase in head circumference, bulging fontanelle, hemiparesis and frequent convulsions [4, 13]. Other forms of clinical presentation include HTIC syndrome and deficiency signs are rare and late [15].

On CT scans, DIG is seen as large, hypodense or slightly hyperdense superficial portion that extends to the overlying meninges and shows intensely contrast enhancement. The cystic portion is usually located deep, whereas the solid portion is peripheral [13, 16, 5].

MRI is the reference radiological examination to diagnose DIG, showing the presence of a supratentorial and large tumor, usually frontoparietal, with a dual component: cystic hypointense in T1, hyperintense in T2, and solid isointense in T1, T2 . Magnetic resonance spectroscopy (MRS) can be very useful in monitoring metabolic changes in the tumor, gangliogliomas were usually hypometabolic [16, 5, 17, 1].

The histological examination reveals a pleomorphic tumor involving the meningeal space and the cortex, characterized on the macroscopic level by its large size, its well-defined contours, with a double component; wide cystic, and fleshy adherent to the meninges. Microscopically, large areas of intense desmoplasia are found adjacent to and intermixed with pleomorphic neuroepithelial tumor cells with mixed Glial and neuronal differentiation and poorly mitosis [2, 1]. In immunohistochemistry, the ganglionic tumor component expressed, in all cases, neurofilamennts, synaptophysin, chromogranin. While the glial tumor quota expresses GFAP, Ki-67 and survivin, these different markers help to pose the diagnosis [2, 18].

At the moment there are no molecular genetic data that would enable us to predict tumor progression DIGs [19]. Comprehensive tumor molecular profiling led to the discovery of a novel BRAF alteration, increasing the number of BRAF alterations identified in DIG This finding suggests that, like other low-grade neuroepithelial tumors, MAPK pathway activation may have a potential oncogenic-driver role in a subset of patients with DIG/DIA [20].

The relative rarity of this tumor and its somewhat indolent course contribute to the difficulty in determining therapeutic strategy; but the majorities of the authors plead for radical resection [21, 22], the location of the tumor and the presence of several large cysts, play a role in the favorable results obtained in these patients, but unfortunately, complete resection is not always easy to, because of the young age, the large size of these tumors and their close attachment to the dura mater [17, 23], and partial resection increases the risk of malignant transformation and recurrence as our case [21].

Adjuvant therapy may be necessary, especially in incompletely resected tumors [22]. Adjuvant chemotherapy is discussed for tumors with anaplastic characteristics; deep with aggressive behavior [22]. Adjuvant RTH is not systematic, and may overtreat patients, it's reserved exclusively to residues or tumor recurrences whose surgical revision is no Feasible or anaplastic forms as our case. Compliant techniques must be used to optimally save normal tissues. The value of radiotherapy has not been established [10], Cranial irradiations at doses of 35 to 60 Gy have been recommending [22, 24-26].

Gangliogliomas are often associated with long survival. A review of 42 cases of supratentorial gangliogliomas surgically treated shows a statistically significant association of survival with younger age, female gender, and lack of histological evidence of

malignancy [6]. The prognosis is generally good in cases of complete resection, and spontaneous regression is possibly. The recurrence-free intervals range from 6 months to 19 years [13, 27].

In the present case, the behavior of the tumor apparently changed during the course of the disease from low-grade DIG to a high-grade primitive tumor. It remains debatable whether this tumor had changed its benign nature into malignant during its course because of incomplete surgery, or whether this tumor had a malignant subpopulation within the DIG from the beginning.

CONCLUSION

Low-grade gangliogliomas (GGs) can be surgically treated with good long-term results. Malignant degeneration of a benign ganglioglioma is a rare occurrence. Further studies of this event are desirable to confirm these findings, and clarify the exact role of each therapeutic weapon.

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