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Case Report

Epilepsy Revealing a Infiltrating Glioma of the Brainstem: A Case Report and Review of the Literature

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Abstract: *Introduction:* Invasive brainstem glioma (IBG) is a rare, very aggressive pontine glial tumour. It remains one of the most devastating diagnoses among childhood cancers. We report the observation of an infiltrating brainstem glioma. *Observation:* This is a 7-year-old girl, with no notable medical and surgical history, who presented in a rapidly progressive context, headaches, vomiting, weakness of the left hemibody, aphasia, tonic-clonic seizures, neurological examination had as particularity, a pyramidal syndrome, a syndrome of intracranial hypertension, and a syndrome of cortical irritation, on the biology one noted a hypercalcemia, encephalic MRI revealed a hyper signal in T2 at the expense of the brainstem, she received corticosteroid therapy, followed by antiepileptic treatment. Evolution resulted in the death of the patient 3 months after the diagnosis. *Discussion:* Invasive brainstem glioma accounts for about 15% of pediatric brain tumors, presumed diagnosis of invasive brainstem glioma has been progressively made based on clinical and radiological aspects only. For lesions with typical clinical presentation and imaging. Surgery and radiotherapy currently remains the standard treatment, *Conclusion:* The management of infiltrating gliomas of the brainstem remains delicate, and the prognosis always remains gloomy, our study is an illustrative case, it is one of the very first case of brainstem gliomas described in children in the republic of guinea. Large-scale studies would be necessary to determine the prevalence of the pathology on a national level.

Keywords: Infiltrating Glioma, Brainstem, Chemotherapy

1. Introduction

Invasive brainstem glioma (IBG) is a rare, very aggressive, infiltrated and diffuse pontine glial tumor usually occurring in children [1]. GITC is a rare type of brain tumour. It represents between 10 and 15% of all brain tumors in children and mainly affects children between 5 and 10 years old with a peak incidence at the age of 6 years [2]. Diagnosis is classically based on a typical magnetic resonance imaging (MRI) appearance in a child presenting with suggestive neurological signs such as pyramidal involvement with hyperreflexia, movement coordination disorders, abnormalities of the cranial pairs responsible for strabismus or facial paralysis [3]. Pathological confirmation is, in this context, considered as not very useful because it only rarely modifies the therapeutic management [4].

Surgical treatment of cerebral GITC is very complex and resection is most often unfeasible, due to the highly infiltrating nature of the lesion [5].

Radiotherapy, in addition to surgery, currently remains the standard treatment [6].

The evolution is done towards an extension of the tumoral

infiltration in all the directions (elongated medulla, cerebral peduncles, cerebellum) then late by leptomeningeal metastases [7]. Almost all children will die of their disease between 6 months and 2 years after their initial diagnosis [8].

We report the case of a patient with an infiltrating brainstem glioma.

2. Case Description

This is a 7-year-old girl, schoolgirl, right-handed, with no notable medical and surgical history, who presented in a rapidly progressive context, headaches, vomiting, weakness of the left hemibody, aphasia, generalized tonic-clonic seizures of unspecified duration and a notion of fever. Neurological examination revealed left hemiparesis rated at 3/5 proportional, pyramidal-type hypertonia, hyper-reflexia, left Babinski's sign; paralysis of laterality of gaze; IV paralysis and convergent strabismus. The rest of the clinical examination was otherwise unremarkable. The usual biological assessment showed as a particularity hyponatremia at 129 mmol/l, discreet hypocalcemia at 1.41 mmol/l. Hepatic and renal function tests were normal.

Figure 2 The patient received Phenobarbital (1 tab * 2/day), Dexamethazone (0.3 mg/kg), Albendazole (1 tab/day), Omeprazole (10 mg/day), prednisone (20 mg 2 tab in PU).

Evolution was marked 3 months after the diagnosis, by a recurrence of convulsive seizures complicated in status epilepticus followed by the death of the patient in a cerebellar engagement table.

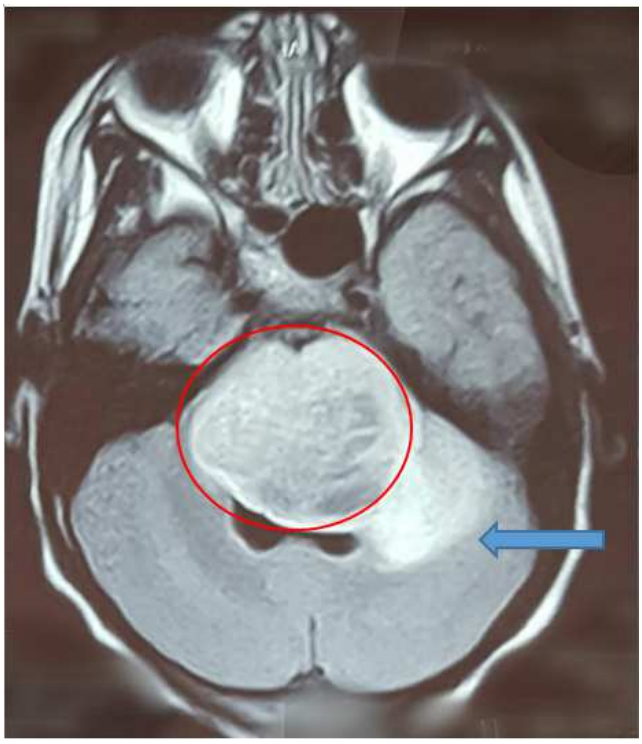

Figure 1. *Well-limited T2 hypersignal involving the brainstem, compressing the IV ventricle with peri-lesional edema, blue arrow.*

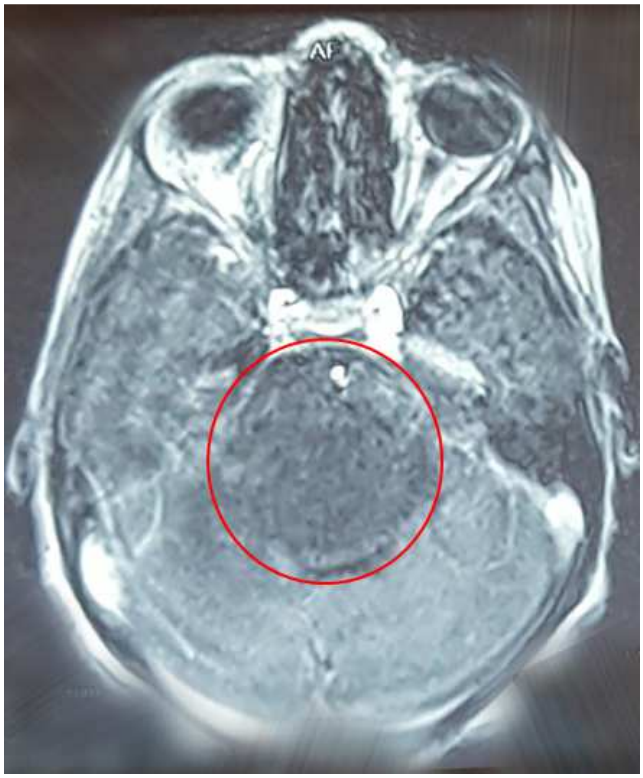

Figure 2. *Hypo signal in T1 gado with absence of contrast uptake, interesting the brainstem.*

3. Discussion

The identification of an infiltrating brainstem glioma (DIPG) in a child remains one of the most devastating diagnoses among childhood cancers [8]. It accounts for approximately 15% of pediatric brain tumours. They include two types of tumors. The first includes infiltrating gliomas of the brainstem, of very reserved prognosis, of infiltrating appearance, located in the pons and often encompassing the basilar artery, is the subject of this article. The second includes focal lesions located in the bulb, rather posterior, sometimes resectable and whose histological type is “pilocytic astrocytoma” with a much more favorable prognosis [9]. The age of our patient was 7 years old. This is slightly higher than that of the literature which states the peak of incidence is around 6 years [2]. Lefebvre J et al, in a study published in 2015 also reported a peak around age 6 [10]. The mode of revelation of the pathology was rapidly progressive, Veldhuijzen van Zanten et al. In the Netherlands found at 96% of their patient's median duration of symptoms before diagnosis was 1 month [11]. The clinical picture was dominated by a pyramidal syndrome, a cortical irritation syndrome, IV and VI paralysis, and an intracranial hypertension syndrome. According to the literature Three symptoms predominate in patients with a DIPG: cranial pair deficits, ataxia and clinical signs of the long tract (hyperreflexia, clonus, increased tone, presence of a Babinski). A subgroup of patients may present with signs of hydrocephalus and intracranial hypertension [12]. Paradoxically, we notice in our patient a notorious absence of

ataxia or movement coordination disorder probably because of the motor deficit which, according to the literature, is a main symptom revealing GITC. This is the case of the study carried out by Schumacher M, et al. In 2007 [13]. In terms of imaging, brain MRI was our first-line examination, MRI is the imaging modality of choice and infiltrating brainstem glioma presents as an expansive lesion centered on the pons, invading more than 50% of the pons, with high signal in T2-weighted sequences and low signal in sequences T1-weighted, enhancement is weak or absent, sometimes a cockade is visible. The expansion of the volume of the protuberance can envelop the basilar artery. Micro-bleeding can be observed [14]. The anatomo-pathological examination and molecular biology were our limits, Since the introduction of imaging methods capable of distinguishing infiltrating gliomas of the brainstem from other diagnoses by its characteristics, the presumed diagnosis of an infiltrating glioma of the brainstem has gradually been made on the basis of clinical and radiological aspects only. For lesions with typical clinical presentation and imaging. Biopsy is considered irrelevant and dangerous, given that the prognosis of these lesions is very poor regardless of the grade according to the WHO classification. Until now, biopsy has been reserved for radiologically or clinically atypical cases [15]. Our patient benefited from corticosteroid bolus treatment accompanied by adjuvant measures to this corticosteroid therapy associated with antiepileptic treatment. The first-line treatment of malignant gliomas is surgery, which must be as complete as possible and the quality of which has a major impact on recurrence-free survival and overall survival. In the case of brain stern localization, excision is the most often impracticable, due to the very infiltrative nature of the lesion in the eloquent zone, and can in no case be complete [5]. It is often combined with focal radiotherapy at a dose of 50–60 Gy, with a classic fractionation of 1.8–2 Gy per session and per day. This irradiation most often leads to a clinical improvement in the patient with a reduction in his symptoms and makes it possible to reduce or even stop the treatment with corticosteroids, generally started as soon as the diagnosis is made in order to limit the clinical consequences of the peritumoral edema [6]. Compared to chemotherapy. The response rate is extremely low, around 4%, including with the use of very high doses followed by an autologous hematopoietic stem cell transplant. Temozolomide has been proposed after radiotherapy in infiltrating gliomas of the brainstem, with a conventional administration schedule (200 mg/m^2/day, 5 days out of 28). This administration schedule did not improve patient survival [16]. Evolution was marked by a resurgence of epileptic seizures complicated in status epilepticus, a disorder of vigilance type of coma followed by death in a table of cerebellar engagement. Data from the literature report that despite aggressive therapeutic approaches, the prognosis of malignant brainstem gliomas remains particularly poor with an overall survival rate of around 10% at 2 years and a median survival of 9 at 12 months depending on the series [5]. In the case presented, the tumoral nature, the location of the tumor associated with the lack of financial means to ensure adequate care probably contributed to the death of the patient.

4. Conclusion

The management of infiltrating gliomas of the brainstem in children has improved over the last decades, but their prognosis remains gloomy because always associated with a high mortality rate, our study is an illustrative case, it is This is one of the very first cases of brainstem gliomas described in children in the Republic of Guinea, large-scale studies would be necessary to determine the prevalence of the pathology at the national level.

Conflicts of Interest

All the authors do not have any possible conflicts of interest.

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