Interdisciplinary Neurosurgery 18 (2019) 100499

Contents lists available at ScienceDirect

Interdisciplinary Neurosurgery

journal homepage: www.elsevier.com/locate/inat

Case Reports & Case Series

Paediatric bilateral thalamic glioma: Case report and literature review

Badejo O.A.[a], Osobu B.E.[b], Salami A.A.[c], Adeyinka A.O.[b], Shokunbi M.T.[a,*]

[a] Division of Neurological Surgery, Department of Surgery, College of Medicine, University of Ibadan, Nigeria
[b] Department of Radiology, University College Hospital and College of Medicine, University of Ibadan, Nigeria
[c] Department of Pathology, College of Medicine, University of Ibadan, Nigeria

ARTICLE INFO

Keywords:
Bilateral thalamic glioma
Diffuse low-grade astrocytoma
Endoscopic transventricular biopsy

ABSTRACT

We report the case of a child with bilateral thalamic glioma. The outcome of his management and a literature review are also presented.

1. Introduction

Primary thalamic tumours are rare [1]. They account for 1–1.5% of all brain tumours and approximately a quarter of them occur in children younger than 15 years [2,3]. Bilateral thalamic gliomas (BTGs) are extremely rare sub-types of thalamic tumours, which are known to have a poor outcome regardless of the treatment modality [4]. About 70 cases have been reported so far in the literature [5]. We present the outcome of treatment of BTG in a three year old Nigerian child and a brief review of the literature on these uncommon type of central nervous system tumours.

2. Case presentation

A three-year-old right-handed boy who presented with a headache, abnormal gait and inability to sit unsupported of a week duration. There was an associated history of drowsiness, excessive sleeping, and multiple episodes of projectile vomiting. Examination revealed a young boy who was fully conscious but drowsy. His pupils were of normal size but reacted sluggishly to light. He had bilateral abducens nerve palsies, bilateral papilloedema, global hypertonia/hyperreflexia and bilateral extensor plantar response. He also had truncal ataxia and dysmetria but no sensorimotor deficit. Examination of other systems revealed normal findings.

A clinical diagnosis of acute onset raised intracranial pressure from an infratentorial space-occupying lesion was made. Cranial Computed Tomography scan showed bilateral symmetrical enlarged thalamic nuclei which were hypodense to isodense and non-contrast enhancing (Fig. 1a and b). There was associated obstructive hydrocephalus. Brain Magnetic Resonance Imaging showed bilateral symmetrical non-enhancing masses involving both thalami (with estimated volumes of 40.17cm^3 on the right and 44.84cm^3 on the left). These were hypointense on T1 weighted images and hyperintense on T2 weighted and FLAIR images. There was associated dilatation of the lateral ventricles and effacement of the quadrigeminal/ambient cisterns bilaterally (Fig. 2a–d). We made a radiological diagnosis of a bilateral thalamic tumour. The patient's management was multidisciplinary involving the neurological surgery, radiology, pathology, paediatric oncology and radiation oncology teams.

A biopsy specimen obtained via an endoscopic transventricular route showed features of a WHO grade II diffuse astrocytoma (Fig. 3a–c). He subsequently received sixteen courses of Vincristine/Carboplatin chemotherapy, which was later changed to Etoposide/Cisplatin on account of clinico-radiological evidence of tumour progression. He had cerebrospinal fluid diversion (via a ventriculoperitoneal shunt) five months after the initial procedure due to worsening hydrocephalus (Fig. 4a–c). The second line chemotherapeutic agents were discontinued after the third cycle on request by the patient's mother because of their side-effects (bone marrow suppression/widespread dermatitis/recurrent chest infection). Radiotherapy was considered unsafe in this patient given his age and the potential for radiation-induced neurocognitive decline (on the advice of the radiation oncology team). Repeat neuroimaging at six months following the initial surgery showed further tumour progression with involvement of the caudate nuclei/brainstem and extension into the lateral ventricles (Fig. 5a–c). Attempt at switching his chemotherapeutic agents were futile due to non-availability of the drugs. At nine months post tumour biopsy, he had recurrent headache/vomiting, expressive aphasia, worsening gait imbalance, ataxia, paraparesis, bilateral ptosis, upgaze paresis and choreoform movement. He subsequently defaulted follow-

* Corresponding author at: Division of Neurological Surgery, Department of Surgery, College of Medicine, University of Ibadan, PMB 5112, Nigeria.
E-mail address: temitayoshokunbi@yahoo.com (M.T. Shokunbi).

https://doi.org/10.1016/j.inat.2019.100499
Received 30 November 2018; Received in revised form 4 January 2019; Accepted 25 May 2019

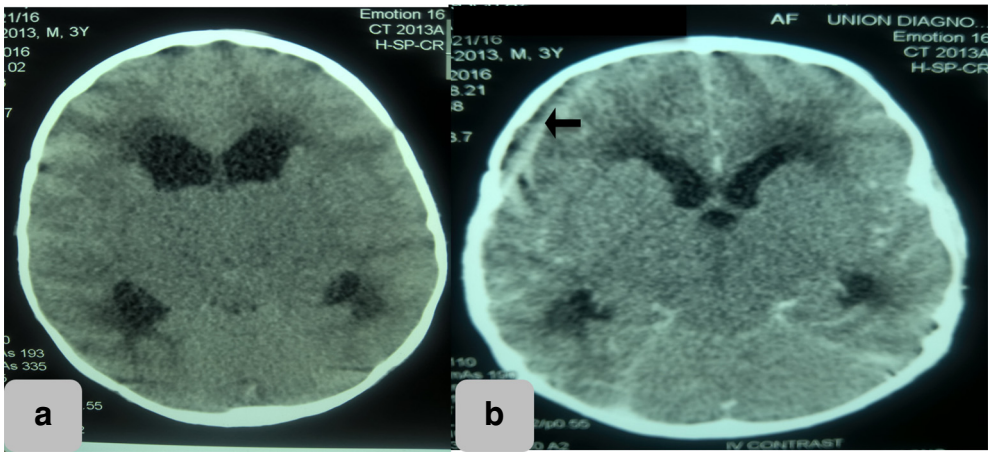

Fig. 1. Axial images, Cranial computerized tomography scan.
a: Non-contrast brain image showing isodense thalamic glioma bilaterally with dilatation of frontal horns of both lateral ventricles.
b: Post-contrast image showing no enhancement of the tumour. There is a motion artifact (black arrow) in the right frontal convexity mimicking a subdural collection.

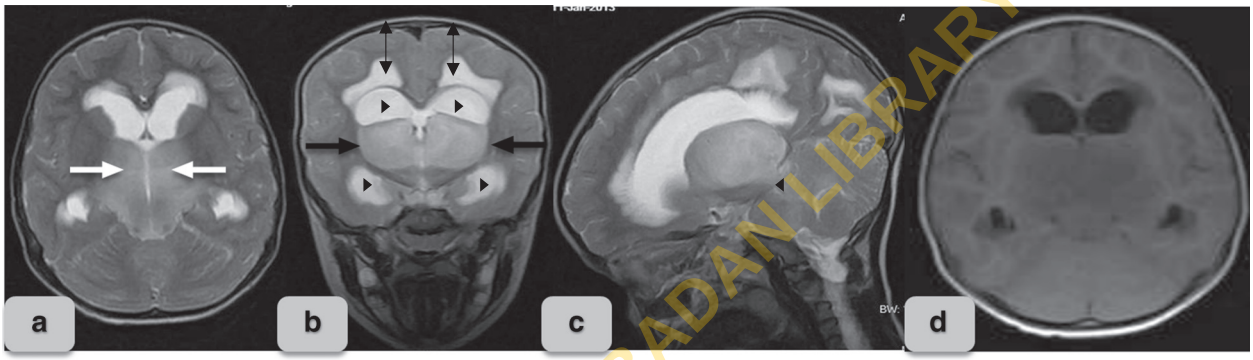

Fig. 2. Brain Magnetic Resonance Imaging.
a: T2 weighted image showing hyperintense diffuse thalamic glioma bilaterally (white arrows) with associated ventriculomegaly.
b: Coronal T2W Brain MRI showing limitation of the tumour to the thalami (thick black arrows). Note the dilatation of the lateral ventricles (black arrowheads) and transependymal seepage of cerebrospinal fluid (thin black arrows).
c: Sagittal T2 weighted brain image showing an extension of the tumour toward the midbrain (black arrowhead).
d: Axial T1 weighted brain MRI post-Gadolinium administration showing no contrast enhancement.

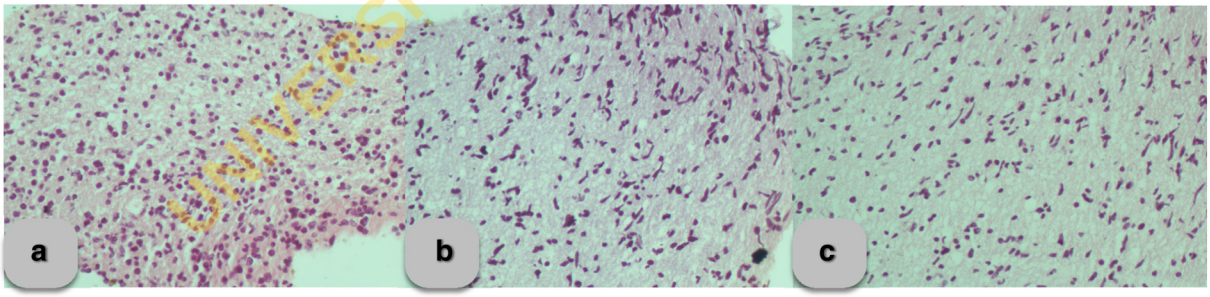

Fig. 3. Hematoxylin and Eosin staining of the biopsy specimen.
a–c: Showing a fairly cellular lesion with moderately pleomorphic cells having round to elongated nuclei and scanty cytoplasm on a fibrillary background. There are microcystic spaces in some areas while other areas appear more compact. Also seen are focal areas of haemorrhage. Note the absence of mitotic figures in these photomicrographs.

up and was confirmed by his mother to have died at home a year after initial evaluation and tumour biopsy.

3. Discussion

Primary bilateral (unlike unilateral) thalamic tumours are incredibly uncommon. Very few case reports exist in the literature [3,5,6]. To the best of our knowledge, this is the first reported case from sub-Saharan Africa. There is no gender predilection, and > 50% of the reported cases fall within the paediatric age group [5]. The occurrence of BTG is not widely accepted, and two schools of thought have been propounded regarding their origin. Some authors have postulated that they arise on one side and subsequently spread to the contralateral side with the passage of time (a phenomenon seen in about 33% of unilateral thalamic gliomas). Others consider them to proliferate from the subependymal region of the third ventricle [3,4,7]. Classical morphological features of these lesions are diffuse symmetrical enlargement of the thalamic nuclei and the absence of bridging tumour between the two [7,8]. They typically involve the dorsomedial and intralaminar nuclei of the thalami and often spare the adjoining third ventricle, temporal lobes, midbrain and pineal gland until late stages of the disease [4,7].

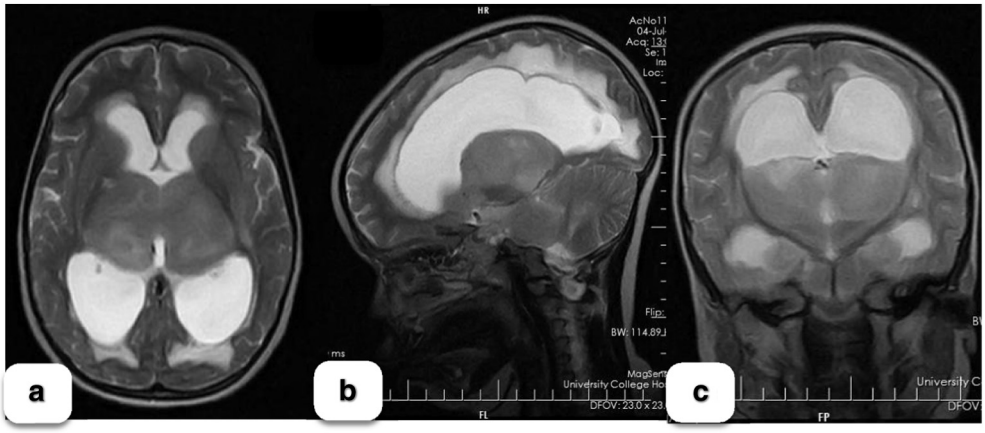

Fig. 4. (a–c): T2 weighted brain MRI showing progressive hydrocephalus five months postoperatively. Note the larger size of the lateral ventricles compared to the initial neuroimaging.

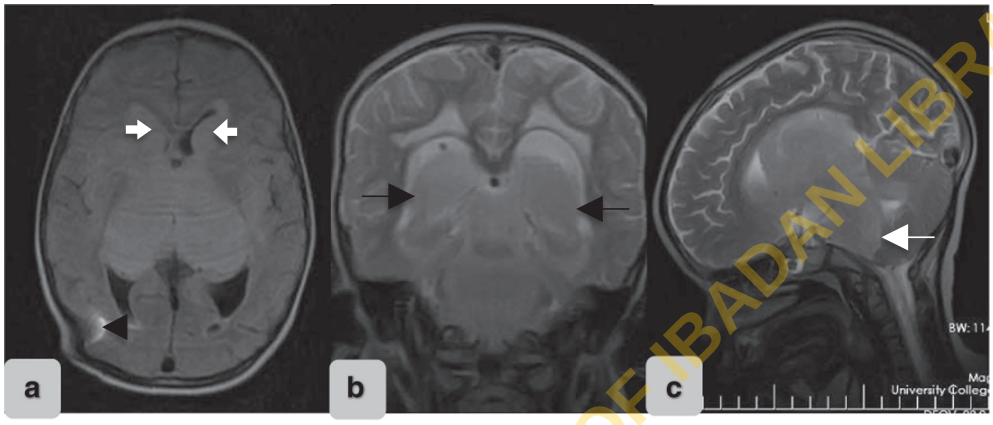

Fig. 5. (a–c): Non-contrast enhanced brain MRI (six months post-endoscopic transventricular tumour biopsy).
a: Axial FLAIR image showing involvement of the caudate nuclei (white arrows) and further increase in tumour size. Note the shunt artifact in the right occipital region (black arrow head) and reduction in the ventricular size.
b: Coronal T2W Brain MRI showing extension of the tumour into the lateral ventricles (black arrows).
c: Sagittal T2 weighted brain image showing brainstem involvement (white arrow).

The age at presentation varies from three months to eighty years with few cases reported below six years of age [3,5]. There is a paucity of literature on the histologic sub-types in children [9]. Most of them have been found to be low grade astrocytomas but anaplastic and glioblastoma sub-types have been reported especially in older individuals [5,7,10]. Some other authors on the subject found no correlation between the histologic type of BTGs and patients' age [10]. The clinical manifestations of primary bilateral thalamic tumours can be explained on the basis of the involvement of the different nuclei or tracts of this region. These include gait unsteadiness, sensory abnormalities, motor deficit, dysmetria, torticollis and nystagmus [3,8]. Affected adults with BTGs may present with personality changes, emotional lability, loss of memory, apathy and cognitive impairment [7]. Features of raised intracranial pressure (ICP) occur late in the disease and are usually due to mass effects of the lesion rather than hydrocephalus which may be mild or absent [3,7,8]. However, a high incidence of increased ICP has been reported to be associated with thalamic tumours [10]. Our patient had headaches, vomiting, abnormal gait, hemiparesis and somnolence. He had hydrocephalus at presentation and this continued to worsen in spite of adjuvant chemotherapy, thus necessitating further neurosurgical intervention. Impairment of the cerebello-rubro-thalamic tract could explain the cerebellar features seen in this patient [8]. Duration of symptoms at the time of diagnosis and WHO grade of tumour have been reported to be independent prognostic indices of survival which has a median/mean duration of four months and thirteen months respectively [5].

Neuroimaging is indispensable in establishing diagnosis in affected individuals. The non-contrast enhancing nature of these tumours and similar density to the normal brain on CT scan make MRI a preferred neuroimaging modality compared to CT scan as they appear hypo- to isointense on T1 weighted images and hyperintense on T2 weighted MRI [3,5]. However, higher grade tumours can show varying degrees of contrast enhancement [7]. Radiological differential diagnosis of BTGs include other brain tumours (Lymphomas, teratomas, germinomas), toxic and metabolic disorders (Wernicke's encephalopathy, osmotic myelinosis), vascular conditions (infarcts, deep venous thrombosis) and infection (viral encephalitis, Creutzfeldt-Jakob disease) [3,7,11]. Magnetic Resonance Spectroscopy (MRS) is useful for differentiating these tumours from other lesions. It also depicts the dissimilarity between primary bilateral thalamic gliomas and unilateral thalamic gliomas with respect to their metabolic characteristics [3]. An MRS pattern distinctive for BTGs is increased creatine-phosphocreatine peak [7,10,12]. The role of this modality for non-invasive diagnosis requires further exploration.

Surgical intervention of primary bilateral thalamic tumours poses a unique challenge because of the diffuse bilateral involvement of the thalamic nuclei by these tumours, which makes radical excision untenable. It is therefore not surprising that no case of radical excision has been reported in the literature [7,8]. The goal of surgery is to obtain biopsy for histopathological diagnosis and this can be safely achieved by endoscopic means [3,7,8]. Adjuvant therapy (radiotherapy ± chemotherapy) have not been shown to be of significant benefit and prognosis remains poor regardless of the treatment modality [3,5,7,8].

4. Conclusion

Bilateral thalamic gliomas are rare neoplasms which differ from unilateral thalamic tumours with respect to their radiological

characteristics, metabolic profile and prognosis. Our patient's clinical features, tumour appearance on neuroimaging, histologic diagnosis, response to treatment and observed duration of survival are in keeping with the literature on BTGs.

Funding acknowledgement

None.

Declaration of Competing Interest

The authors declare none.

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