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Bilateral Thalamic Tumors: Two Case Report and Literature Review

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Abstract

Primary thalamic tumours are rare, bilateral thalamic tumors are extremely rare sub-types of thalamic tumours, account for approximately 10% of reported thalamic tumor cases. Clinical manifestations of primary bilateral thalamic tumours depends on the involvement of the different nuclei or tracts of this region. Radiological investigations show bilateral thalamic lesions with varying features suggestive of gliomas or other infiltrative tumors. Its intricate intrinsic circuitry and connections with eloquent adjacent structures has made the thalamus a seemingly surgically inaccessible structure. Radiotherapy is the main mode of treatment since surgical intervention is limited to a role of biopsy and management of secondary effects. This case report illustrates the complexities involved in managing bilateral thalamic lesions and emphasizes the importance of tailored approach considering the clinical context and radiological features in the absence of biopsy confirmation.

Introduction

Primary thalamic tumours are rare. Thalamic tumors compose 2% to 5% of the pediatric brain tumor population, and only 1% of the adult brain tumor population [1,2]. Based on anatomic localization and spread, two distinct types of primary thalamic tumors have been identified: Unilateral and Bilateral. Over 80% of thalamic tumors are unilateral, however Bilateral Thalamic Tumors (BTT) are extremely rare sub-types of thalamic tumours. BTT account for approximately 10% of reported thalamic tumor cases in which approximately a quarter of them occur in children younger than 15 years with no sex predominance and the rest in adults (mean age for children is 6.6 years and for adult is 43.1 years) [2]. The occurrence of BTT is debated and two schools of thoughts have been postulated regarding their origin. Some authors have hypothesised that they arise on one side and subsequently spread to the contralateral side over 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]. Classical morphological features of these lesions are, diffuse symmetrical enlargement of the thalamic nuclei and the absence of a bridging tumour between the two. They typically involve the dorsomedial and intralaminar nuclei of the thalami and often bypass the adjoining third ventricle, temporal lobes, midbrain and pineal gland until late stages of the disease. Its complex intrinsic circuitry and connections with eloquent adjacent structures has made the thalamus a surgically challenging structure.

The clinical manifestations of primary bilateral thalamic tumours can described with involvement of the different nuclei or tracts of this region. Manifesting symptoms can be headache, memory loss, personality changes, apathy, emotional lability, cognitive impairment, gait unsteadiness, sensory abnormalities, motor deficit, dysmetria, torticollis and nystagmus. Features of raised intracranial pressure (ICP) occur late in the disease and are usually due to mass effects of the lesion rather than the uncommon effects of hydrocephalus. However, a high incidence of increased ICP has been reported to be associated with thalamic tumours [1,4]. On histopathology examinations, gliomas have consistently proven to be the most common pathologic observation in the thalamus for adults. Of gliomas, low-grade tumors are the most common about 65% or higher in some studies and include pilocytic astrocytomas, pilomyxoid astrocytomas, diffuse astrocytomas, and nonspecific low-grade astrocytomas. High-grade gliomas seen in the thalamus are most often anaplastic astrocytomas or glioblastomas, but others such as anaplastic gangliogliomas have been reported. Other rare tumor types include ependymomas, PNETs, gangliogliomas, ganglioneurocytomas [5].

In the pediatric population, tumor types and prevalence prove quite similar to adults, with low-grade gliomas being the most common. Similarly for bilateral lesions, although known to be a distinct entity from their unilateral counterparts, they too

are most commonly diagnosed as an low-grade lesions usually a diffuse low grade astrocytoma WHO grade II [5].

The prognosis of BTT is poor with a rapid and fatal evolution, the longest period of survival reported as 3 years in patients with WHO grade II glioma [1,6].

CASE 1

A 19-year-old female presented with headache, giddiness, right ear tinnitus, and forgetfulness. On neurological examination patient has right lower limb weakness and papilledema. MRI examination revealed large area of altered signal intensities within bilateral thalami. The lesion which is homogeneously hypointense on T1 Figure 1(A) and hyperintense on T2 Figure 1(B) and FLAIR. Entire lesion is measuring 68 x 57 x 49mm with no abnormal contrast enhancement. Findings are suggestive of low grade infiltrative bilateral thalamic glioma with involvement of the posterior half of midbrain including tectal plate. Compression of third ventricle is seen with dilatation of bilateral lateral ventricles with periventricular ooze indicative of hydrocephalus. Patient underwent for VP shunt procedure to decompress the intraventricular pressure. Computed Tomography (CT) scan of the brain showed non-enhancing hypodense enlarged thalami bilaterally and did not reveal any hemorrhagic components or calcifications within the tumor with decompressed ventricles with VP shunt Figure 1(C). Followed by open biopsy procedure via transcortical transventricular route. Histological examination was inconclusive. After oncology consultation patient was advised for radiotherapy but relatives refused for the same. The clinical follow-up over a one and half year period showed a stable disease without significant change in the clinical state. Follow-up MRI also showed stationary signal intensities but an increase in bilateral thalamic mass with left thalamus is more severely affected without any contrast enhancement, entire lesion is measuring 81 x 79 x 62mm Figure 1(E,F). Follow up CT also showed increased thalamic mass with stationary density with VP shunt in situ Figure 1 (D).

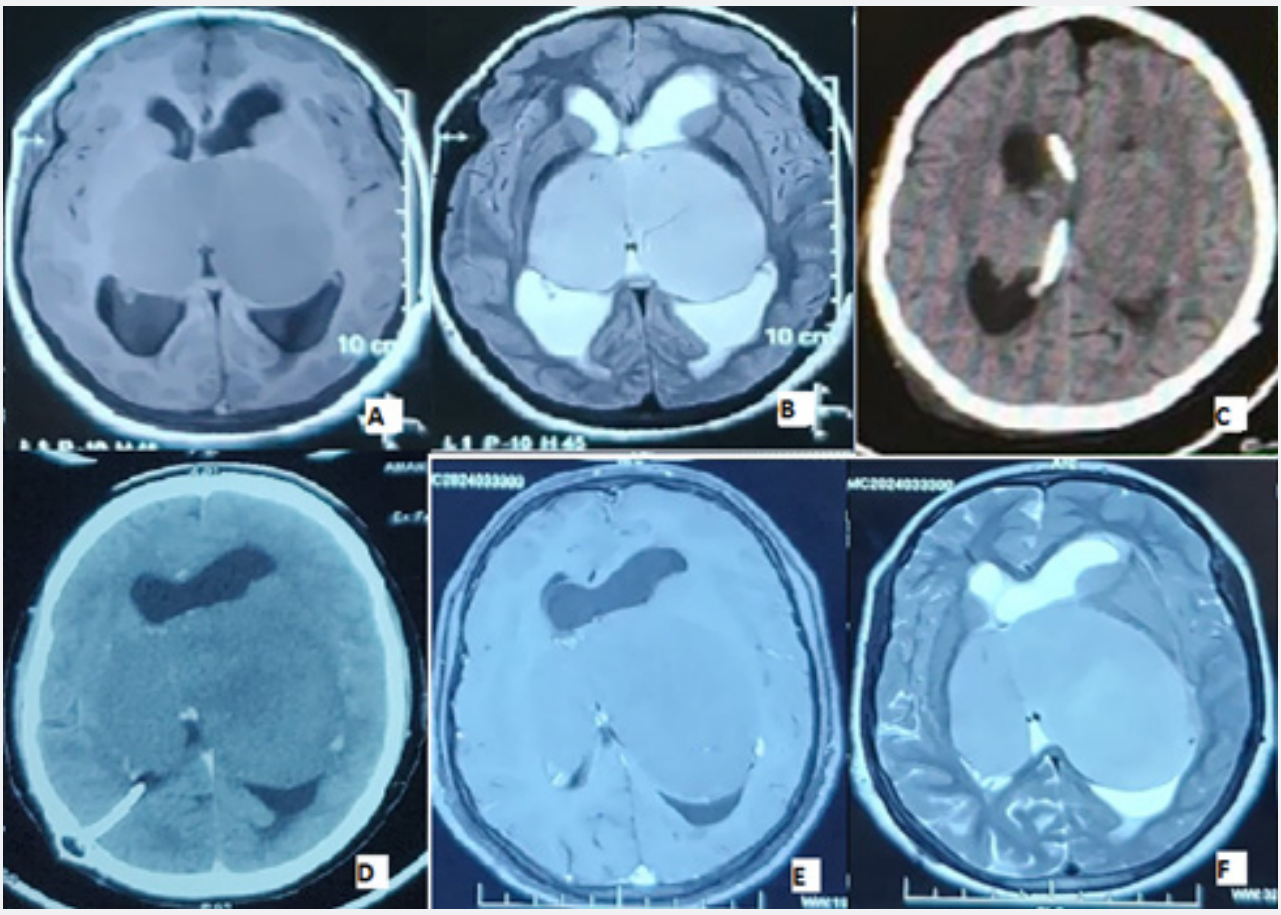

Figure 1: MRI examination revealed large isointense area on T1 (A), homogeneously hyperintense on T2 involving bilateral thalami with dilated lateral ventricles with periventricular ooze indicative of hydrocephalus(B), CT scan shows non-enhancing hypodense enlarged thalami bilaterally and without any hemorrhagic components or calcifications within the tumor with decompressed ventricles with VP shunt in situ(D). Follow up CT (D)and MRI(E,F) with stationary intensity with increased lesion with no abnormal contrast enhancement.

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CASE 2

A 11-year-old male presented with headache, vomiting and difficulty in speech and walking. On neurological examination, patient had all four-limb weakness (R>L) with dysarthria, and fundoscopy examination revealed papilledema. CT scan of the brain showed non-enhancing hypodense lesion involving bilateral thalami, with involvement of the midbrain causing significant compression on the third ventricle, resulting in obstructive hydrocephalus with no hemorrhagic components or calcifications within the tumor Figure 2(A). MRI examination revealed lesion of size 56 x 50 x 48 mm involving bilateral thalami, adjacent part of basal ganglia, right half of mid brain, foramen of Monro, causing compression of aqueduct of Sylvius, resulting in moderate dilatation of lateral ventricles and 3rd ventricle. Appearing heterogenous intensity lesion with hypointense on T1 Figure 2(B) and hyperintense on T2 and FLAIR images Figure 2(C), shows patchy restrictions on DWI Figure 2(E) with no obvious blooming on GRE Figure 2(D). MRS of the lesion, showed significantly elevated choline peaks with mildly reduced creatine and NAA peaks, indicating increased cell membrane turn over as well as neuronal loss elevated lipid and lactate peaks, indicating necrosis or anaerobic glycolysis. These findings are suggestive of mitotic pathology possibly gliomatous or neoplastic etiology. The patient underwent VP shunt procedure and one month later open biopsy was done via transcortical transventricular route and found to be a diffuse high-grade glioma (WHO grade IV). After oncology consultation patient was referred for radiotherapy. Over the period of three months patient deteriorated and became disoriented showed increased weakness in all 4 limbs, leading to inability to walk or sit with development of aphasia. Patient developed an infection in shunt and biopsy operative site with involvement of shunt tract involvement, later progressing to meningitis and ultimately succumbed.

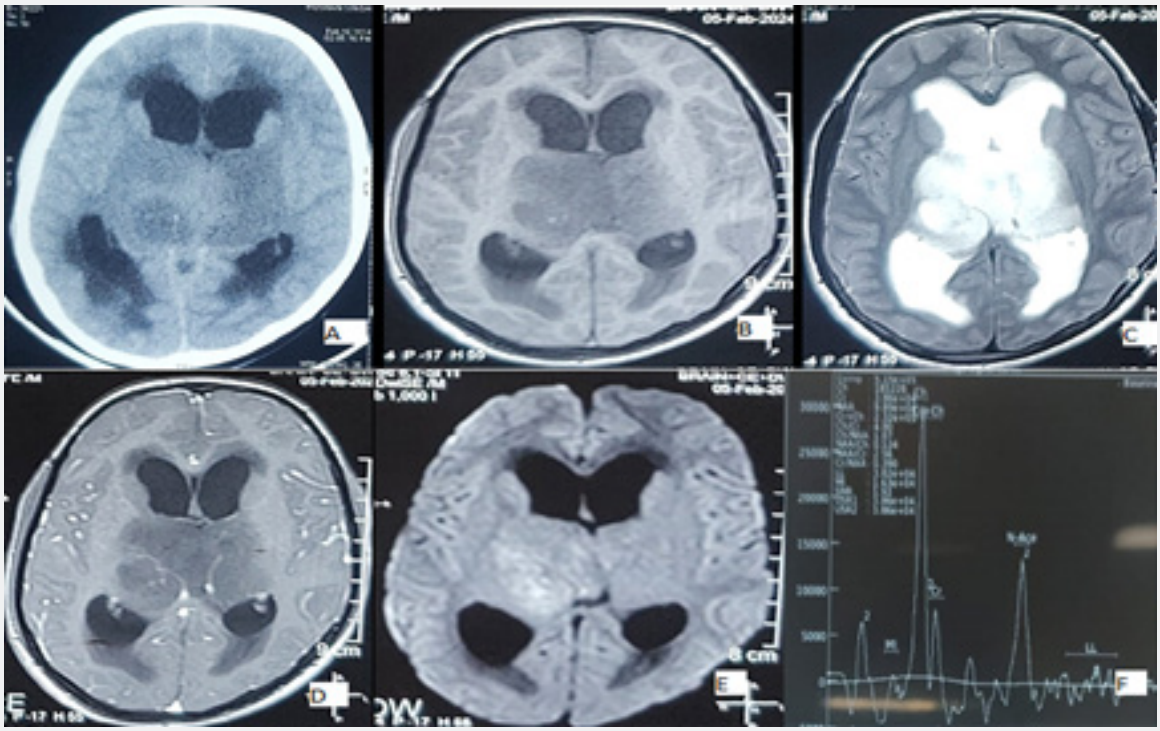

Figure 2: (A) non-enhancing hypodense enlarged thalami bilaterally, with involvement of midbrain on CT. MRI shows lesion involving bilateral thalami, adjacent part of basal ganglia, right half of mid brain, foramen of monro, causing compression of aquaduct of sylvius, resulting in moderate dilatation of lateral & 3rd ventricles, appearing hypointense on T1W (B) while hyperintense on T2W & FLAIR images (C), shows patchy restriction on DWI(E) with no obvious blooming on GRE (D) with significantly elevated choline peaks with mildly reduced creatine and NAA peaks (F).

Discussion

BTT is an extremely rare sub-type of thalamic tumors. Only a few case reports exist on this entity. They tend to have poor outcome regardless of the treatment and have a variable presentation and progression.

After extensive review of available literature we found that the age range of the BTT is very wide, varying between 3 months and 80 years. There are too few cases reported to correctly assess the prevalence of BTT for a certain age. Neverthless we have found that there are only few cases reported in patients over 65, possibly implying an earlier onset the disease [7].

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The presentation of BTT is extremely varied due to the involvement of the thalamic tracts, nuclei, and pathways of this region. Unfortunately, the early stages of the disease are silent, with the patients having mild signs and symptoms even with large tumors. Symptoms mainly comprise of mental deterioration, personality changes, and apathy, rather than focal neurological signs. Pediatric patients who rarely present with personality disorders even in the case of large tumors. As found in our study adult female patient presented with mild symptoms and personality disorder with slow pregression whereas, pediatric patient presented with more severe symptoms without personality disorder and with rapid progression [8].

We also found that BTT presents with characteristic imaging features. CT scans show symmetrical isodensity lesions and MRI reveals hypointense to isointense lesion in T1-weighted images and a homogeneous hyperintense lesion in T2 and FLAIR sequences. Gadolinium enhancement is not present in BTT as found in our case study, to help in diagnosis of tumor [9].

The histology of BTT is variable with a predilection being for WHO grade II. Astrocytomas accounted for 89.1% cases whereas glioblastoma multiforme was observed in 10.1% of patients [10]. The occurrence of BTT grade III and IV at ages over 65 years is extremely rare with more predilection for young age as in our study with one case having WHO grade IV diffuse glioma and other case biopsy is inconclusive [11].

Due to Diffuse and bilateral involvement of thalamus by these tumor makes surgical therapy arduous and some cases of debulking of tumor reported but no case of radical removal has been documented [12]. The role of surgery is to get tissue for histological diagnosis which can be obtained by open surgery with different approaches such as interhemispheric transcallosal, infratentorial suprasellar, transsylvian transinsular or transcortical transventricular approach or endoscopically or under stereotactic guidance. In cases of associated obstructive hydrocephalus, cerebrospinal fluid (CSF) diversion i.e VP shunt may offer symptomatic relief of elevated intracranial pressure but does not otherwise alter the course of the disease [4,12].

Generally, these gliomas are low-grade astrocytomas (grade II of WHO classification), but limited anaplastic areas may be encountered. Radiotherapy and chemotherapy are sometimes utilized as adjuvant therapy, but their role is controversial. Results are generally poor. Rapid fatal evolution of the disease makes these tumors a steep challenge [2,4,11]. Integrated meta analysis by Xiaodong Niu et al., included 70 cases from 32 publications on survival and treatment showed that Radiotherapy and chemotherapy can provide short-term advantage in clinical and radiographic standards and had better median survival than those without adjuvant therapies. The mean overall survival was 13.0 months, whereas the median overall survival was 4.0 months with longest documented survival was 3 years after diagnosis [6,10]. However, neither radiotherapy nor chemotherapy were independent predictive factor for survival [10].

Conclusion

Bilateral thalamic gliomas are extremely rare. The origin is presumably unlike unilateral thalamic gliomas. Patient presentation may vary with mild cognitive dysfunctions in large tumors including varied disease progession. Radiological investigations are insufficient to conclude the diagnosis, so this study illustrates the importance of tissue diagnosis. Its intricate complex circuitry and connections with eloquent adjacent structures has made the thalamus a seemingly surgically inaccessible structure. Radiotherapy and chemotherapy is the main modality of treatment since surgical intervention is limited to a role of biopsy and management of secondary effects. For the future recommendations, multicentre and collaborative research with larger series and in-depth studies needed to understand this rare disease and its predictive factors and treatment modality and survival.

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