Bilateral Thalamic Glioma : A Case Report

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Abstract

**Objects:** A 4-year-old boy is here presented with the signs of increased intracranial pressure including headache, vomiting, and sixth cranial nerve palsy.

**Methods:** The magnetic resonance imaging revealed symmetrically enlarged thalami with increased intensity on T2-weighted images. Magnetic resonance spectroscopy revealed decreased N-acetylaspartate levels with normal choline, creatinin, and lactate levels. The definite diagnosis of bithalamic glioma was made by lesional biopsy.

**Conclusion:** Bilateral thalamic gliomas are uncommon tumors with wide variety of symptoms. Typical radiological features, however, might suggest the diagnosis.

**Keywords:** Bithalamic glioma, intracranial childhood tumors

INTRODUCTION

Primary thalamic tumors account for 0.8–1.5% of all central nervous system tumors, and approximately 25% of them arise in children under the age of 15 years (Cheek WR et al., 1966; McKissock W et al., 1958; Partlow GD et al., 1992; Villarejo F et al., 1994). These tumors, are usually unilateral astrocytomas (McKissock W et al., 1958; Tovi D et al, 1961). A distinct type, bilateral thalamic gliomas are rare among the thalamic tumors, and has a very poor prognosis despite treatment (Partlow GD et al., 1992). Bilateral thalamic glioma is a large tumor appearing symmetrically in both thalami and is accompanied by personality change and/or mental deterioration, relative motor and sensory sparing with a very poor prognosis despite treatment (Estève F et al., 1999; Partlow
The diagnosis of bilateral thalamic tumors are difficult because of their nonspecific and nonfocal clinical signs and symptoms. Furthermore, neuroimaging features atypical for neoplasms can be mistaken for other diseases such as inflammatory, mitochondrial or vascular diseases (Carter DJ et al., 1989; Krouwer HGJ et al., 1995; Partlow GD et al., 1992; Rogers LR et al., 1994; Russell DS et al., 1989; Yoshida M et al., 1998). Here we present a 4-year-old child with bilateral thalamic glioma manifesting as left-sided sixth cranial nerve palsy and headache.

CASE PRESENTATION
The patient was brought to our neurology clinics because of internal deviation of his left eye. His past medical history revealed that four- 4-year-old boy had headache, nausea and vomiting on awakenings for the last one month, and he developed left-sided sixth cranial nerve palsy one week ago. He was the first child to non-consanguineous family, and born following an uncomplicated pregnancy. He had normal motor and mental developmental milestones. His family history was unremarkable. Systemic examination was within normal limits. Neurological examination was also normal beside the sixth cranial nerve palsy on his left eye and bilateral grade I papillaoedema.

His cranial MRI demonstrated that thalami were symmetrically enlarged, and homogeneously hyperintense on T2-weighted images, obstructing the third ventricle. The lesions had no contrast enhancement by the administration of intravenous gadolinium (Figure 1A, 1B). Because of the presence of enlargement of ventricles and progressive clinical picture, the patient was first consulted with the neurosurgeons and a ventriculo-peritoneal shunt catheter was placed at second day of admission. Following shunting surgery, headache and vomiting were disappeared, and the sixth cranial nerve palsy in addition to papillaoedema was totally recuperated.

A detailed biochemical tests were all normal. The proton MR spectroscopy (MRS) revealed diffuse decreased N-acetylaspartate (NAA) levels corresponding to bilateral thalami. These findings in addition to MRI features were supportive for low-grade glial tumors. For definitive diagnosis, biopsy was then performed from thalamus by interhemispheric tranecallosal approach. The sections of brain biopsy specimen showed diffuse permeative infiltration of tumor cells into natural brain parenchyma. Tumor was composed of ‘rod, shaped elongated cells with hypercromatic nuclei and bland cytoplasm (Figure 2A). Low mitotic activity (1-2 / 10HPF) was observed but no vascular endothelial proliferation and necrosis. The MIB-1 index was calculated as 4% (Figure 2B). The diagnosis was low grade astrocytoma with pathologically suggestive for gliomatosis cerebri pattern.

During the follow-up period, however, the patient developed fever and confusion, and diagnosed to have shunt infection. He had an urgent reoperation for reshunting, and treated for shunt infection appropriately with full recovery. Radiotherapy or chemotherapy has planned, but after one and a half months, he was rehospitalized in our neurology department being fed by nasogastric tube, unable to talk, and bed-written. Because of rapid deterioration in his medical condition, radiotherapy or chemotherapy could not be initiated. The patient died 7 months after the first symptoms appeared.
DISCUSSION

Bilateral thalamic glioma are very rare, and only 17 cases have been reported to our knowledge (Estève F et al., 1999; Hirano H et al., 2000; Ruel JH et al., 1992; Uchino M et al., 2002; Yoshida M et al., 1998; Ziegler DK et al., 1977). The age of patients were ranged between 8 to 70 years with a mean of 39 years, however our patients was four years of age, being the youngest patient with the diagnosis of bilateral thalamic glioma in the literature.

The clinical signs and symptoms are mainly composed of personality changes, mental deterioration, memory loss, apathy, or emotional lability rather than focal neurological signs or signs of increased intracranial pressure (ICP) (Partlow GD et
The prognosis of bithalamic gliomas are very poor, leading to death within about one year following first presenting symptoms (Estève F et al., 1999; Yoshida M et al., 1998; Uchino M et al., 2002). Although headache and signs of ICP are frequent in unilateral thalamic masses, such symptoms are uncommon in bilateral thalamic glioma, especially at presentation (Uchino M et al., 2002). Only 2 of the previously presented cases had headache (Estève F et al., 1999; Ziegler DK et al., 1977), and only one exhibited moderate hydrocephaly (Ziegler DK et al., 1977). The presented case had enlarged ventricles on admission, and he also had sixth cranial nerve palsy, which has not reported before. The differences in presentation has been proposed to be related with the origin of the tumor (Smyth EG et al., 1938). According to this, peduncular, capsulothalamic tumors invading lateral or ventrolateral thalamus result in sensory disturbances at an early stage of the disease; while intrinsic thalamic gliomas originating in the subependymal glia in the third ventricle expand laterally from the medial nuclei and cause early mental deterioration (Smyth EG et al., 1938). It might be suggested that the presence of hydrocephaly may be related with subependymal origin, as anatomical relationship supports the expansile growth of the tumor in to the third ventricle, but not laterally towards internal capsule.

As in the presented case, the symmetrically distributed thalamic lesions revealed by MRI at an early stage favor the diagnosis (Yoshida M et al., 1998). Tissue density abnormalities can better be observed as diffuse hyperintense areas on T2-weighted images (Carter DJ et al., 1989; Rogers LR et al., 1994; Yanaka K et al., 1992). In reported case, typical MRI findings and progressive clinical picture aided the diagnosis of bilateral thalamic tumor, with the exclusion of other diseases such as mitochondrial cytopathies or acute necrotising encephalopathy.

MRS findings are also important in both diagnosis of bilateral thalamic tumors, and for the exclusion of other diseases of inflammatory, vascular or metabolic in origin. The proton MRS NAA signal is considered a marker of neuronal tissue (Luyten PR et al., 1990; Williams SR, 1992), and therefore expected to decrease in destructive lesions. The proton MRS choline levels correlate with membrane biosynthesis, especially in proliferating tissue (Luyten PR et al., 1990; Williams SR, 1992). The creatinin signal is either low or nearly unchanged in low-grade gliomas (Kuggel H et al., 1992). In our patient, the proton MRS NAA was decreased with normal choline, creatinin, and lactate levels, supporting low-grade glioma. Histological analysis of thalamic tumors is not easy, because diagnosis by biopsy, operation or autopsy is not always possible (Yoshida M et al., 1992). In our patient, we had the histopathological diagnosis of low-grade astrocytoma, confirming the diagnosis of bithalamic glioma radiologically.

In this report, we presented a 4-year-old boy presented as bilateral thalamic tumors manifesting with headache and the sixth cranial nerve palsy. This is the youngest patient with bilateral thalamic tumors so far reported in literature. This case shows bilateral thalamic tumors have a broad range of signs and symptoms. Although infrequent, typical radiological features of the tumor suggest the diagnosis in the presence of variable symptoms.

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REFERENCES