Case Report

Intraventricular glioblastoma multiforme: Case report

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ARTICLE INFO

Article history:
Received 18 March 2014
Accepted 6 September 2014
Available online 10 February 2015

Keywords:
Glioblastoma multiforme
Intraventricular tumor

ABSTRACT

Glioblastoma multiforme (GBM) is the most common primary brain tumor, but pure intraventricular location is extremely rare for GBM in neurosurgical practice. To our knowledge, there are only 19 reported cases to date. We present an additional case of intraventricular GBM with detailed clinical course, radiological and pathological findings.

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Glioblastoma multiforme intraventricular: informe de caso

RESUMEN

El glioblastoma multiforme (GBM) es el tumor cerebral primario más común que existe, pero una localización netamente intraventricular es muy rara en la práctica neuroquirúrgica. Según nuestro conocimiento, hasta la fecha solo se han publicado 19 casos intraventriculares. Presentamos un nuevo caso de GBM intraventricular con una descripción detallada de la evolución clínica y los hallazgos patológicos y radiológicos.

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1. Introduction

Glioblastoma multiforme (GBM) is a common primary brain tumor, which constitutes about 25% of all intracranial tumors in adults.1,2 Being known as the most malignant primary brain tumor, GBM may arise theoretically anywhere within the central nervous system (CNS).1,2 However, ventricular system is a very rare location for their occurrence. To our knowledge, there
are only 19 cases of intraventricular GBM reported to date.\textsuperscript{1-11} Herein, we report an additional case of intraventricular GBM, in order to discuss clinical and radiological findings, surgical strategy and clinical course.

2. Case report

A 65-year-old woman was admitted to the outpatient clinic, with severe headache, gait disturbance, drowsiness, vomiting and urinary incontinence. Her neurological examination revealed a slight paraparesis in lower extremity examination without reflex abnormality and bilateral papilledema. A non-contrast computed tomography (CT) scan was obtained and a right lateral ventricular mass lesion was seen. Magnetic resonance imaging (MRI) revealed an intraventricular, partly solid and partly cystic, rim-enhancing, 38 mm × 31 mm sized heterogeneous mass lesion around the body of the lateral ventricle (Figs. 1 and 2). The patient underwent surgery via posterior transclososal approach. The aim was to totally remove the mass lesion. The tumor seemed to arise from the medial occipital ventricular wall, and the lateral wall seemed to be intact. However, a severe arrhythmia occurred during surgery, which did not respond to medical therapy. The surgery had to be stopped after the advice of the anesthesiologist and cardiologist. Only a limited resection and biopsy were achieved. Histopathological examination of the tumor showed a mitotic index of 7/10, necrosis, atypical nuclei and endothelial proliferation (Figs. 3–5). Most of the tumor cells were stained positive for GFAP. The pathological diagnosis was in consistency with GBM WHO Grade 4. The patient was taken in to the coronary intensive care unit, for regulation of the cardiac arrhythmia. An additional surgery was planned in order to resect the rest tumor, but with the pathological diagnosis of GBM the patients’ family refused for a secondary surgery, after they were informed about the prognosis of the disease. The patient was discharged from the hospital without any additional neurological deficits. The patient received regional radiotherapy and chemotherapy with temazolamide afterwards. However, three months after surgery, the patient died.

3. Discussion

GBM is the most common and the most malignant primary CNS tumor in adults. This WHO Grade 4 tumor shows

Fig. 1 – Axial T1-weighted MRI showed a 38 mm × 31 mm sized mass lesion with heterogenous intensity and rim-like contrast enhancement.

Fig. 2 – Coronal T1-weighted MRI created a suspicion that the lesion arose from the medial occipital wall.

Fig. 3 – Photomicrograph shows focal necrosis and surrounding characteristic palisading necrosis. Hemotoxilen & Eosin (H&E), original magnification ×100.
nuclear atypia, high mitotic index, endothelial proliferation and necrosis. GBM constitutes approximately 25% of all intracranial tumors and 50% of all glial tumors in adults. Despite microscopic total surgical removal, radiotherapy and chemotherapy, the prognosis is worst among all intracranial malignancies with a median survival of 25–35 weeks. Cerebral cortex of frontal and temporal lobes are the most common locations for their occurrence. However, intraventricular GBMs are relatively rare. We did a PubMed search using the keywords “intraventricular glioblastoma multiforme” and “intraventricular GBM”. We investigated the results which contain related case reports or case series. We found out only 19 cases of intraventricular GBM reported to date.

In addition to these case reports some series may contain additional cases of intraventricular GBM; for example, a series of 112 patients with lateral ventricle tumors by Gokalp et al. contains five cases of GBM. Secer et al. presented nine cases of intraventricular GBM and stated that a total of 46 cases of lateral ventricle tumors were treated at their institution during the study period. The majority of the intraventricular tumors are benign and only 13% were reported as malignant lesions, including GBM, metastatic carcinoma, melanoma, etc. On the other hand, in a relatively large series of 267 patients with GBM by Stark et al. there was not even a single case of intraventricular GBM. The true incidence of intraventricular GBM among all intracranial tumors or among all GBM is still uncertain or varies greatly, but it is not wrong to say that these lesions are relatively rare.

A lesion that originates from the ventricular wall or from the structures within the ventricle is considered primary ventricular tumor and a lesion that originates from the adjacent brain parenchyma and exophytically grows into the ventricle is considered secondary ventricular tumor. Lee and Manzano stated that the intraventricular GBM may arise from the neoglial cells of septum pellucidum or fornix. Especially, fornix and the limbic system due to their close relationship with the ventricular system have been postulated as the origin of the ventricular GBM formation. Kim et al. underlined the importance of subependymal zone and the pluripotent stem cells within. We also think that abnormal glial proliferation of neural stem cells might be the cause of intraventricular occurrence of GBM.

Intraventricular GBMs have typical MRI findings such as irregular borders, heterogeneous or ring-like contrast enhancement, a broad surrounding edema on T2-weighted images and central hypointensity which is indicative of necrosis. Our case also shared the same radiological features. GBM usually presents in the sixth or seventh decade of life with a median age of 61 years. Usually, these tumors do not cause any symptoms or signs until they reach a considerably large size. The most common clinical presentations are due to the signs of increased intracranial pressure, signs that related to obstruction of CSF circulation or signs caused by compression of eloquent areas. Our case was presented with similar symptoms and signs.

The surgical approaches that may be used for the resection of lateral ventricle tumors include a large variety. Transcallosal and transcortical approaches are most commonly used. The aim of the surgery is to increase length of survival as well as the quality of life of the patients. Gross debulking compared to minimal resection and biopsy provides better results. However, there are controversial results about the prognosis of patients with intraventricular GBM. Especially, worse clinical results and poor prognosis were reported for some pediatric cases. On the contrary, Sarsilmaz et al. reported 24 months of disease-free survival of a 16-year-old boy and “butterfly” recurrence afterwards. Secer et al. reported a median survival of 18.8 months in a series of nine patients. This number is considerably long compared to survival of parenchymal GBM patients, considering that total resection was only achieved in one of these nine patients. We aimed a total resection via posterior transcortical approach as the MRI suggests that the lesion more likely arose from the medial occipital ventricular wall. However, due to the above-mentioned cardiac problems during surgery, only a limited subtotal resection and biopsy could be achieved in our case. Therefore, the prognosis was poor with only three months of survival.
4. Conclusions

Intraventricular GBM is relatively rare. GBM usually shows rapid progression and recurrence after treatment. However, intraventricular GBM might have a better prognosis in adults. Therefore, gross total resection should be aimed to increase the length of survival and quality of life of these patients.

REFERENCES