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CASE REPORT

Intraventricular glioblastoma multiforme mimicking meningioma and review of the literature

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ABSTRACT

Glioblastoma multiforme (GBM) is the most common primary brain tumor and is usually found in aged persons in the cerebral hemispheres particularly the frontotemporal region. But intraventricular GBM is rare and only few cases have been reported in the literature. We report a case of a 27-year-old man who presented with headache, vision loss in both eyes, and other signs and symptoms of increased intracranial pressure. Computed tomography and magnetic resonance imaging showed an intraventricular, well-circumscribed lesion with *homogeneous* enhancement of contrast, suggestive of meningioma that is more common than GBM in this location. The patient underwent surgical removal through transcortical route. The final pathologic diagnosis was GBM. We present the clinical features, radiological findings, and surgical management of this case and discuss the pathogenesis and review of the literature of intraventricular GBM.

Key words: Glioblastoma multiforme, intraventricular, meningioma

Introduction

Intraventricular tumors are uncommon, accounting only 1-10% of all brain tumors and include a variety of benign tumors, malignant tumors, and cyst formations. Glioblastoma multiforme, although, is the most common primary tumor of brain accounting for nearly 20-25% of cases, it rarely occurs within ventricles. They are actually secondary tumors of ventricles arising from the structures adjacent to it and secondarily invading into the ventricles in an exophytic manner. There have been reports of glioblastoma multiforme in trigone, body, and occipital horn of the lateral ventricle[1-7] and in the third ventricle.[8] However, we could not find any report of frontal horn glioblastoma multiforme (GBM) in the literature. We report a case GBM in a young male patient showing radiological features suggestive of a more common benign lesion such as meningioma. The lesion was completely excised and the histopathological study was consistent with GBM. There is a gross difference in the postoperative adjuvant therapy as well as overall prognosis of these two lesions.

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Case Report

A 27-year-old male patient presented to us with headache for last 2 years. The headache was gradually increasing in severity and since last 5 months, has become intense with associated intermittent vomiting. There was progressive decline in visual acuity in both eyes for last 3 months and patient became blind 1 month prior to admission. On physical examination, the patient was conscious, oriented with loss of perception of light in both eyes. Optic fundi showed secondary optic atrophy. There was no other significant neurologic finding.

Computed tomography (CT) scan showed a well-marginated, iso- to hyperdense lesion in frontal horn and body of right lateral ventricle with intense, homogeneous enhancement on contrast. MRI showed a well-circumscribed lesion with regular margins showing gross enhancement and compressing the foramen of Monroe producing hydrocephalus [Figures 1a-d]. MRS showed the decreased NAA level with the elevated choline peak. But the ratio choline/creatinine was less than 2 which

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was not typical of GBM. Ratio more than 3 is suggestive of a highly malignant lesion such as GBM.

A right frontal transcortical approach taken. A highly vascular, soft, friable tumor found protruding into the frontal horn of right lateral ventricle. Near total tumor removal done in piece meal. Histopathology showed highly pleomorphic tumor cells with increased cellularity, endothelial cell proliferation, and nuclear palisading around central zone of necrosis suggestive of glioblastoma multiforme [Figures 2 and 3]. Immunohistochemistry showed strong positivity toward Glial Fibrillary Acidic Protein [Figure 4]. The final diagnosis was glioblastoma multiforme.

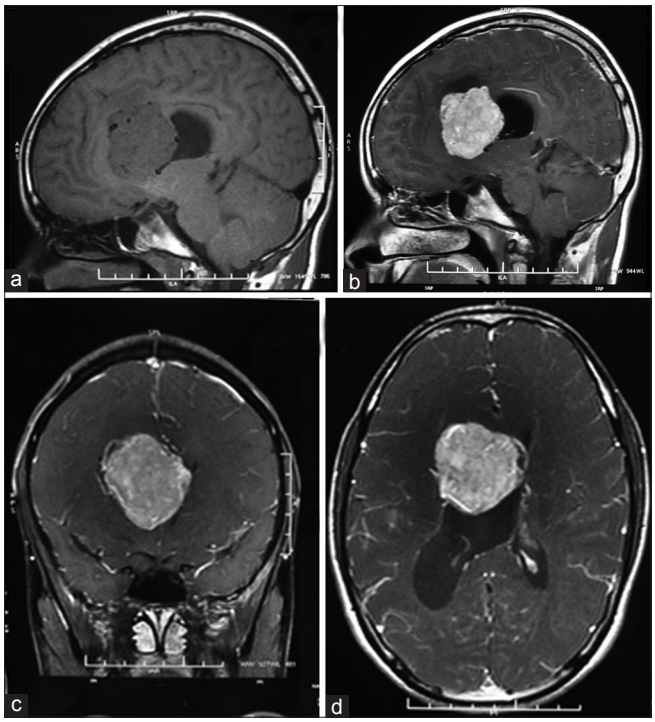

Figure 1: (a) T1-weighted MRI showing the tumor in frontal horn (b) T1 weighted sagittal MRI with gadolinium contrast showing well circumscribed tumor with intense enhancement (c) Coronal contrast MRI (d) Axial contrast MRI of the tumor showing the extension of tumor to opposite frontal horn and body

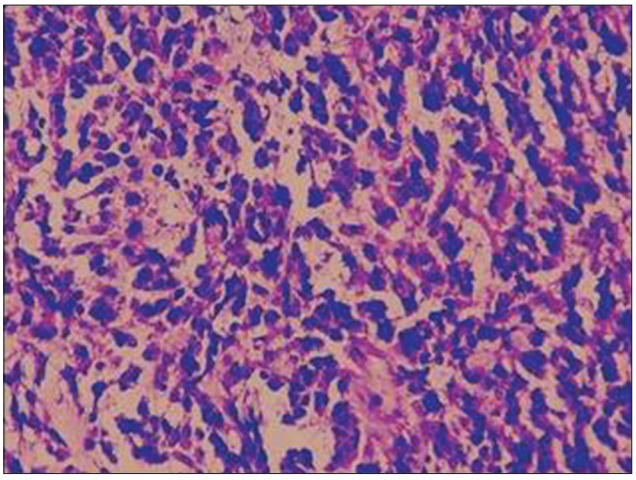

Figure 3: Highly pleomorphic tumor cells and increased cellularity

Discussion

Glioblastoma multiforme is the most common primary brain tumor in adults and accounts for 15-20% of all intracranial tumors and approximately 50% of gliomas. Although it can arise anywhere in the CNS, nearly two-thirds are found in the fronto-temporal region. But, intraventricular GBM is relatively rare. It has been mentioned to be usually found predominantly in the frontal horn or body of lateral ventricle in some reports; however, such finding we could not find in any literature.

Intraventricular tumors have been classically divided into two types according to their origin: Primary and secondary. Neoplasms that arise from the ventricular wall and its lining or from structures within the ventricles are considered primary ventricular tumors, and ependymoma, choroid plexus papilloma, choroid plexus carcinoma,

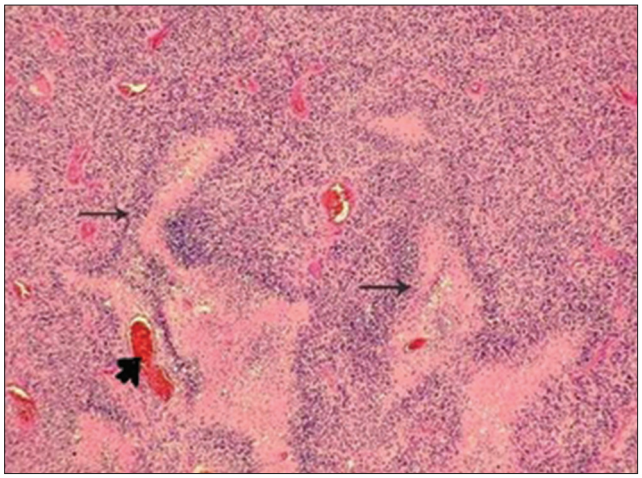

Figure 2: Nuclear palisading around areas of necrosis (arrow marks) with endovascular proliferation (arrow head)

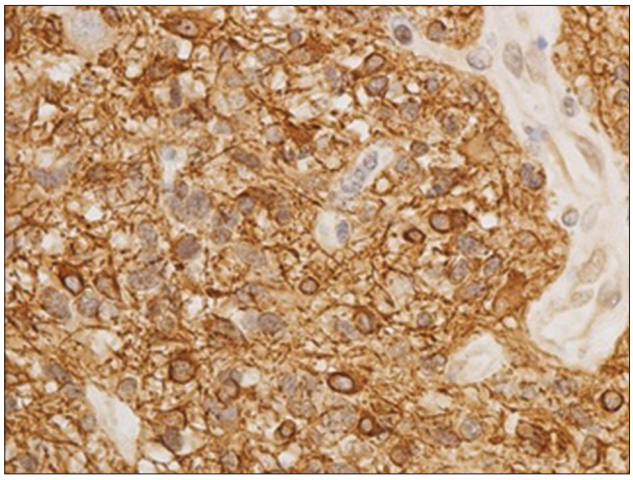

Figure 4: Immunohistochemistry showing intense Glial Fibrillary Acidic Protein positivity

meningioma belong to this group. Tumors that arise from structures adjacent to the ventricle and gradually enlarge, so that more than two-thirds of them reside within the ventricle, are considered secondary ventricular tumors with transependymal development. These tumors have their origin primarily from the cerebral tissue. Gliomas that include astrocytomas, subependymal giant cell astrocytomas, other less common variants, including choroid glioma, glioblastoma multiforme, and mixed glial neuronal tumors are included in this group.

The area just below the ependymal lining of the ventricles is called the subventricular zone (SVZ) and contains pluripotent stem cells that can give rise to ependymal cells as well as various cells upon stimulation. According to Doetsch *et al*. this zone contains as many as four distinct cell types; ependymal cells, astrocyte like type B cells, type C cells, and oligodendrocyte progenitors. Genetic mutation followed by uncontrolled proliferation could lead to pathogenesis of malignant tumors such as glioblastoma multiforme in this rarer site. Once originated, these tumors could secondarily invade the ventricle through transependymal migration.

Most lateral ventricular tumors enlarge slowly and typically cause no symptoms until reaching a size large enough to cause obstructive hydrocephalus or compression of surrounding eloquent structures. Due to their aggressive behavior, glioblastomas usually become symptomatic within a short interval. This usually manifests as headache, vomiting, and other signs of increased intracranial pressure. In this case the extreme degree of it led to complete blindness due to secondary optic atrophy.

Radiologically, the glioblastomas usually show irregular, infiltrative borders with heterogeneous enhancement in form of rim or peripheral enhancement of tumor tissue and areas of necrosis. However, in this case there was a well defined, round to oval, well circumscribed lesion showing *homogeneous* intense enhancement. This feature in a relatively young patient with long-standing symptoms strongly suggested the diagnosis as meningioma. Intraoperatively the tumor was highly vascular, friable with lots of neo-vascularization suggestive of a malignant lesion. Histopathology confirmed the nature of lesion as glioblastoma multiforme. We could not find a report of glioblastoma in the frontal horn of lateral ventricle in the literature. Although Park *et al*.[4] and Kim *et al*.[2] mentioned that the glioblastoma in intraventricular location are common in frontal horn or body, we could not find any report of a GBM in frontal horn in the literature. In both of their cases the tumor was located in trigone area. Sarsilmaz *et al*.[5] reported a pediatric GBM located in posterior body and occipital horn of the left lateral ventricle. Seccer *et al*.[6] reported nine cases of intraventricular GBM, out of which seven were in body and rest arising in septum pellucidum.

Conclusion

Intraventricular tumors are relatively rarer, heterogeneous group of tumors, presenting with nonspecific signs and symptoms delaying their diagnosis. Glioblastoma multiforme, although the most common primary intracranial tumor, is relatively uncommon in this location in comparison to their benign counterparts. In addition, they can present quite differently from their intracerebral component, both in clinically and radiologically, thereby making their preoperative diagnosis difficult. This case illustrates this by resembling a meningioma, which shows a great difference in the prognosis. So glioblastoma multiforme should be kept in the differential diagnosis of intraventricular tumor during preoperative management planning.

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Conflicts of interest

There are no conflicts of interest.

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