A Rare Case Report of Central Neurocytoma

Abstract

Central neurocytoma (CN) is a slow-growing, benign brain neoplasm with a favorable prognosis and, using clinical and imaging findings, one can significantly limit the differential diagnosis. They develop preponderantly in young adults, most often in the lateral ventricles. This report describes a case of CN in a 19-year-old patient, the signs and symptoms, imaging, histopathology including immunohistochemistry, and the treatment applied.

Keywords: *Central neurocytoma, lateral ventricle, prognosis*

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Introduction

Central neurocytoma (CN) is a slow-growing, benign neoplasm, typically located in the lateral ventricles of brain, near the foramen of Monro, with a characteristic attachment to the septum pellucidum, mainly affecting young adults and bearing a favorable prognosis.[1] It was first described by Hassoun *et al*. in 1982.[2] We report here the case of a 19-year-old boy with CN who had a large, intraventricular brain mass with characteristic imaging features on magnetic resonance imaging (MRI). He underwent surgery and no adjuvant therapy was considered. After 3 years, when the residual tumor started increasing in size, he was treated with radiotherapy.

Case Report

An otherwise well, 19-year-old boy, presented with a headache and diminished vision in both eyes for 1 year. Ophthalmic examination revealed diminished visual acuity in both eyes and right abducens nerve palsy. Fundoscopy revealed papilledema in both eyes, which was suggestive of increased intracranial pressure. Other physical and neurological examinations were normal. Contrast-enhanced MRI (CE MRI) of brain revealed a large (63 mm × 50 mm × 49 mm), lobulated lesion in the third ventricle and extending to the left lateral ventricle which was hyperintense on T2-weighted (T2W) image, hypointense on T1W image, and moderate heterogeneous intense enhancement of lesion with dilatation of both lateral ventricles [Figure 1]. Then, he underwent endoscope-assisted transcortical transventricular near-total excision of tumor in the neurosurgery department. Histopathology and immunohistochemistry (IHC) were suggestive of atypical neurocytoma (WHO Grade II). Tumor cells were immunopositive for synaptophysin, Microtubule Associated Protein-2 (MAP-2), neu N, and immunonegative for GFAP and IDH 1. MIB labeling index was 6%. Imaging in postoperative period showed residual disease [Figure 2]. He was not offered any adjuvant therapy owing to its indolent nature and subsidence of symptoms. Three years postsurgery, follow-up CE MRI brain showed increased size of residual lesion [Figure 3]. Hence, he was administered radiation dose of 54 Gy in 30 fractions over 6 weeks by three-dimensional conformal radiotherapy.

Discussion

Initially described in 1982 by Hassoun *et al*.,[2] CN is a rare neuronal tumor which corresponds to 0.25%–0.5% of brain tumors.[3] The initial description classified them as WHO Grade I lesions; however, this was upgraded in 1993 to WHO Grade II as it was recognized that at least some of these tumors exhibited more aggressive behavior.[4] CN typically affects young adults around the third decade. Our patient is a 19-year-old adolescent. Central Neurocytoma is characteristically located in the supratentorial ventricular system. Nearly 50% of the cases involve the lateral

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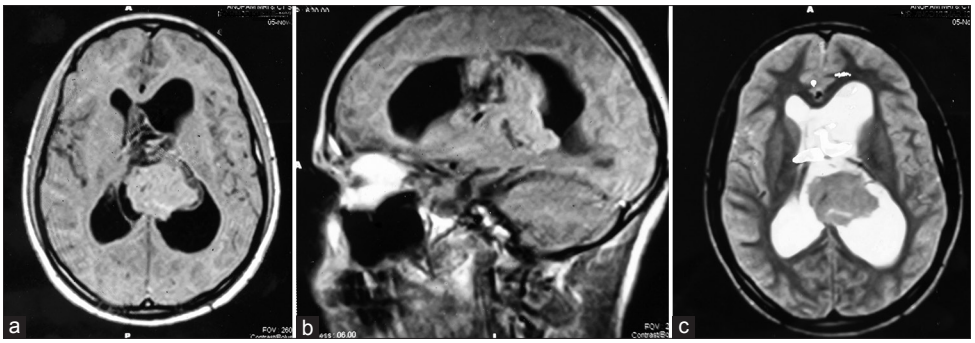

Figure 1: Contrast-enhanced magnetic resonance imaging showing tumor in (a) T1-weighted (axial view; postcontrast), (b) T1-weighted (sagittal view), (c) T2-weighted (axial view) images

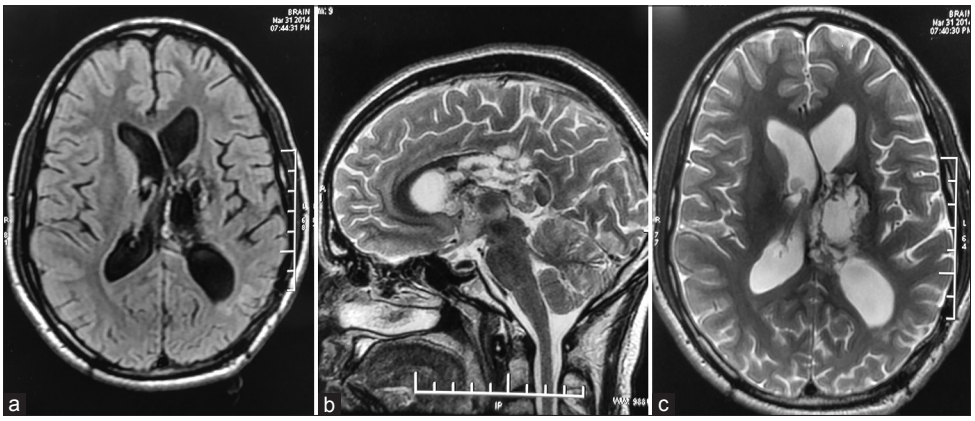

Figure 2: Contrast-enhanced magnetic resonance imaging showing residual tumor in (a) T2-weighted fluid-attenuated inversion recovery (axial view), (b) T2-weighted (sagittal view), (c) T2-weighted (axial view) images

ventricles near the foramen of Monro, whereas 15% are located in both the lateral and third ventricles. About 13% of CNs are bilateral and only 3% occur in the third ventricle as an isolated location. Our patient presented with tumor at the third ventricle which extended to the left lateral ventricle. There is a single case report of a CN that arose in the fourth ventricle.[5] They typically present with signs and symptoms of increased intracranial pressure induced by obstructive hydrocephalus,[6] as is in our case. On MRI, CN is typically heterogeneous on all sequences, but it is isointense to cerebral cortex on T1W images and isointense to hyperintense on T2W images. Areas of low signal intensity or absent signal on both T1W and T2W images can represent calcification, cyst, hemorrhage, and tumor vessels. Most tumors show some degree of enhancement.[7] The presented case fit into the radiological features of CN as reported in the literature. Histopathologically, the nuclei of these neurocytes are round or oval with finely speckled "salt and pepper" chromatin. Neurocytes are typically not immunoreactive for Glial Fibrillary Acidic Protein (GFAP). The diagnosis must be based on IHC for neuronal antigens such as synaptophysin and neuron-specific enolase.[8] The majority of CN are benign though 25% of cases are more aggressive with an MIB-I labeling index more than 2% and atypical histological features. These cells are strongly immunoreactive for synaptophysin.[9] The current case perfectly fits to the pathological features of CN as evident in literature.

CNs have good prognosis. As a benign tumor of slow growth, CN still has surgery with complete resection as its gold standard treatment. Patients with incomplete excision may benefit from radiotherapy.[6] In contrast with the more aggressive atypical neurocytomas, well-differentiated neurocytomas are associated with an excellent long-term survival. Our patient lived with residual disease postsurgery without any progression for 3 years. The decision to proceed with irradiation needs to be made on the patient's risk tolerance for needing another craniotomy and considerations of potential radiation toxicity. Three years postsurgery, when our patient had radiological progression, he was subjected to radiotherapy to a total dose of 54 Gy which appears to be sufficient.[10]

Conclusion

CN is a benign intraventricular brain tumor that typically originates from the neuronal cells of the septum pellucidum and bears an excellent prognosis. By using clinical, demographic, and imaging findings, one can significantly limit the differential diagnosis for most of these intraventricular tumors. Surgery with gross total resection is the most preferable option, correlated with the best

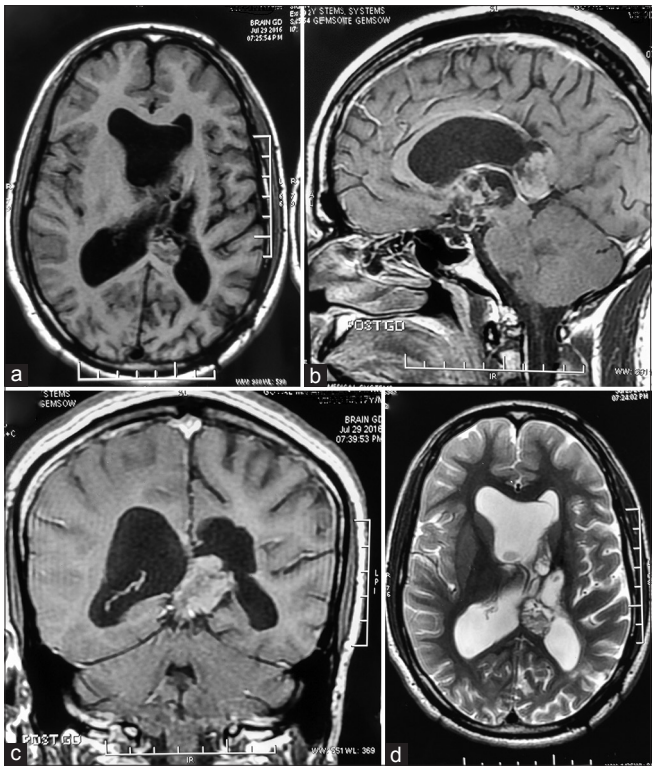

Figure 3: Contrast-enhanced magnetic resonance imaging showing increased residual tumor size in (a) T1-weighted (axial view), (b) T1-weighted (sagittal view; postcontrast), (c) T1-weighted (coronal view; post contrast), (d) T2-weighted (axial view) images

long-term survival rates and local tumor control. Adjuvant radiotherapy may be considered for residual CN following subtotal resection, large CN size, or CNs near inoperable regions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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