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Central neurocytoma of the third ventricle: Case report and treatment review☆

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ABSTRACT

Central neurocytoma is a rare tumor of neuronal differentiation which is typically seen in young adults. Here, we report a case of a 25 years old female patient who presented with central neurocytoma of the third ventricle. Sub-total resection of the tumor was carried out via left parieto-occipital craniotomy. Afterwards, the patient received conventional radiotherapy (i.e., 54 Gy/27 fractions). We observed marked improvement in terms of tumor size reduction (i.e., from 5.2×4.8 cm^2 to 3.4×2.1 cm^2; ~71%) and patient performance status.

Introduction

Central neurocytomas (CN) are rare tumors of neuronal differentiation which are typically seen in young adults. Such tumors when located within the ventricles of the brain are called *intraventricular* while those arising/present in the cerebral hemispheres and spinal cord are called *extraventricular* neurocytomas. Having a relatively good prognosis, early recognition of these tumors is crucial towards more favorable treatment outcomes. Radiological studies, in particular magnetic resonance imaging (MRI), provides the first step in diagnosis of these tumors [1]. However, histopathology ascertains the definite diagnosis. The prominent features of these tumors have been elucidated [2,3], which, among others, include: morphology comprising cells with uniform round contours, round nuclei with fine speckled chromatin, cell clusters with nuclear free fibrillary areas mimicking neurophil and delicate arborizing capillaries and foci of calcification. At the therapeutic end, total surgical resection is considered as the treatment of choice [3–5]; nevertheless, radiotherapy [3–7] and chemotherapy are sometimes used in adjuvant settings [3,8,9], particularly when total resection is not possible.

Herein, we present a case of intraventricular neurocytoma (IVN), assess the response of our treatment protocol (i.e., sub-total tumor resection followed by conventional radiotherapy) and review the treatment options from the available literature. Specifically, we report a case of 25 years old female diagnosed with IVN. The patient showed marked improvement to sub-total tumor resection followed by radiotherapy.

Case report

A 25-year-old female presented to the radiology department with the complaints of headache for three months duration. The patient was blind from both eyes. The Eastern Cooperative Oncology Group (ECOG) performance status of the patient was one at the time of presentation. Magnetic resonance (MR) images of the brain demonstrated tumor in the lateral ventricular region (Fig. 1). Specifically, MR images revealed moderately enhancing abnormal signal intensity area involving the septum pellucidum and affecting the anterior part of both lateral ventricles, as presented in Fig. 1. Moderate dilation of posterior parts of lateral ventricles was also noted. No further differential diagnostic or therapeutic work up was done for nine months, due to patient negligence. After nine months, the patient again presented to the radiology department; repeat MR study illustrated a large heterogeneously enhancing mass of size 5.1×3.8 cm^2 in the midline region, with bulk of mass at the left. Specifically, the mass occupied the region of third ventricle. Further, mild communicating hydrocephalus was also noted. The mass also affected adjacent ventricles. However, posterior fossa of the brain stem was normal (Fig. 1). Sub-total resection

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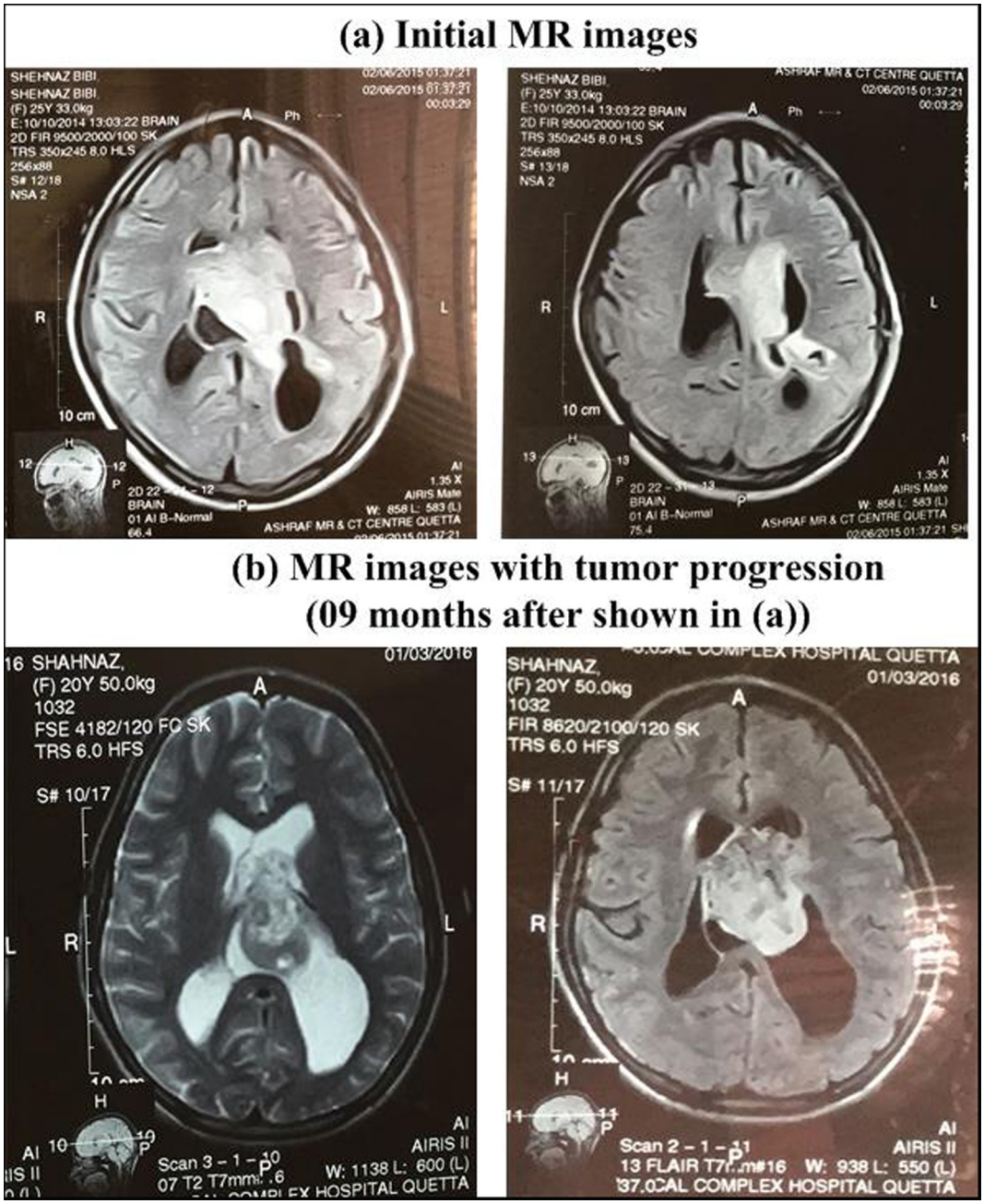

Fig. 1. Pre-treatment magnetic resonance (MR) images of the brain showing tumor in the lateral ventricular region. MR images with largest tumor size are shown only.

of the tumor was implemented via left parieto-occipital craniotomy.

Histopathology revealed fragments of a neoplastic lesion (size = 2 × 1.5 cm^2) composed of small to intermediate sized cells arranging in the form of sheets seen around arborizing blood vessels (Fig. 2). The individual neoplastic cells had scant cytoplasm and pleomorphic nuclei with open coarse chromatin showing few (approximately 1/10 HPF) mitoses. In foci, fibrillary nuclear free zones were seen. However, definite necrosis was not identified. Immunohistochemical stains showed following patterns; GFAP and EMA were negative, synaptophysin was diffuse positive and Ki 67 (Mib-1) was low. These features favored central neurocytoma, WHO grade II, according to WHO classification of central nervous system (CNS) neoplasms [10]. The summary of clinical, radiological and histopathological features has been presented in Table 1.

MR study after one month of craniotomy demonstrated evidence of mass (size = 5.2 × 4.8 cm^2) in the septum pellucidum and frontal horn of third left lateral ventricle. It was isointense on T1 and intermediate signal on T2 and FLAIR images. Moreover, there was evidence of minimal dilatation of all the ventricles (Fig. 3). Re-surgery was refused by the surgeon at this stage.

Conventional radiotherapy alone was decided at the therapeutic end. Specifically, 54 Gy in 27 fractions was delivered via two lateral opposing portals. Chemotherapy was not included in the treatment protocol. The tumor was re-assessed radiologically after three months of radiotherapy completion. Post-radiotherapy MR images illustrated mass in the frontal horn of left lateral ventricle with

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