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Case Report

Oncology Section

A Rare Clinical Scenario of Central Neurocytoma of Brain among Two Young Adult Patients

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ABSTRACT

The Central Neurocytoma (CN) is a rare tumour of brain which is commonly seen in young adults. Two cases, A 24-year-old-male and a 37-year-old-female with CN of the lateral ventricles are reported here. Both patients presented with complaints of headache and seizures with no focal neurological deficits. Gross total excision of tumour was carried out via right frontoparietal craniotomy. Afterwards, they received adjuvant radiotherapy by Intensity Modulated Radiotherapy (IMRT), 56Gy in 28 fractions for residual lesion. There was marked improvement in terms of tumour response and improvement of patient's performance status and quality of life.

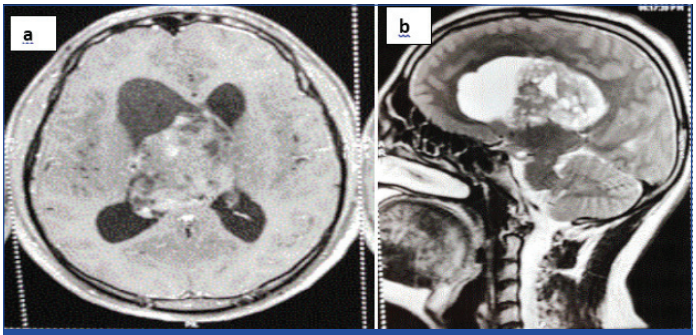
Keywords: Frontoparietal craniotomy, Radiotherapy, Tumour, Ventricle

CASE REPORT

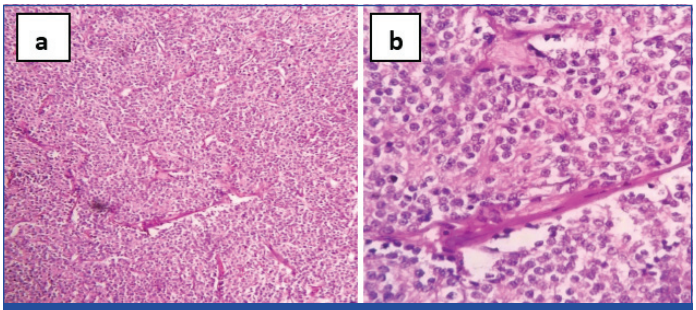
Case 1

A 24-year-old-male patient complained of headache for two months duration and seizures of two episodes in the previous month. No familial history of malignancies, seizures and brain tumours were there.

Magnetic Resonance Imaging (MRI) brain findings revealed large mixed intensity lesion in intra ventricular location in lateral ventricle area in midline with areas of calcification, solid and cystic areas measuring 4.4×7.1×4.6 cm causing moderate hydrocephalus [Table/Fig-1a,b] and asymmetric dilatation of lateral ventricles. Features were in favour of ependymoma with differential diagnosis of craniopharyngioma. Patient underwent right frontoparietal craniotomy and gross total excision of tumour. Postoperative histopathological report was suggestive of Central Neurocytoma World Health Organisation (CN WHO) grade II [Table/Fig-2a,b].

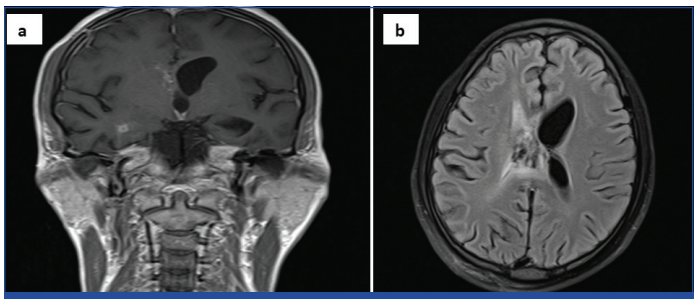


[Table/Fig-1]: Preoperative MRI showing the lesion with moderate hydrocephalus and in intraventricular location: (a) Axial view; and (b) Sagittal view.



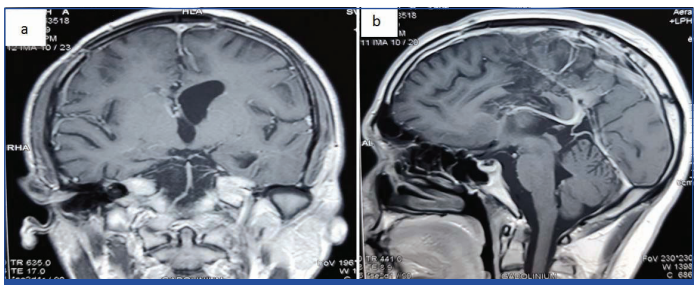
[Table/Fig-2]: (a) Under H&E section low power view showed cells arranged diffusely with intervening capillaries (10x); and (b) High power view showed cells are round to oval, monomorphic nucleus speckled chromatin (40x).

The patient later underwent ventriculo-peritoneal shunt two months after surgery as he developed tubercular meningitis and was offered anti-tubercular drugs. Postoperative MRI findings [Table/Fig-3a,b], done after three months, revealed residual lesion in right lateral ventricle measuring 1.5×2.6 cm.



[Table/Fig-3]: Postoperative MRI showing residual lesion: (a)coronal view; and (b) Axial view.

He was planned for adjuvant radiotherapy by IMRT and was given a total dose of 56 Gy in 28 fractions at 2Gy/fraction. He had marked symptomatic improvement and response in terms of tumour size and there was no residual lesion seen in follow-up Magnetic Resonance Imaging (MRI) [Table/Fig-4a,b], five months after radiotherapy.



[Table/Fig-4]: Postradiotherapy MRI coronal (a) and sagittal (b) views.

Case 2

A 37-year-old female, with no co-morbidities, was evaluated for complaints of headache, weakness of lower limbs and seizures of one month duration. There were no cranial nerve palsies. No familial history of malignancies and brain tumours.

MRI brain findings revealed large heterogenous space occupying lesion in midline within the lateral ventricles measuring 3.5×5.2×4.5 cm and extending to both sides, more towards right lateral ventricle with indentation over 3rd ventricle and dilated occipital and temporal horn of right lateral ventricle. She underwent right frontal craniotomy with

middle frontal gyrus approach and decompression. Postoperative histopathological report was suggestive of CN WHO grade II.

Postoperative MRI showed good decompression with enlarged right ventricle and temporal horn, areas of right-sided parenchymal bleed was seen. She was planned for adjuvant radiotherapy by IMRT. Patient was given adjuvant radiotherapy with a total dose of 56 Gy in 28 fractions at 2Gy/fraction. Patient tolerated the treatment well and discharged under stable condition. Patient had marked symptomatic improvement and response in terms of tumour size as no evidence of residual detected in follow-up MRI five months after radiotherapy.

DISCUSSION

The CN is a group of rare brain tumours with characteristic indolent growth [1], usually seen in adolescents and young adults. It 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 [2], mainly affecting young adults and bearing a favourable prognosis [3]. It was first described by Hassoun J et al., in 1982 [4]. Incidence is rare and comprises only 0.25-0.5% of brain tumours [5,6]. CN typically affects young adults around the third decade [7].

It is characteristically located in the supratentorial ventricular system. Nearly, 50% of the cases involve the lateral 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 [8]. The index patients presented with tumour in the lateral ventricles. Usually, patients present with signs and symptoms of increased intracranial pressure induced by obstructive hydrocephalus [8].

Histopathologically, the nuclei of these neurocytes are round or oval with finely speckled salt and pepper chromatin. Neurocytes are typically not immune reactive for Glial Fibrillary Acidic Protein (GFAP) [9]. The diagnosis must be based on Immunohistochemistry (IHC) for neuronal antigens such as synaptophysin and neuron-specific enolase [9].

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 [10]. Surgery remains the main stay of treatment [11]. However, addition of adjuvant radiotherapy improves response rates and survival. Chemotherapy seems to have limiting role in the management of neurocytomas [12].

CONCLUSION(S)

To conclude, Central Neurocytoma (CN) being an extremely rare, ordinarily benign intraventricular brain tumour may be managed successfully by a combined modality approach of surgery and adjuvant radiotherapy to achieve better local control and survival. However, multi institutional studies and longer follow-up is needed to validate this therapeutic approach.

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