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Case Report on Neurocytoma

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Introduction

Among all the brain tumors, Neurocytoma is the utmost rare intraventricular brain tumor that affects younger age population. It was first described in 1982 by Hassoun et al. as a midline neuronal tumor located in the intraventricular region seen frequently among young adults (1). Despite their increased recognition, central Neurocytoma still remain rare tumors of the central nervous system, comprising only 0.25–0.5% of all the brain tumors(2). The main features of central Neurocytoma are their location in the ventricular region with specific neuronal origin, specific radiological characteristics, their resemblance to oligodendroglioma or Ependymoma on light microscopy and most importantly, their favorable prognosis with benign biological behavior .

Case Report

A patient 8 year old male, initially presented with complaints of headache which were followed by multiple episodes of vomiting for the past one year. The patient had experienced 3 episodes of seizures in the past 3 months.

Primary Radiological evauation

MRI Brain was done which showed an ill defined hyper intense T1 and T2 weighted mass lesion in the periventricular region with predominant cystic component and peripheral soft tissue component showing heterogeneous areas of contrast enhancement in the periphery with peripheral calcification restricted diffusion and mass defect.

Surgical Intervention

The patient underwent Right frontal craniotomy and tumor decompression on 23rd February 2017.

Histopathological Evaluation

The frozen section of the stained tumor specimen showed features of Glial tumor with differential diagnosis of 1. Astrocytoma 2. Oligodendroglioma.

The post operated H and E stained tumor specimen histopathology revealed the tumor to be composed of shears of uniform cells with a clear cytoplasm and small, round nuclei through an abundant fibrillary stroma. There were multiple calcifications seen. Homer-Wright rosettes were not present. In view of resemblance of the h and e stained tumor cells to oligodendroglima, the slides were reviewed at Tata Medical College, which were consistent with features of extraventricular neurocytoma (WHO grade II).

Radiological Investigation after Surgery

Post operated CT Scan of Head was done which showed craniotomy defect with bone flap replacement involving bilateral parietal bones with drainage tube along with underlying post operated changes.

The patient then presented to the Department of radiation Oncology for further management.

At presentation, the Kernofsky Performance Status of the patient was 80%, conscious, well oriented to time, place and person. Power in all four limbs were maintained with score of 5/5. Bowel and bladder control of the patient was intact. The patient however had left 6th cranial nerve palsy.

MRI Brain showed Hyper intense signals measuring 1.6 x 1.4 cm in right frontal lobe with underlying Post operated changes.

Radiation Details

The case was discussed and patient was planned for Adjuvant Radiotherapy. Patient received External Beam Radiotherapy 36Gy/18# to Brain on 6 MV Linear Accelerator, from 3 April 2017 to 6 May 2017 in the Department of Radiation Oncology at Himalayan Institute of Medical Science, Dehradun

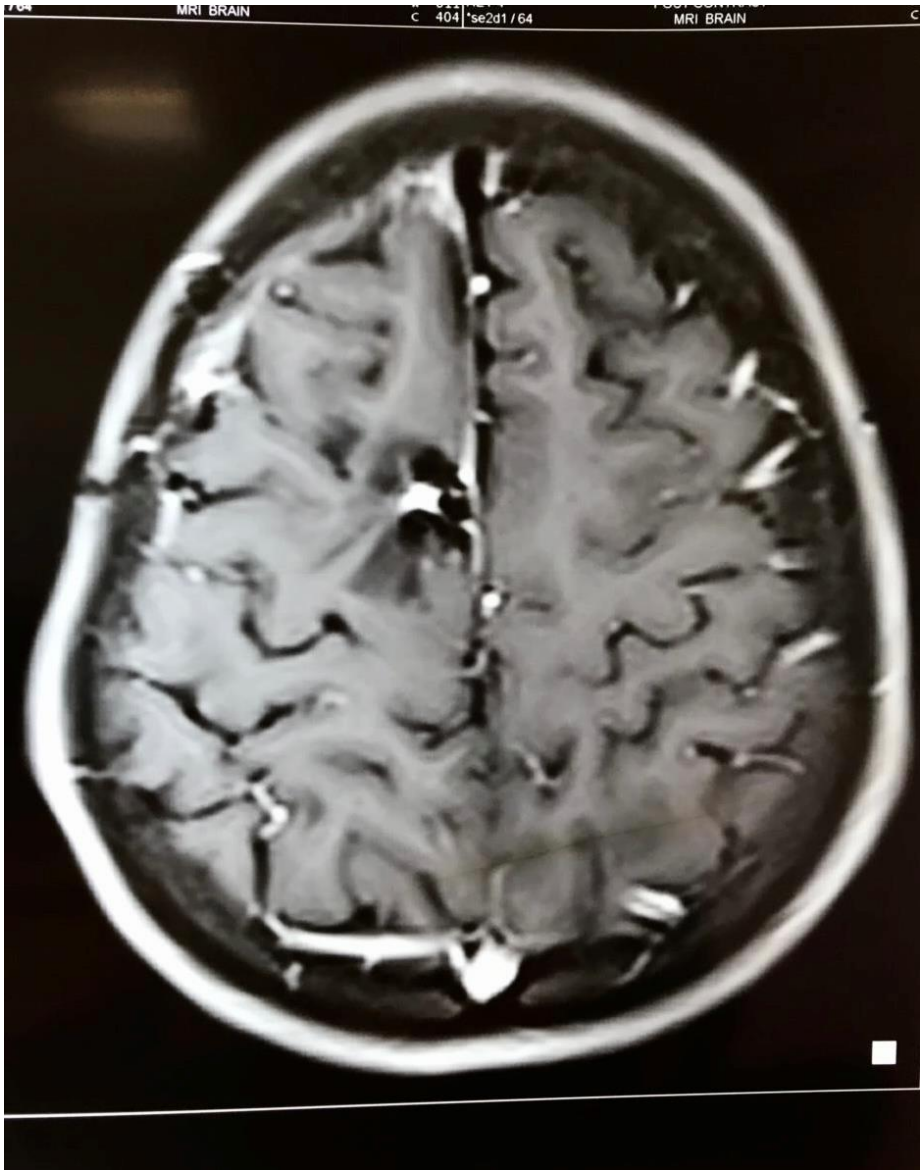
The patient is on regular follow Up with anti epileptic support with Tablet Leveracetam 250 mg once daily.

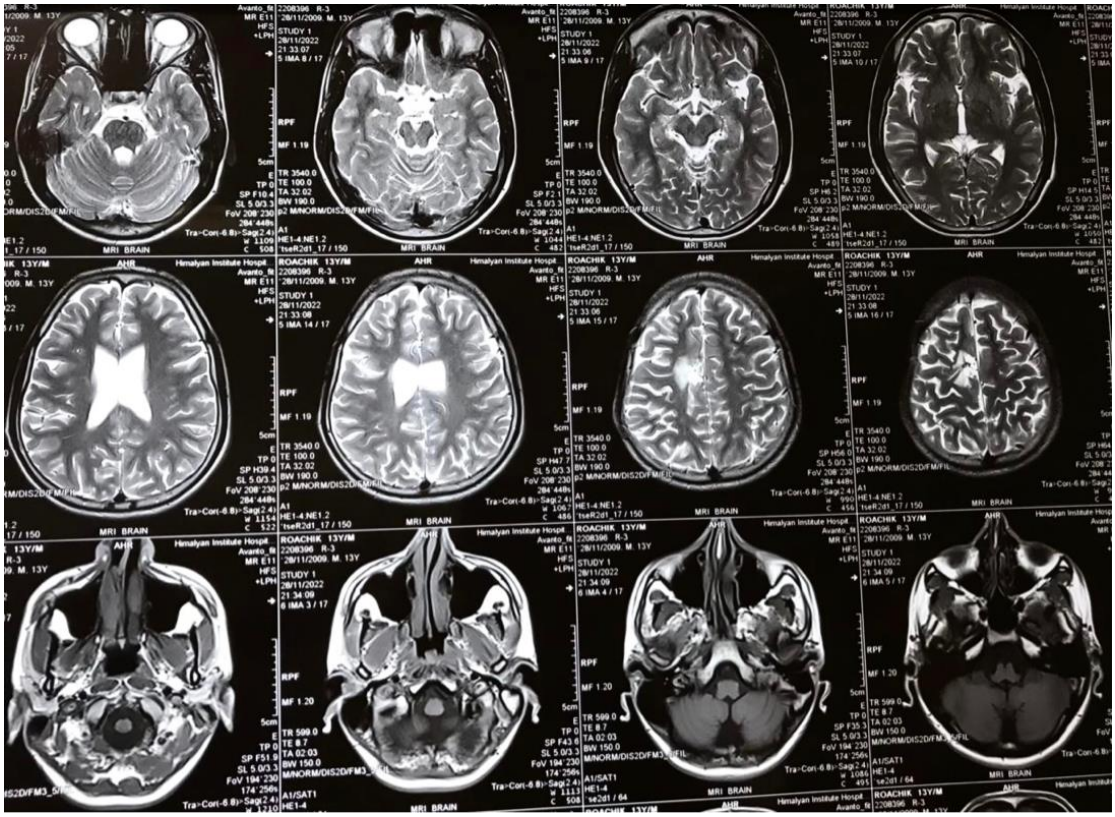
Follow up

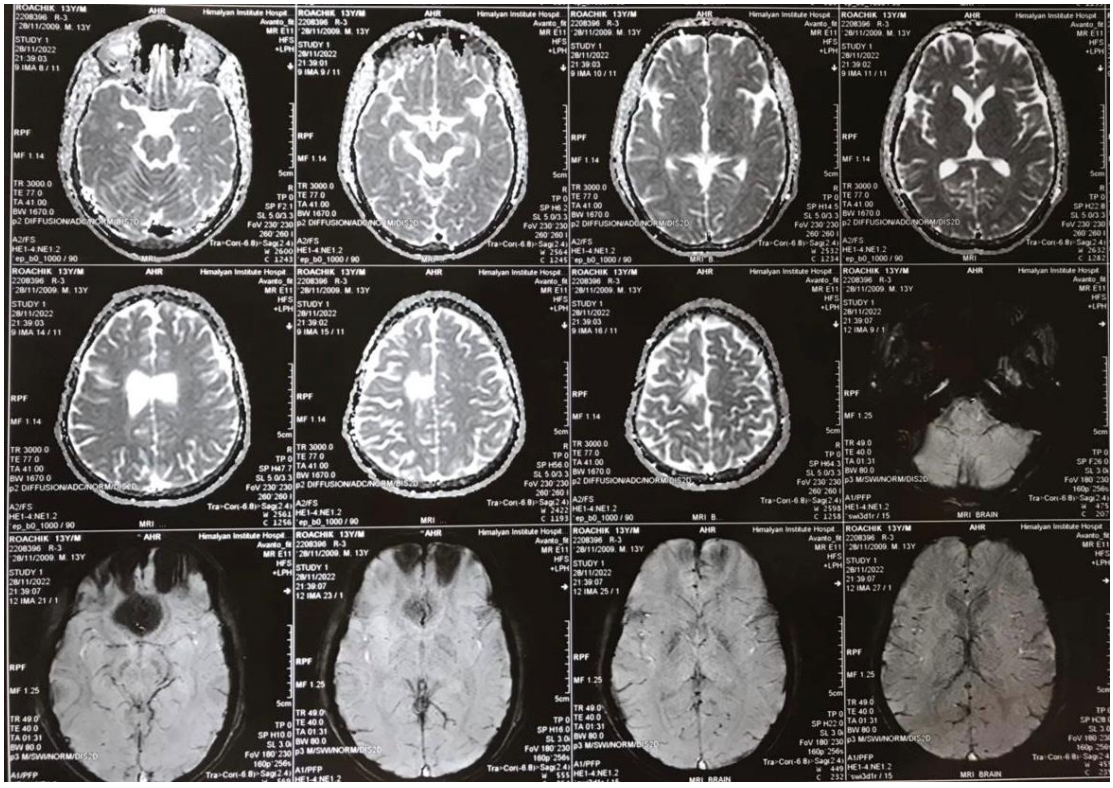
The patient is on regular follow up.

On examination: Kernofsky performance status of the patient is 90 percent, conscious, well oriented to time, place and person. Power in all four limbs were maintained with score of 5/5. Bowel and bladder control of the patient was intact. The patient did not show any neurological deficit.

Follow up MRI Brain was done which showed no evidence of disease.







Discussion

Neurocytoma is the a rare intra ventricular brain tumor which was first described by Hassoun et al. in 1982 as a midline neuronal tumor located in the intra ventricular region seen frequently among younger population(1). The tumor comprises only 0.25–0.5% of all the brain tumors(2). The tumor is known to arise from the septum pellucidum, fornix, or the walls of the lateral ventricles due to increased intracranial pressure(3). The main features of Neurocytoma are their location in the ventricular region with specific neuronal origin, specific radiological characteristics, their resemblance to oligodendroglioma or Ependymoma on light microscopy and most importantly, their favorable prognosis with benign biological behavior. Although central neurocytoma closely resembles oligodendroglioma under the light microscope, electron microscopy reveals clear differences such as origin of the neurocytoma cells from neuronal cell, with others features such as uniform cells with clear cytoplasm, small and round nuclei, and dense fibrillary stroma(4). Mitosis is not common and there is generally no pleomorphism. Therefore, These observations indicate that the tumor is benign and differs from other neuronal tumors, especially neuroblastoma.

Neurocytoma is seen frequently in younger population, affecting both the genders equally. Due to raised intracranial pressure, the patients commonly present with severe headache vomiting and gait disturbances(5). In severe cases, the patients may also present with features of hemiparesis, seizures or hemorrhage. No specific risk factors or genetic predilections have been identified .

Management of Neurocytoma is a multimodality approach, which includes Surgical resection, (with different approaches such as trans-callosal, transcortical, and endoscopic resection) and Adjuvant Radiotherapy. The choice of surgical approach is largely dependent on the surgeon experience and tumor location and therefore, the surgical approach should be individualized. In general, surgical resection of tumor carries challenge because of its huge size, location in the deep midline close to critical intraventricular structures, and occasional hypervascularity of the tumor. Therefore, the trans-callosal approach is mainly performed to operate in a tumor in the third ventricle or both lateral ventricles with normal ventricular size and to avoid cortical incision. In recent advances, neuro-navigation system has been developed to provide better accuracy, better localization and better delineation of brain lesions, therefore, ensuing maximum safe resection.

Adjuvant Radiotherapy has been used to reduce the chances of recurrences in young individuals. According to a study conducted by Mahavadi et al, the recurrence rate after gross total resection was

23.9%, whereas 6.9% was the recurrence rate after gross total resection with radiotherapy(6). Therefore, the study concluded that the Adjunctive radiotherapy reduces the risk of tumor progression after incomplete tumor resection and improve patient overall survival rate with maximum safe resection, which is 93.2% in al level compared to surgery alone(7). In our patient, due to intraventricular location of the tumor, the transcortical approach was favored over trans-callosal approach by the neurosurgery team, and a subtotal tumor resection was accomplished via transcortical approach because of the high vascularity of the tumor. Postoperatively, the patient was managed with Adjuvant Radiotherapy.

Conclusion

Despite being a well recognized tumor of the brain effecting the younger population, Neurocytoma is a rare entity comprising only 2.5 percent of all the brain tumors(2). It is a slow growing benign neoplasm of the central nervous system that has an excellent prognosis. Although the best long term survival rates and local tumor control correlates with total resection which is the most preferable management, Adjuvant Radiotherapy is considered following subtotal resection with residual Neurocytoma.

Therefore, the case of 8 year old male, presented with headache, vomiting and multiple episodes of seizures Neurocytoma should always be considered as a differential diagnosis in slow growing neoplasms located in the ventricles. A multimodality treatment approach with surgery and Adjuvant Radiotherapy should always be kept in mind. An attempt of Total safe resection should be carried out . In cases where a total resection is not feasible, a subtotal resection should be done followed by Adjuvant radiotherapy.

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