

CASE REPORT

Pan ventricular neurocytoma

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ABSTRACT

Central neurocytomas are intraventricular benign neoplasms located within the lateral ventricle adjacent to foramen of Monro. A rare case of a central neurocytoma extending throughout the entire ventricular system at the time of presentation is reported. The clinical course of this tumor in a 25-year-old male and its management are presented with relevant review of the literature.

Key words: Central neurocytoma, intraventricular tumors, panventricular, transcallosal

Introduction

Intraventricular tumors account for 10% of central nervous system neoplasms. The pathological characteristics of the tumor vary with patient age and location. Central neurocytomas are intraventricular tumors located within the lateral ventricles, usually next to the foramen of Monro and septum pellucidum. They may extend into the third ventricle and periventricular brain parenchyma. A pan ventricular extension of a neurocytoma as noted in the present case is rare, and such tumors are difficult to treat.

Case Report

A 25-year-old male presented with progressive weakness of both lower limbs since 3 months. He had headaches and vomiting 1-week prior to admission. On neurological examination, the patient had spastic paraparesis. Both fundi revealed papilledema. Magnetic resonance imaging demonstrated a well-defined lobulated intraventricular mass lesion occupying lateral ventricles, third ventricle, fourth ventricle and extending into cisterna magna through foramen of magendi [Figures 1 and 2]. The tumor literally formed a cast

of the entire ventricular system. The tumor was isointense on T1-weighted images, hyperintense on T2-weighted images and significant flow voids were noted. On contrast administration, heterogeneous enhancement was seen. The patient underwent right fronto-parietal parasagittal craniotomy with transcallosal radical excision of the tumor within the lateral ventricles. The tumor was soft, easily suckable and extremely vascular. Haemostasis was achieved with great difficulty. The patient deteriorated in sensorium within a few hours after surgery and had to be put on ventilatory support. Postoperative computed tomography scan demonstrated postoperative changes with residual tumor in third and fourth ventricle. There was no hydrocephalus. The patient was treated conservatively with decongestants and anticonvulsants. The patient succumbed on second postoperative day. Histological examination confirmed the lesion to be a central neurocytoma.

Discussion

Central neurocytomas (WHO Grade II) are low-grade, slow-growing primary brain tumors of neuronal origin. They comprise 0.25–0.5% of all primary brain tumours.^[1] They develop predominantly in young adults between second and fourth decades.

The typical central neurocytoma is located in the supratentorial ventricular system adjacent of the foramen of Monro. Tumor extension into the third ventricle occurs in 26% of central neurocytomas. Isolated involvement of third or fourth ventricle and even extra-ventricular sites are rarely reported. There are case reports of central neurocytomas with cranio spinal and extra central nervous system dissemination.^[2] Holo ventricular involvement with extra ventricular extension of a central neurocytoma as noted in the present case is extremely rare and has been earlier described only once.^[3]

Although pan ventricular involvement by neoplastic process is a rare event, however such cases have been reported

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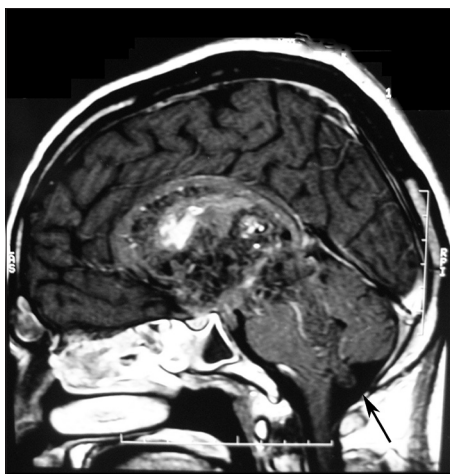


Figure 1: Postcontrast sagittal magnetic resonance image showing a heterogeneously enhancing tumor involving the lateral, third and fourth ventricles. Tumor is noted to extend into the cisterna magna (arrow)

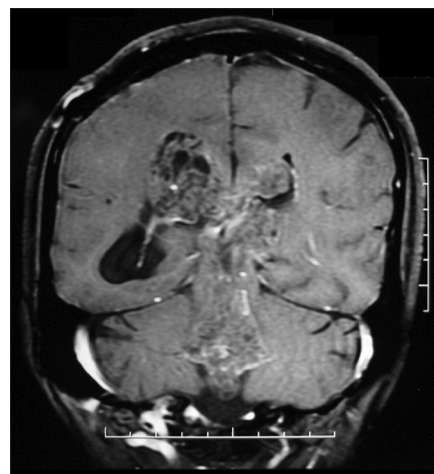


Figure 2: Postcontrast coronal magnetic resonance image demonstrates tumor extension into both lateral ventricles. It extends through the third ventricle into the fourth ventricle

before. Hercules *et al.* have described a multicentric low-grade astrocytoma involving all four ventricles.^[4] Similarly, a case of en plaque ependymoma involving ependymal lining of whole ventricular system was described by Muzumdar *et al.*^[5] Roche *et al.*, described a giant pan ventricular neurocytoma with extension to the interpeduncular and prepontine cisterns.^[3] The clinical significance of such pan ventricular extension remains an enigma.

Gross pathology of central neurocytoma is that of a lobulated well circumscribed, gray color mass with cystic areas and few calcifications. On light microscopy, they have small homogenous uniform cells with rounded nuclei and scant cytoplasm resembling perinuclear halos giving fried egg appearance. Immunostaining for neuron-specific enolase and synaptophysin confirm the neuronal nature of the neoplasm. Electron microscopy demonstrates clear dense core vesicles, microtubules, and synapse formation.

The treatment options of extensive tumors involving different compartments of the ventricular system such as a panventricular central neurocytoma remain a neurosurgical challenge. Surgical decompression, establishment of histological diagnosis and appropriate reduction of intracranial pressure either by tumor removal or cerebrospinal fluid diversion are the main treatment goals. As seen in the present case, these tumors can be highly vascular and can extensively involve choroidal vasculature. In the present case as more tumor was removed more extensive bleeding was encountered with inability to control choroidal blood supply. Few lessons were learned and worth noting while dealing with such extensive intraventricular lesions. It is always advisable to study major blood supply in advance and take early control of blood supply. A postoperative ventricular drain is advisable in such cases with residual tumors. Lastly, a

multistage resection can be planned to minimize morbidity related to long extensive surgery. However, the extensive spread of a benign tumor is a formidable challenge even for a multi-stage removal. Stereotactic-guided neurosurgery, endoscopic surgery and neuronavigation have improved the rate of the total tumoral resection in recent times.^[1] Surgical treatment of these tumors should take into consideration the invasion of the periventricular tissue.

Several authors report extent of resection as a key factor in the prognosis. A subtotal resection would lead to higher rates of recurrence and lower survival.^[1] Extra ventricular extension has been considered to have a bad prognosis. The adjuvant use of radiation therapy and chemotherapy in subtotally resected central neurocytomas is not universally accepted. A pan ventricular central neurocytoma is a rare case. Such a case is a neurosurgical challenge and tumor excision may be achieved by multistage procedures.

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