Central Neurocytoma- A Rare Brain Tumor

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Abstract

Central neurocytoma are rare, slow growing, intraventricular tumors of neuronal origin, typically located in the lateral ventricles, near the Foramen of Monroe with characteristic imaging features. They generally occur in young age with favorable prognosis. With clinical, histological and immunohistochemical background, we report a case of central neurocytoma.

Introduction

Central neurocytoma is a slow growing, benign neoplasm with a favorable prognosis and affects mainly young adults.[1-3] These low grade and slowly growing primary brain tumor were firstly described by Hassoun et al in 1982.[4] They comprise 0.25% to 0.5% of all primary brain tumors.[5] There is no reported gender predilection.[6] Central neurocytoma is typically located in the lateral ventricles, near the Foramen of Monroe. We report a case of central neurocytoma histologically and immunohistochemically proved in 33-year-old female.

Case Report

33-year-old female, no major comorbidities, presented with chief complaints of headache since 15 days which got worsened a day back was associated with non-bilious vomiting and diplopia. Headache was diffuse in nature and present throughout the day. She had no history of fever or loss of consciousness.

On examination, she was well nourished and adequately built. She was afebrile, pulse was 80 beats/min, regular, blood pressure in right brachial artery 120/80mm of Hg. Neurological examination did not reveal any focal deficit or any cranial nerve involvement. Laboratory parameters were within normal limits. Within few hours, she became unconscious with decorticate rigidity, pulse rate dropped to 40 beats/ min, pupils were semi dilated bilaterally with sluggish reaction to light. She was shifted to Intensive Care Unit (ICU), was intubated and mechanical ventilatory support given due to irregular breathing, features suggestive of acute decompensated hydrocephalus. Her MRI brain could not be done due to acute deterioration of neurological condition and hence CT brain was done and patient was taken for surgery without waiting for MRI brain.

Computed Tomography of Brain revealed hyper dense lesion in mid line in the region of Foramen of Monroe measuring approximately 2.1 X 1.6cm, causing third ventricle obstruction leading to bilateral lateral ventricles hydrocephalus and peri ventricular ooze with diffuse cerebral edema (Figure 1).

Right Parasagittal craniotomy was done, decompression of tumor was done and biopsy taken and was sent for histopathological examination. Macroscopically it was multiple small gray white pieces of tissue. Microscopically, the tumor composed of monotonous round cells with perinuclear halos, nuclei contain fine chromatin granules. On immunohistochemistry, tumor cells were positive for Synaptophysin, Neuron specific Enolase and GFAP (Glial fibrillary acidic protein), negative for Chromogranin A (Figure 2) and Ki-67 less than 5% in maximum proliferating area. Histological and immunohistochemical profile compatible with diagnosis of central neurocytoma.

Discussion

Central neurocytoma is a rare WHO grade II neuroepithelial intraventricular tumors, constituting only 0.25%- 0.50% of all intracranial tumors.[5] Initially described in 1982 by Hassoun et al, central neurocytoma is a rare tumor of neuroglial origin.[4] 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.[6] These tumors typically affect young adults around third decade. They are characteristically located in the supratentorial ventricular system. Half of the cases involve the lateral

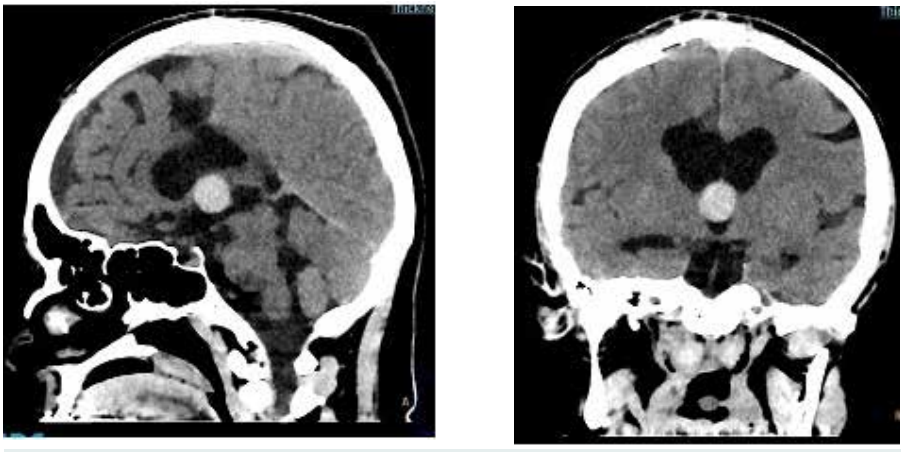

Fig. 1: Computed tomography showing hyper dense lesion in mid line in the region of Foramen of Monroe causing dilatation of bilateral lateral ventricles

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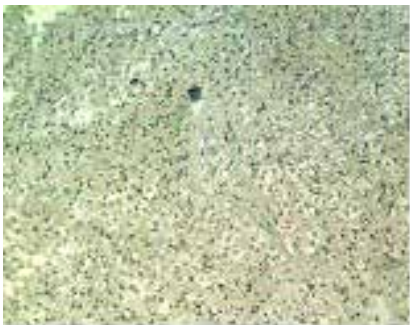
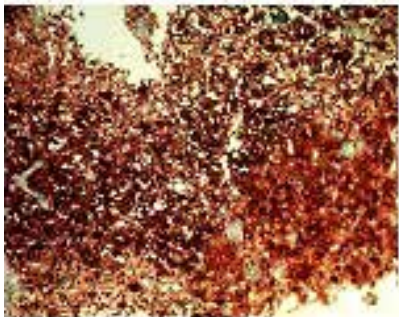
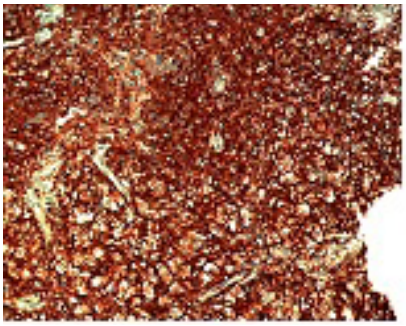


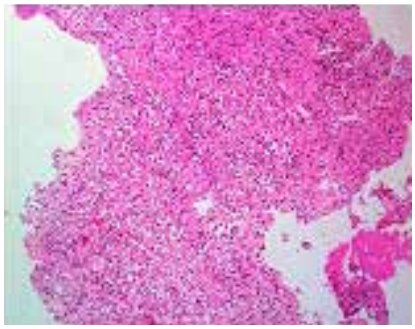
Chromogrannin A negative



NSE positive



Synaptophysin positive



H and E stain

Fig. 2: Histology and immunohistochemistry markers of Central neurocytoma

ventricles near the Foramen of Monroe, whereas 15% are located in both the lateral and third ventricles. About 13% of central neurocytoma are bilateral and only 3% occur in third ventricle as an isolated location.[7]

The typical clinical presentation is with signs and symptoms of increased intracranial pressure induced by obstructive hydrocephalus. Patients may present with acute symptoms related to sudden development of ventricular obstruction and elevated intracranial pressures, generally there is insidious onset of symptoms. Schild et al analyzed 27 patients with central neurocytomas regarding their presenting symptoms, and 93% of patients complained of headaches, 37% had visual changes, and 30% experienced nausea and vomiting at presentation.[8] In another study by Wang et al, out of 27 patients, 21 presented with headache and 6 with vomiting.[9]

The imaging method such as CT and MRI are used to evaluate location and assist on the diagnosis of the tumor, but the definite diagnosis is established by pathology analysis (electron microscopy and immunohistochemical studies).[10] Based on location and histomorphology, the differential diagnosis of masses located in ventricular system, are oligodendroglioma, ependymoma and neuroblastoma.[11] In light of the cellular monotony, peri nuclear halos and frequent calcification, oligodendroglioma becomes the principal entity in the differential diagnosis. In most of the cases, these two are virtually indistinguishable at H and E stain. Immunostaining for Synaptophysin is the simplest way to distinguish neurocytoma from oligodendroglioma since the latter is nonreactive.[11]

Central neurocytomas carries good prognosis. The best treatment for central neurocytoma appears to be complete surgical resection. Patient with incomplete excision may benefit from radiotherapy.[12] Well differentiated neurocytomas are associated with good 5-year survival rate.

Conclusion

Central neurocytomas are slow growing, rare, benign intraventricular tumors of neuronal origin. The diagnosis is established by typical location of tumor, histology and immunohistochemistry. The treatment of choice is complete surgical removal of tumor, radiotherapy in incomplete excision. In our case of typical neurocytoma, confirmed with microscopic examination and immunohistochemistry, treated with surgical decompression of tumor.

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