Case Report

Pediatric central neurocytoma: Case report and review of literature

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ABSTRACT

Central neurocytomas are slow-growing primary brain tumors of neuronal origin having a predilection to arise mostly in the lateral ventricles. We report a case of a 9-year-old girl who presented with headache and vomiting of 1-month duration. Her magnetic resonance imaging was suggestive of central neurocytoma of the third ventricle and was surgically managed, and tumor tissue was sent for histopathology and immunohistochemistry which confirmed the diagnosis.

Key words: Central neurocytoma, immunohistochemistry, synaptophysin

Introduction

Central neurocytomas are slow-growing neuronal primary intracranial tumors, found mostly in young adults, usually located in the lateral ventricles.[1,2] Pediatric presentation of central neurocytoma is very rare, and no significant data have been published regarding their incidence. Rades et al. in their series of neurocytoma in children reviewed 59 patients, which is so far considered the largest case series of this rare neoplasm of children under 18 years, reported only 13 (22%) patients who were in the first decade.[3] We are reporting this case because of its rarity, especially in this age and its location. Our case is of a 9-year-old girl who was diagnosed with central neurocytoma of the third ventricle.

Case Report

A 9-year-old previously healthy girl presented with headache of 1-month duration, which was continuous and holocranial in nature. There was also associated nausea and several episodes of vomiting for the past 1 month. On detailed clinical examination, the only positive finding was mild bilateral papilledema. The vital signs were within normal limits, and there were no focal neurological deficits. Her blood reports were unremarkable.

On magnetic resonance imaging of the brain, a well-defined globular mass with mixed intensity was noted within the third ventricle, thereby causing its distension. The tumor measured 47 mm × 59 mm × 55 mm in craniocaudal and craniocaudal dimensions. Bilateral thalamic compression was noted along with upstream hydrocephalus. On T2 imaging [Figure 1b], the mass appeared heterogeneously hyperintense, and on T1-weighted [Figure 1a], a hyperintense area with multiple cystic areas was noted. The solid part of the lesion showed diffusion restriction. Postcontrast the tumor showed enhancement [Figure 1c]. Magnetic resonance spectroscopy revealed high choline and glycine peaks with low N-acetylaspartate peaks.

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Cite this article as: Baishya BK, Singh RK, Dutta D, Hussain Z. Pediatric central neurocytoma: Case report and review of literature. J Pediatr Neurosci 2016;11:348-50.
The patient was operated with right frontoparietal craniotomy using the anterior interhemispheric transcallosal approach. Corpus callosotomy of around 2 cm was done and the choroid plexus was seen and followed to reach the foramen of Monro, and the tumor was approached transforaminally. Total removal of the tumor was achieved by piecemeal excision through the foramen of Monro. Watertight dural closure was done using Vicryl 4-0. Bone flap was replaced, and wound was closed in layers after achieving absolute hemostasis. Intraoperatively, the tumor was found to be globular, pale, grayish-white, soft, suckable, and moderately vascular. The tumor sample was sent for histopathologic examination and immunohistochemistry (synaptophysin and neuron-specific enolase [NSE]).

On histopathology [Figure 2a], small sheets of atypical round-/oval-/epithelial-like cells with enlarged round/oval/hyperchromatic nuclei with occasional nucleoli with definite cellular-nuclear pleomorphism were seen, and rare mitotic activity was noted. On immunohistochemistry [Figure 2b and c], both synaptophysin and NSE were positive helping us to arrive at the diagnosis of central neurocytoma.

The patient was discharged without any neurological deficits and with relief of headache on the 10th postoperative day. Till date, the patient is on regular follow-up with us clinico-radiologically.

Discussion

Central neurocytoma was described in 1982 by Hassoun et al.\(^4\) The incidence of central neurocytoma is only 0.25%–0.5% of all brain tumors.\(^5\) Initially, it was termed as a WHO grade I tumor and was updated to grade II in 1993.\(^6\) Characteristically, they are located in the ventricular regions supratentorially. Fifty percent are located in the lateral ventricles, 13% in both lateral and third ventricles, and solitary third ventricle central neurocytomas amount to only 3%.\(^7\)

Classically, central neurocytomas present with features of increased intracranial pressure associated with obstructive hydrocephalus. In a study, Schild et al. reviewed 27 patients of which 93% had headache, 37% had visual problems, and 30% had nausea and vomiting as their chief complaints,\(^8\) which was similar to the complaints of our patient.

The most commonly used approaches are anterior transcallosal approach and anterior transcortical approach.\(^9\) In our case, the previous approach, i.e., the anterior transcallosal approach was done.

Central neurocytoma is confirmed diagnostically by immunohistochemistry for neuronal antigens such as synaptophysin and NSE. Among the two, synaptophysin is most remarkable and NSE is considered nonspecific.\(^10\) Both these tests were positive in our study.

Central neurocytomas generally carry a good prognosis. The ideal treatment is the total surgical removal of the tumor. Radiotherapy might prove beneficial in adult cases, in which there is incomplete tumor removal although it is still a matter of concern in children.\(^3\)

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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