Original Research Paper

Volume - 15 | Issue - 04 | April - 2025 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Oncopathology

UNRAVELING A RARE CASE OF CENTRAL NEUROCYTOMA: A HISTOPATHOLOGICAL AND IMMUNOHISTOCHEMICAL EVALUATION

Dr. Chinju Thomas*	Post Graduate, Department of Pathology, JNMC, Belagavi. *Corresponding Author
Dr. Ramesh Chavan	Professor, Department of Pathology, JNMC, Belagavi.
Dr. Sajo Elamattom	Senior Resident, Department of Pathology, JNMC, Belagavi.

ABSTRACT Central neurocytomas are rare brain tumors, accounting for approximately 0.25% to 0.50% of all brain tumors. They typically affect young adults between the ages of 20 and 40. These tumors are predominantly intraventricular, most commonly located in the lateral or third ventricles near the foramen of Monro. The clinical presentation is primarily due to increased intracranial pressure and includes symptoms such as headache, nausea, vomiting, seizures, visual disturbances, and papilledema. Accurate diagnosis and histological confirmation are essential, with immunohistochemical studies aiding in the differentiation from other similar neoplasms. Surgical excision is crucial for achieving favorable outcomes.

KEYWORDS : Central neurocytoma, intraventricular tumor, neurocytic rosettes.

INTRODUCTION

Central neurocytoma (CN), first described by Hassoun et al. in the 1980s[1], is a rare intraventricular tumor accounting for 0.25–0.5% of central nervous system neoplasms, typically affecting young adults aged 20–40[2]. It commonly arises near the foramen of Monro within the lateral or third ventricles. Although the exact cell of origin remains uncertain, proposed sources include neuronal cells, neuronal progenitor or stem cells, and multipotent precursor cells. Clinically, CN presents with symptoms of raised intracranial pressure such as headache, nausea, visual disturbances, and hydrocephalus. Diagnosis involves neuroimaging and histopathology. Gross total resection (GTR) offers excellent prognosis and low recurrence.

CASE STUDY

A 28-year-old female presented with a two-day history of severe headache, along with vomiting and giddiness. She also reported progressive blurring of vision over the past 7–8 days. Notably, she had a history of intermittent headaches over the last 3 months, but there was no significant personal or family history. On examination, the patient was vitally stable, and her general and systemic examinations were unremarkable. Ophthalmologic evaluation revealed signs of chronic papilledema, suggesting raised intracranial pressure.

Neuroimaging with CT **(Fig:1)** of the brain demonstrated a large intraventricular mass lesion in the right lateral ventricle, characterized by heterogeneous enhancement, with internal cystic and necrotic areas as well as calcified specks. There was a significant mass effect, causing deviation of the septum pellucidum and resulting in obstructive hydrocephalus, with evidence of periventricular cerebrospinal fluid (CSF) seepage and cerebral edema. Based on radiological features, a diagnosis of an intraventricular neoplasm was considered. The differential diagnoses included central neurocytoma, glioma, ependymoma, oligodendroglioma, glioblastoma (GBM), and pineocytoma.

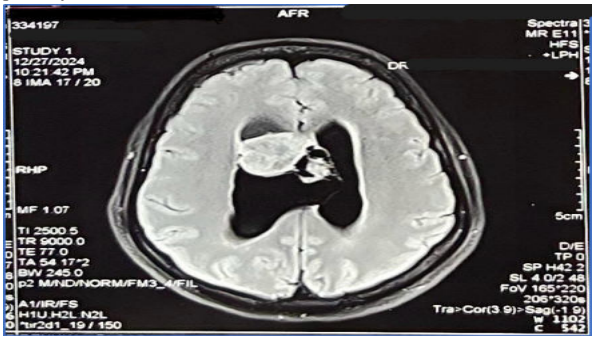

Fig 1: MRI Brain shows a large right intraventricular mass with enhancement, cystic areas, calcifications, deviation.

The patient underwent gross total resection, and the specimen was received in the histopathology department. Gross examination revealed multiple grey-white to grey-brown soft tissue fragments of varying sizes. Microscopic evaluation **(Fig:2, 3, 4)** showed a benign neoplasm composed of small to medium-sized uniform cells arranged predominantly in sheets, with occasional linear patterns. The tumor cells exhibited indistinct, clear cytoplasm and round nuclei with regular contours, finely stippled "salt and pepper" chromatin, and occasional micronucleoli. A characteristic feature was the presence of perinuclear clearing in most tumor cells.

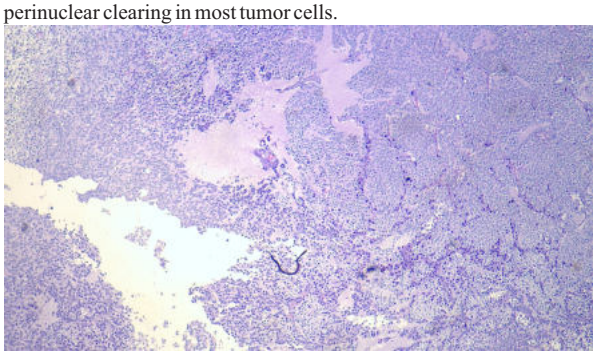

Fig 2: Pictomicrograph showing Moderately cellular neurocytic tumor with uniform cells, pseudorosettes, fibrillary background (H&E, **X10)**

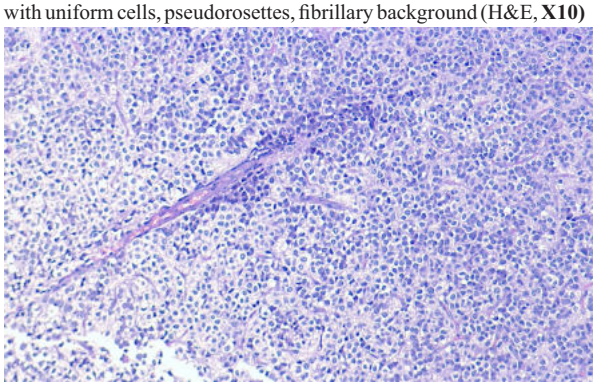

Fig 3: Pictomicrograph showing Uniform neurocytic cells with pseudorosettes and fibrillary neuropil-like background (H&E, **X40)**

Numerous arborizing capillaries were observed, along with occasional hyalinized blood vessels. Areas of fibrillar neuropil-like matrix were present, in addition to neurocytic rosettes and occasional perivascular pseudorosettes. No calcifications were identified. These histomorphological features were suggestive of central neurocytoma, WHO Grade , II, oligodendroglioma or ependymoma.

Immunohistochemistry was done for final diagnosis which showed diffuse cytoplasmic positivity for synaptophysin **(Fig:5)**, whereas S100 **(Fig:6)** was negative, which rules out Oligodendroglioma, confirming neuronal differentiation. GFAP negative and S100 negative, rules out Ependymoma. Additional markers such as NeuN and Olig-2 may be used to further support the diagnosis.

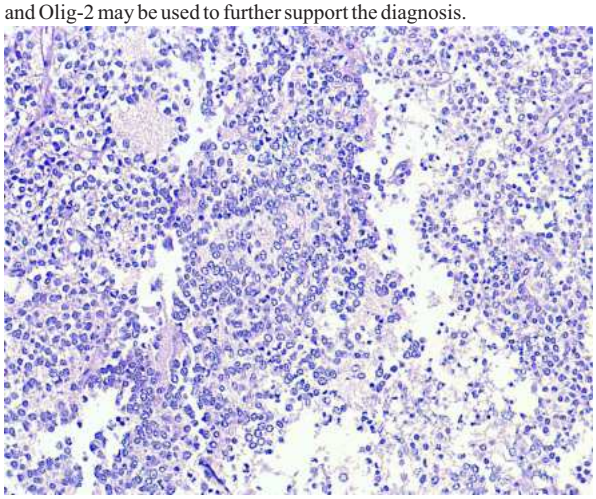

Fig 4: Pictomicrograph showing round uniform cells, clear perinuclear halo, delicate vascular network (H&E, **X40)**

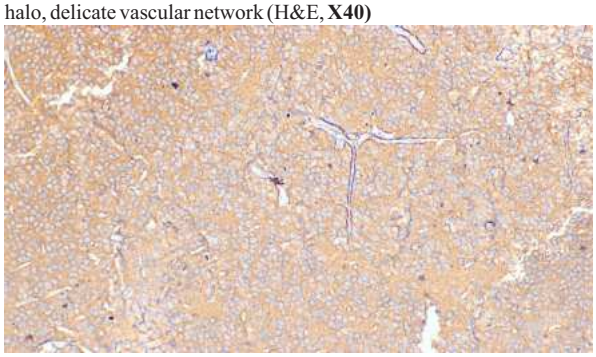

Fig 5: Pictomicrograph showing Synaptophysin IHC with diffuse cytoplasmic positivity in tumor cells (IHC, **X40)**

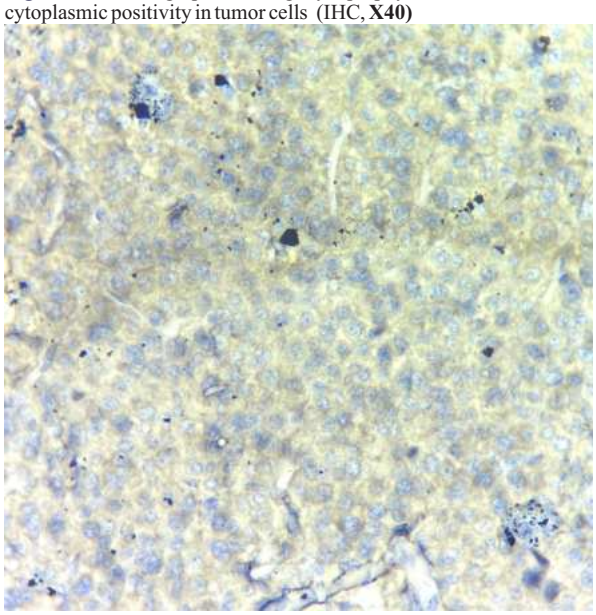

Fig 6: Pictomicrograph showing S 100 IHC with no cytoplasmic staining in tumor cells (IHC, **X40**)

DISCUSSION:

Central neurocytoma is a rare, typically benign intraventricular neuronal tumor that predominantly affects young adults and is most commonly located near the foramen of Monro. Histologically, it is characterized by uniform round cells with perinuclear halos that resemble oligodendroglioma, set within a fibrillary, neuropil-rich background. In our case, microscopic examination revealed small round cells with round to oval nuclei, fine granular ("salt and pepper") chromatin, inconspicuous nucleoli, and clear perinuclear halos—creating honeycomb-like appearance. The tumor cells exhibited scant cytoplasm, contributing to the characteristic "empty halo" appearance. Notably, neurocytic rosettes and neuropil islands, both hallmark features of central neurocytoma, were readily appreciated.

Based on radiological features, a diagnosis of an intraventricular neoplasm was initially considered. The differential diagnoses included central neurocytoma, oligodendroglioma, ependymoma, glioma, glioblastoma (GBM), and pineocytoma. However, these alternatives were subsequently excluded based on distinct gross, histopathological and immunohistochemical characteristics.

Oligodendroglioma are poorly circumscribed with an infiltrative border ,Typically not located in the ventricle, Lacks salt-and-pepper nuclei, neuropil islands, and ganglion cell differentiation, Synaptophysin negative where as the Ependymomas have cells have long fibrillary processes, Characteristically shows true rosettes, Typically protrudes from ventricular lining, GFAP positive and synaptophysin negative

Immunohistochemistry (IHC) played a pivotal role in confirming the neuronal lineage of the tumor cells. Synaptophysin, a presynaptic vesicle protein, is a sensitive and specific marker for neuronal differentiation and is widely employed in the diagnosis of central neurocytoma[3,4]. NeuN, another neuronal nuclear marker, further substantiated the neuronal origin. Most central neurocytomas have a benign course and lack features of anaplasia. However, approximately 20% of cases demonstrate atypical or high-grade features, including increased mitotic activity, cellular pleomorphism, vascular proliferation, necrosis, and a high MIB-1 (Ki-67) labeling index, all of which indicate a more aggressive clinical behavior[5]. In our case, mitotic figures and necrosis were absent, and the MIB-1 index was less than 2%, suggesting a low-grade tumor with a favorable prognosis[6]. Values exceeding 2–4% have been associated with an increased risk of recurrence and may necessitate adjuvant therapies, such as radiotherapy[7,8]. Therefore, regular long-term follow-up is essential, even in cases without atypical features, as late recurrence can occur.

Given the rarity of central neurocytoma and its overlapping features with other intraventricular neoplasms, accurate diagnosis depends on a combination of detailed histopathological examination and confirmatory immunohistochemical profiling. This integrated approach is vital for guiding appropriate management and prognostication.

CONCLUSIONS

Central neurocytoma is a rare, typically benign intraventricular tumor affecting young adults, often presenting with symptoms of obstructive hydrocephalus such as headache and visual disturbances. Surgical resection is the primary treatment, relieving symptoms and allowing for diagnostic confirmation. Histopathological evaluation reveals uniform round cells with neuronal features. Immunohistochemistry, including synaptophysin positivity, is crucial for differentiation from other intraventricular tumors[9]. The MIB-1 labeling index serves as an important prognostic tool[10]; values exceeding 2–4% suggest a higher risk of recurrence and may warrant adjuvant therapy. Accurate diagnosis and monitoring are essential for optimal long-term management and improved clinical outcomes.

REFERENCES:

1. Hassoun J, Gambarelli D, Grisoli F, Pellet W, Salamon G, Pellissier JF, et al. Central neurocytoma: An electron-microscopic study of two cases. Acta Neuropathol. 1982;56(2):151–6.
2. Louis DN, Perry A, Wesseling P, Brat DJ, Cree IA, Figarella-Branger D, et al. The 2021 WHO Classification of Tumors of the Central Nervous System: a summary. Neuro Oncol. 2021;23(8):1231–51.
3. Sharma MC, Deb P, Sharma S, Sarkar C. Neurocytoma: a comprehensive review. Neurosurg Rev. 2006;29(4):270–85.
4. Nishio S, Morioka T, Suzuki S, Fukui M. Immunohistochemical and ultrastructural study of central neurocytoma. Acta Neuropathol. 1993;85(6):586–93.
5. Soylemezoglu F, Scheithauer BW, Esteve J, Kleihues P. Atypical central neurocytoma. J Neuropathol Exp Neurol. 1997;56(5):551–6.
6. Rades D, Schild SE. Treatment recommendations for the various subtypes of neurocytoma. Expert Rev Neurother. 2006;6(10):1371–9.
7. Wang M, Zhang R, Han Z, Liu W, Xing B, Ma Z, et al. Clinical features and treatment outcomes of central neurocytoma: 15-year experience at a single institution. J Clin Neurosci. 2017;35:93–8.
8. Furtado SV, Ghosal N, Hegde AS. Central neurocytoma: a clinicopathological and radiological review. Neurosurg Rev. 2010;33(3):291–302.

9. Leenstra JL, Rodriguez FJ, Frechette CM, Giannini C, Pollock BE, Link MJ, et al. Central neurocytoma: diagnostic and therapeutic considerations in modern practice. Neurosurgery. 2007;60(4):701–8.
10. Rossi ML, Jones DM, Beale DJ, Hughes JT, Schiffer D. Clinicopathological study of 10 neurocytomas with recurrence and malignant progression. Br J Neurosurg. 1997;11(3):232–40.