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Clinical Presentation And Management Of Central Neurocytoma In Pediatric Age Group

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ABSTRACT

Central neurocytoma is a rare, typically benign intraventricular tumor primarily affecting young adults, but its occurrence in the pediatric population is exceptionally uncommon. This case report aims to highlight the clinical presentation, radiologic features, surgical management, and the outcome of a pediatric central neurocytoma. Herewith, we report the case of a toddler who presented with signs of raised intracranial pressure. Magnetic resource imaging (MRI) brain revealed heterogenous soft tissue showing enhancement in right lateral ventricle lesion extending into third ventricle and aqueduct of Sylvius leading to resultant obstructive hydrocephalus. Patient underwent endoscopic septostomy with maximal safe resection and was diagnosed with Central Neurocytoma (WHO Grade 2) based on immunohistochemistry (IHC) analysis of intraoperative tissue sample. Despite the rarity of central neurocytomas in children, early diagnosis and appropriate management can lead to excellent long-term prognosis, emphasizing the importance of awareness among clinicians of this entity.

Keywords: *Central Neurocytoma, Pediatric Brain Tumor, Intraventricular Tumor, Neuro-oncology, Rare Pediatric Tumor*

1. INTRODUCTION

Neurocytomas are WHO grade 2 primary CNS neoplasms that are derived from neural cells. These are of two types, intraventricular and extra-ventricular neurocytomas. Intraventricular neurocytoma is much more common and is called a central neurocytoma(CN) when located centrally within the ventricles, often near foramen of Monro.. It comprises less than 1% of primary brain tumors and predominantly affects young adults aged 20-40 years old. It is extremely rare in children with limited data available globally in pediatric age group. According to WHO classification, CN is a grade 2 brain tumor [1]. The tumor is speculated to arise from neuronal cells of septum pellucidum and subependymal cells of lateral ventricles. Due to its location in ventricles, CN often causes symptoms related to raised intracranial pressure, such as headache,

vomiting, altered mental status, visual and gait disturbances.Ophthalmic examination findings may reveal decreased visual acuity, diplopia and papilledema.MRI is preferred imaging modality but confirmatory diagnosis is based on histologic and immunophenotypic features.The mainstay of treatment is surgical removal of the tumor. Complete resection is associated with a good prognosis, though subtotal resection may require adjuvant radiotherapy.

Herein we report a case of a toddler, diagnosed with central neurocytoma who showed excellent recovery despite post operative complications.

2. CASE PRESENTATION

A toddler , born of non-consanguineous marriage with normal birth and developmental history was brought by her mother to our emergency department with complaints of fever and projectile vomiting since 3 days . Mother also complained of increased drowsiness and lethargy since 1 day. There was no significant family and past history. General examination revealed persistent hypertensive readings for her age (95-99th centile) , bradycardia, irregular breathing pattern. Neurological examination revealed altered level of consciousness and brisk deep tendon reflexes. Bilateral pupils were equal and reactive to light. Other systemic examination was within normal limits . Ophthalmic examination revealed blurring of optic disc margins of bilateral eyes with flame shaped hemorrhages along supranasal arcade and a blot hemorrhage in inferior quadrant of right eye. A final impression of bilateral disc oedema with hypertensive changes was made. Our examination and ophthalmic findings were consistent with signs of increased intracranial pressure.

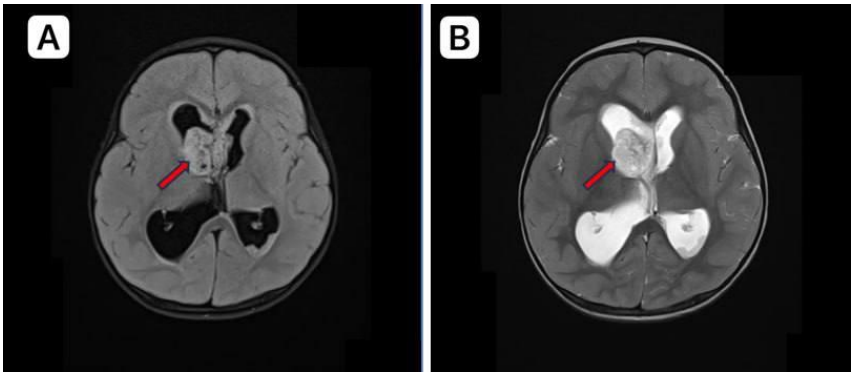

Figure 1: T1-weighted MRI. T1WI axial (A) and T2W1 axial (B) showing soft tissue lesion in right lateral ventricle ,extending into third ventricle with obliteration of foramen of Monroe and aqueduct of Sylvius leading to obstructive hydrocephalus leading to dilatation of third ventricle and asymmetrical dilatation of bilateral lateral ventricles.

MRI: Magnetic resonance imaging; T1WI: T1-weighted image, T2WI: T2-weighted image MRI revealed large altered signal intensity soft tissue lesion measuring 33 x 20 x 41 mm in its largest dimensions with mixed signal intensities exhibiting intermediate to high T2/ Fluid-attenuated inversion recovery (FLAIR) signal with patchy areas of mild restricted diffusion within and mild heterogenous post-contrast enhancement in right lateral ventricle, extending into third ventricle with obliteration of foramen of Monroe and aqueduct of Sylvius leading to obstructive hydrocephalus (Figure 1). As a result, there was dilatation of third ventricle and asymmetrical dilatation of bilateral lateral ventricles (right more than left) thereby causing left sided displacement of septum pellucidum.

A neurosurgery opinion was taken and patient underwent endoscopic septostomy with maximal safe resection which was done under navigation guidance following which an omega reservoir was placed in right ventricle. Intraoperative findings confirmed raised intraventricular pressure with space occupying lesion in right lateral ventricle obscuring right foramen of Monroe and extension into third ventricle. Excised specimen of intraventricular tumor was sent for frozen section and histopathological studies. Microscopic examination (Figure5)of biopsy sample revealed nervous tissue with tumor composed of clusters, nests and true rosettes of round cells which had round nuclei, stippled chromatin and scanty cytoplasm. Surrounding gliotic nervous tissue was present containing few lymphocytes. Immunohistochemistry (IHC) was strongly positive for Synaptophysin and Ki-67 proliferation index of 2-4%. The background gliotic nervous tissue show GFAP positivity. A confirmatory diagnosis of Central Neurocytoma (WHO Grade 2) as suggested by IHC studies.

Post operatively patient showed significant clinical improvement but one week later, the patient started developing signs of raised intracranial pressure again and a repeat brain CT scan revealed development of Subdural hygroma in right fronto-temporo-parietal convexity of maximum 16 mm thickness, causing mass effect in the form of midline shift. This warranted placement of external ventricular drain (Figure 2). One week later patient was reoperated again for unresolving hydrocephalus and a ventriculoperitoneal (VP) shunt was placed (Figure 3) to relieve intracranial pressure .

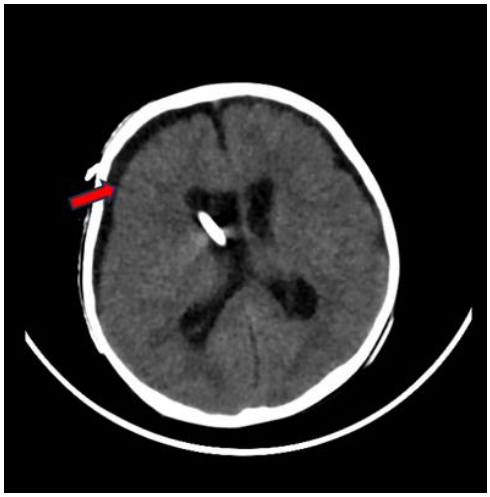

Figure 2: Non-contrast CT brain: subdural hygroma with extraventricular drain in situ traversing through right frontal region with it's tip in the body of the right lateral ventricle.

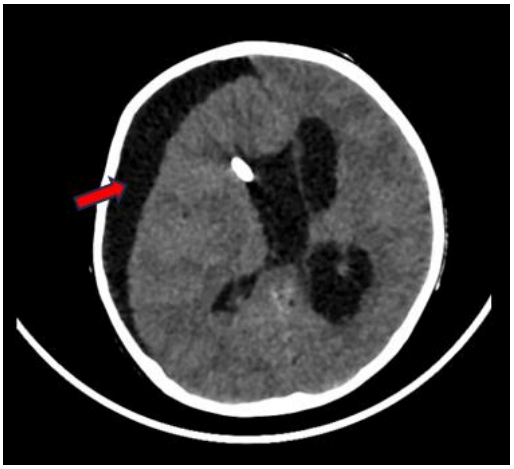

Figure 3: Non-contrast CT brain: subdural hygroma and pneumocephalus along right cerebral convexity with ventriculoperitoneal shunt in situ traversing through right frontal region with it's tip in right frontoparietal region.

2 weeks later after placement of VP shunt , a repeat CT Scan of brain revealed reduction in size of hydrocephalus , subdural hygroma and pneumocephalus(Figure 4). Patient was discharged with no neurological deficit and is being followed up every 6 months to look for recurrence of tumour.

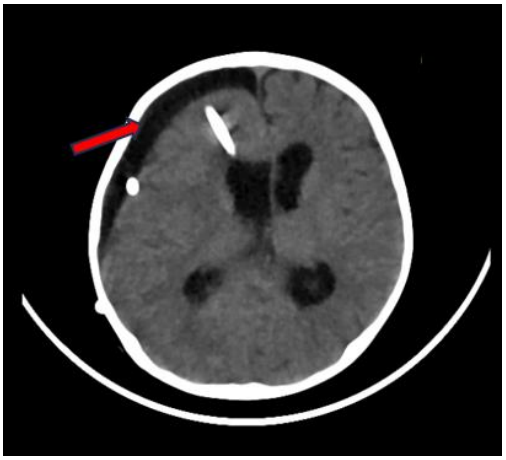

Figure 4: Non-contrast CT brain: follow up scan showing Subdural hygroma and pneumocephalus reduced in size as seen along the right cerebral convexity.

3. DIFFERENTIAL DIAGNOSIS

In a child with intraventricular brain tumour, following differentiatials should be taken into consideration: ependymoma, mixoid glioneuronal tumour, subependymal giant cell astrocytoma, and to a lesser extent, oligodendroglioma, and choroid plexus papilloma.

Table 1: Differential diagnosis of Central Neurocytoma based on histopathology, Immunohistochemistry markers and other typical features [2].

Feature	Central Neurocytoma (CN)	Ependymoma	Myxoid Glioneuronal Tumor (MGT)	Subependymal Giant Cell Astrocytoma (SEGA)	Oligodendroglioma	Choroid Plexus Papilloma (CPP)
HPE: Morphology	Uniform round cells, salt-and-pepper chromatin, delicate vascular network, occasional rosettes or pseudorosett	Perivascular pseudorosett, true ependymal rosettes, monomorphic cells	Myxoid stroma with embedded oligodendrocyte-like and neuronal cells; floating neurons	Large, pleomorphic astrocytes with abundant cytoplasm, multinucleated cells, and prominent nucleoli	Uniform round nuclei, perinuclear halos ("fried egg" appearance), chicken-wire capillaries	Papillary architecture with fibrovascular cores lined by cuboidal to columnar epithelial cells
Synaptophysin	Positive	Negative or patchy	Positive (neuronal component)	Negative	Negative (usually)	Negative
NeuN	Positive (neuronal marker)	Negative	Positive (neuronal component)	Negative	Negative	Negative
GFAP	Negative or weak (variable, depending on glial component)	Positive (glial processes near rosettes)	Positive (glial component)	Positive	Positive	Negative
EMA	Negative	Positive (dot-like pattern)	Negative	Positive (dot-like or diffuse)	Negative	Positive (apical membrane staining)
S100	Positive (variable)	Positive (diffuse)	Positive (diffuse)	Positive	Positive	Positive
IDH1 R132H	Negative	Negative	Negative	Negative	Positive	Negative
INI1/SMARCB1	Retained	Retained	Retained	Retained	Retained	Retained
Ki-67 (Proliferation Index)	Low (<2-5%)	Variable (can be higher in anaplastic variants)	Low	Low	Low to moderate	Low
Other Features	Typically found in lateral ventricles, may show	Occurs along ependymal lining, including posterior	Typically cortical location	Typically in the lateral ventricles, associated with tuberous	Diffuse cerebral hemispheres, especially frontal lobe	Lateral ventricles, third or fourth ventricle,

	calcification	fossa, may calcify		sclerosis		papillary tumor

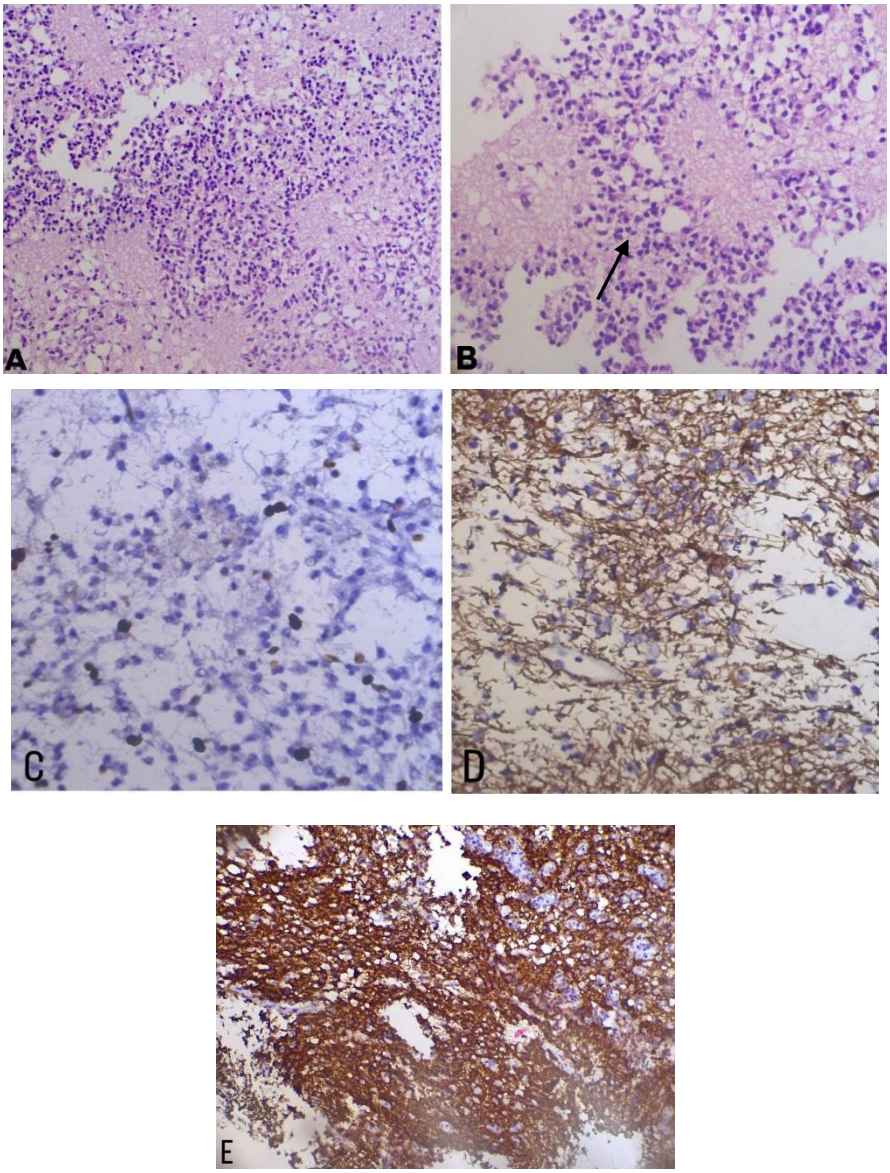

Figure 5: A) H&E stained slide showing small round nuclei with stippled chromatin and scanty cytoplasm (20x)

B) Nervous tissue composed of clusters, nests and true rosettes (black arrow) of round cells (40x)

C) Tumours cells expressing Ki-67 with proliferation index of 2-4%

D) The background gliotic nervous tissue showing GFAP positivity

E)Tumour cells strongly expressing Synaptophysin

The final diagnosis of these differentials relies significantly on histopathological and IHC studies which help determine tumour's origin and classify it accurately as per WHO tumour grading system.

In our case Immunohistochemistry (IHC) was strongly positive for Synaptophysin and Ki-67 proliferation index of 2-4%. The background gliotic nervous tissue showed GFAP positivity, confirming our diagnosis of central neurocytoma (WHO grade 2)

4. OUTCOME AND FOLLOW-UP

After 3 weeks of stay, our patient showed excellent recovery and was discharged with VP shunt in situ. The patient was able to return to daily activities with no neurological symptoms or neurological deficits. The patient is being followed up every 6 months for first few years to look for any tumour recurrence or shunt related complications.

5. DISCUSSION

Central neurocytomas are indolent brain tumours that are infrequently seen in the paediatric age group, and are typically observed in young adults without a sex preponderance [3]. Comprehensive data on the total number of pediatric cases are limited and exact global figures are challenging to ascertain due to the rarity of pediatric CN. The table below summarizes available data:

Study/Source	Number of Pediatric Cases Reported	Age Range (Years)	Key Notes
Retrospective study (1993-2004)	59	Median age -16 years Only 7% children were less than 5 year old	Institutional analysis over 11 years; emphasized rarity in pediatric cases.[4]
Case report	1	9	Central neurocytoma of the third ventricle. [5]
Case report	1	12	Clinical presentation and management of atypical central neurocytoma [6]
Frontiers in Oncology (2022)	no exact number provided	Pediatric age (unspecified)	Emphasizes rarity and lack of comprehensive epidemiological data in children. [7]

As of 2025 , the total number of pediatric cases in the literature is likely fewer than 70. Given the scarcity of cases, each instance contributes valuable insights into the clinical presentation, management, and outcomes of pediatric central neurocytomas. Ongoing documentation and analysis are essential to enhance understanding and treatment strategies for this rare pediatric tumor.

Headache, visual disturbance, lethargy, irritability, vomiting, paraesthesia, difficulty maintaining posture, papilledema, and occasional intraventricular haemorrhage are associated findings which indicate raised intracranial pressure [8]. The World Health Organisation (WHO) has classified CN as a low-grade tumour (Grade II) [9] and is usually deeply seated in midline tissues, which may extend to the third ventricle. It is frequently found in the frontal horn of the lateral ventricle next to the foramen of Monro and is connected to the septum pellucidum [10]. Some cases of CN have been documented extraventricularly in the brain parenchyma , spinal cord, fourth and third ventricle. There have been reports of malignant variations, despite the tumour being primarily benign [11]. In the lateral ventricle, CN manifests as a clearly defined lobulated mass on neuroimaging. In 25% to 50% of instances, it manifests as a mixed density and heterogeneous mass on a computed tomography (CT) scan brain, along with cystic degeneration and calcification [12]. Haemorrhagic alterations are rarely inside the tumour .

Microscopically , the tumour consists of uniform, small round cells with abundant cytoplasm and round nuclei.Immunohistochemistry is investigation of choice for confirmatory diagnosis of CN in order to verify the tumour's neuronal origin. In both fibrillary and perivascular regions, antibodies to synaptophysin are specific for CN. Additionally, glial fibrillary acidic protein reactivity and neurone-specific enolase positivity in IHC help with diagnosis [13].

Total surgical resection is the standard treatment for CN, and it can be carried out using endoscopic , transcallosal or transcortical neurosurgical approaches[14]. Compared to subtotal resection, which was linked to an 86% 5-year survival rate, total resection accomplished in 30–50% of cases was linked with 99% 5-year survival rate [15]. Nonetheless, vigorous and excessive tumour removal may pose a substantial danger of neurological degeneration; therefore, the pros and cons of the procedure should be weighed.

The transcallosal approach is primarily used to operate on a tumour in the third ventricle or both lateral ventricles with normal ventricular size and to avoid making a cortical incision. The transcortical approach is used to operate on large tumours, to prevent damage to the parasagittal vein and fornix, and to avoid making an incision in the corpus callosum. It is important to weigh the benefits and drawbacks of each strategy when making a decision. The most common consequences linked to

the transcallosal procedure are hemiparesis, memory loss, aphasia, mutism, and disconnection syndromes. By contrast, transcortical approach problems include hemiparesis, aphasia, mutism, seizures, disorientation, and memory loss [65].

According to Mahavadi et al., the recurrence rate following gross total resection was 23.9%, while the recurrence rate following gross total resection with radiotherapy (GTR+RT) was 6.9%. While overall survival does not appear to be altered, GTR+RT may increase morbidity [17]. In comparison to GTR and GTR+RT (95.5% and 95.3%, respectively), adjuvant radiotherapies reduce the risk of tumour development following incomplete tumour resection and increase the overall survival rate of patients with maximum safe resection, which is 93.2%. [18]. Accordingly, the major objectives of the procedure are to restore the CSF flow, give a sample for a conclusive histological diagnosis, and achieve maximum tumour removal with minimal neurological consequences.

6. LEARNING POINTS

- Central neurocytoma is a rare slow growing brain tumour which is benign and usually affects young adults but can also develop in early childhood like our case.
- surgical resection is primary treatment and confirmatory diagnosis is based on histopathological and IHC studies.
- Early intervention provides favourable prognosis if tumour is completely resected which can prevent complications impacting early childhood development.

REFERENCES

[1] David N. Louis, Hiroko Ohgaki, Otmar D. Wiestler et al. WHO Classification of Tumours of the Central Nervous System. (2016) ISBN: 9789283244929 - Google Books

[2] Louis, D. N., et al. (2007). "The 2007 WHO classification of tumours of the central nervous system." *Acta Neuropathologica*, 114(2), 97–109

[3] Hallock A, Hamilton B, Ang LC, Tay KY, Meygesi JF, Fisher BJ, et al. Neurocytomas: long-term experience of a single institution. J Neurooncol 2011;13(9):943–9.

[4] Rades D, Schild SE, Fehlauer F. Defining the best available treatment for neurocytomas in children. Cancer. 2004;101:2629–32. doi: 10.1002/cncr.20695. [DOI] [PubMed] [Google Scholar]

[5] Baishya BK, Singh RK, Dutta D, Hussain Z. Pediatric central neurocytoma: Case report and review of literature. J Pediatr Neurosci. 2016 Oct-Dec;11(4):348-350. doi: 10.4103/1817-1745.199485. PMID: 28217163; PMCID: PMC5314854

[6] Sun S, Malicki DM, Levy ML, et alAtypical central neurocytoma with aggressive features in a childBMJ Case Reports CP 2020;13:e236262

[7] Front. Neurol., 22 December 2021 Sec. Neuro-Oncology and Neurosurgical Oncology.Volume 12 - 2021 | https://doi.org/10.3389/fneur.2021.790641

[8] Patel DM, Schmidt RF, Liu JK. Update on the diagnosis, pathogenesis, and treatment strategies for central neurocytoma. J Clin Neurosci 2013;20 (9):1193–9.

[9] Rai P, Nayak R, Anand D, Menon G: Central neurocytoma in the posterior fossa. BMJ Case Rep. 2019, 12:10.1136/bcr-2019-231626

[10] Y. Song, X. Kang, G. Cao et al., "Clinical characteristics and prognostic factors of brain central neurocytoma," Oncotarget, vol. 7, no. 46, pp. 76291–76297, 2016.

[11] D. M. Patel, R. F. Schmidt, and J. K. Liu, "Update on the diag nosis, pathogenesis, and treatment strategies for central neuro cytoma," Journal of Clinical Neuroscience, vol. 20, no. 9, pp. 1193–1199, 2013

[12] A. M.Richardson, V. L. Armstrong, J. E. Gernsback, S. H. Gul tekin, and R. J. Komotar, "Central neurocytoma: rare presenta tion in fourth ventricle and review of literature," World Neurosurgery, vol. 123, pp. 357–361, 2019.

[13] S. J. Lee, T. T. Bui, C. H. Chen et al., "Central neurocytoma: a review of clinical management and histopathologic features," Brain Tumor Research and Treatment, vol. 4, no. 2, pp. 49 57, 2016.

[14] Li X, Guo L, Sheng S, et al. Diagnostic value of six MRI features for central neurocytoma. Eur Radiol 2018; 28(10): 4306–4313.

[15] Collins VP, Jones DT and Giannini C. Pilocytic astrocy toma: pathology, molecular mechanisms and markers. Acta Neuropathol 2015; 129(6): 775–788.

[16] M. Wang, P. Zhou, S. Zhang et al., "Clinical features, treat ment, and long-term outcomes of central neurocytoma: a 20 year experience at a single center," World Neurosurgery, vol. 109, pp. e59–e66, 2018.

[17] M. H. Schmidt, O. N. Gottfried, C. S. von Koch, S. M. Chang, and M. W. McDermott, "Central neurocytoma: a review," Journal of Neuro-Oncology, vol. 66, no. 3, pp. 377–384, 2004.

[18] A. K. Mahavadi, P. M. Patel, M. Kuchakulla et al., "Central neurocytoma treatment modalities: a systematic review asses sing the outcomes of combined maximal safe resection and radiotherapy with gross total resection," World Neurosurgery, vol. 137, pp. e176–e182, 2020.