ORIGINAL RESEARCH PAPER

Radiotherapy

ATYPICAL PAEDIATRIC EXTRA VENTRICULAR NEUROCYTOMA– A RARE CASE REPORT

KEY WORDS: Paediatric tumors, extra ventricular neurocytoma, radiotherapy

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ABSTRACT

Central neurocytoma (CN) and extra ventricular neurocytoma (EVN) are rare intracranial tumors. Central neurocytomas are well-differentiated, intraventricular lesions mainly seen in adults. Similar tumors may develop in the brain parenchyma or spinal cord referred to as extra ventricular neurocytomas. Here we are reporting a case of 10-year-old boy with intracranial mass lesion for which left parieto-occipital craniotomy was done with histopathology showing atypical extra ventricular neurocytoma, recurred after for which gross total excision was done and histopathology report confirmed neurocytoma recurrence. Postoperative MRI showed residual disease and treated with radiation therapy 50 Gray in 28 fractions using IMRT technique. Post radiation MRI showed no residual disease and was kept under follow up.

INTRODUCTION:

Central neurocytoma (CN) and extra ventricular neurocytoma (EVN) are rare intracranial tumors First identified as histologically distinct entities in 1982[1]. Central neurocytomas are well-differentiated, constituting approximately one-half of all intraventricular lesions in adults[2] . Similar tumors have occasionally been identified in the brain parenchyma or spinal cord; in which case they are referred to as extra ventricular neurocytomas. They are classified under glioneuronal and neuronal tumors according to 2021 WHO classification of tumors of the central nervous system [3]. Microscopically sheets, clusters, ribbons or rosettes of monotonous tumor cells with round and regular vesicular nuclei and distinct nucleoli [4].

Immunohistochemically, extra ventricular neurocytomas are characterized by the robust expression of synaptophysin, but with a lack of oligodendrocyte transcription factor 2, isocitrate dehydrogenase enzyme isoform 1 (IDH1) R132/IDH2 R172 mutations and p53 immunoexpression[5]. Atypical lesions are characterized by a MIB-1 labelling index >3% and atypical histologic features such as necrosis, increased mitotic activity, and vascular proliferation [6]. Intracerebral haemorrhage and transformation to a more aggressive malignant lesion are considered serious complications [7]. The treatment for extra ventricular neurocytomas in paediatric and adult populations is gross total resection, with post- operative radiation reserved for subtotal resection or recurrent disease [8]. There is a lack of data on chemotherapy for both typical and atypical extra ventricular neurocytoma.

Case Report:

10-year-old boy with normal developmental milestones presented with persistent headache and projectile vomiting in 10/2021. MRI brain showed left parieto-occipital cystic lesion. Left parieto-occipital craniotomy done under general anaesthesia and histopathology came as atypical extra ventricular neurocytoma (figure 1,2). Immunohistochemistry shows Ki 67- 45%(figure 3). Synaptophysin(figure 4:), Glial fibrillary acidic protein(GFAP) negative (figure 5) and neuron specific enolase (NSE) positive (figure no:6).

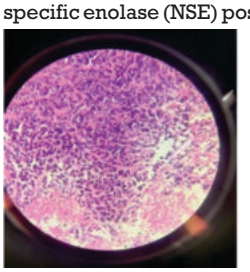

Figure.1

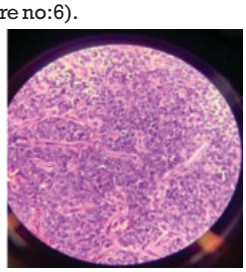

Figure.2

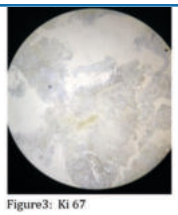

Figure3: Ki 67

Figure 4: Synaptophysin Positive

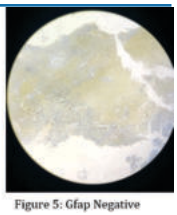

Figure 5: Gfap Negative

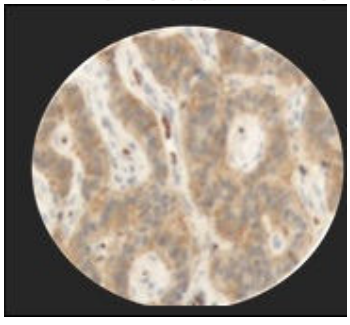

Figure 6 : NSE positive

Post-operative MRI showed no residual disease and he was kept under follow up. Follow up MRI after 6 months showed peripherally enhancing lesion with heterogeneously enhancing solid component measuring 46 x 48 x51 mm involving left parieto- occipital lobe-suggesting recurrence (figure no:7). Gross total excision was done on 13-8-22 with histopathology report atypical extra-ventricular neurocytoma- recurrence. Post-surgery MRI detected 1.9x1.4x0.5 cm lesion with diffusion restriction – most likely residual lesion. Post operatively he was treated with radiotherapy 50Gy in 28 fractions using IMRT technique. (figure no:8 and 9) MRI taken after 6 weeks of Radiation showed no residual mass lesion. Patient was clinically better with no neurological deficits, and kept under follow up.

Clinical Diagnosis : Extra ventricular neurocytoma

Differential Diagnosis:

1. Ependymoma,
2. Oligodendroglioma,
3. High grade astrocytoma.

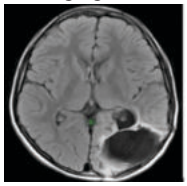

Figure 7

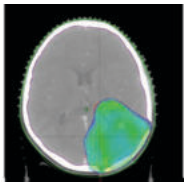

Figure 8

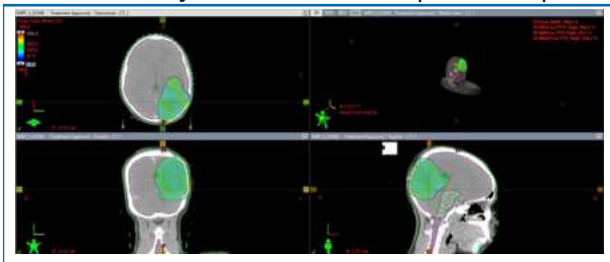

Figure 9

RESULTS:

Patient was clinically better post radiation with no neurological deficits, and MRI suggestive of no significant recurrent or residual disease and is under follow up for past 1 year 10 months.

DISCUSSION ON MANAGEMENT

Extra ventricular neurocytoma is classified under glioneuronal and neuronal tumors by WHO. It is due to alteration in FGFR (FGFR1-TACC1 fusion), and is of IDH-wild type [3]. Paediatric extra ventricular neurocytoma accounts for approximately 0.1% of central nervous system tumors[8]. Tumors are classified into typical or atypical categories, and atypical histological criteria. Extra ventricular neurocytoma generally have a good clinical outcome if completely resected. However, extra ventricular neurocytoma with atypical features are associated with an aggressive clinical outcome and higher recurrence rates as compared with extra ventricular neurocytoma without atypical features. In a study comprising 35 patients with extra ventricular neurocytoma it was found that when MIB-1 ranged from 3% to 18% in the context of cellular atypia, the recurrence rate was the highest[9]. The 5-year overall survival rates were 91% after complete tumor resection, 93% after complete tumor resection and radiotherapy, 46% after incomplete tumor resection and 69% after incomplete tumor resection followed by radiotherapy[9]. The higher rates of recurrence and mortality is detected in neurocytoma with atypical features. 40% recurrence and 20% mortality in atypical central neurocytoma compared with 68% recurrence and 44% mortality, respectively, in atypical extra ventricular neurocytomas [10].

The treatment of atypical neurocytoma has not been distinguished from treatment of typical extra ventricular neurocytoma. Complete resection would provide better local control and survival rates than incomplete resection and that local control and survival rates are improved with postoperative radiation therapy in cases of incomplete resection. In adults, 50–54 Gy is sufficient for typical lesions, whereas atypical lesions require 56–60 Gy. After incomplete tumor resection in children, post-operative radiation significantly improves local control, but not overall survival. Doses of 50 Gy appear appropriate[11]. The evidence for use of chemotherapy is low due to rarity of the condition. Brandes et al. reported a favourable outcome of chemotherapy in progressive and recurrent central neurocytoma; they used a platinum-based agent in three patients. However, there is a lack of data on chemotherapy for both typical and atypical extra ventricular neurocytoma [11].

In this case it was an atypical neurocytoma with synaptophysin positive and Ki 67- 45%. The child was initially treated with gross total excision, which is considered the standard of care, but recurrence occurred within 6 months. Repeat excision was done, but residual disease persisted. In view of age and recurrence in spite of resection multidisciplinary tumor board decided for radiation therapy. He was treated with 50 Gy in 28 fractions using IMRT technique. He had a good disease control and was kept under follow up. Clinical experience of neurocytoma treatment with different modalities like chemotherapy and targeted therapy has not been explored to the fullest due to the rarity of this type of tumor.

Final Diagnosis

Extra ventricular neurocytoma

REFERENCES

1. Chen CL, Shen CC, Wang J, Lu CH, Lee HT. Central neurocytoma: a clinical, radiological and pathological study of nine cases. Clinical neurology and neurosurgery. 2008 Feb 1;110(2):129-36.
2. Hassoun J, Söylemezoglu F, Gambarelli D, Figarella Branger D, von Ammon K, Kleihues P. Central neurocytoma: a synopsis of clinical and histological features. Brain Pathology. 1993 Jul;3(3):297-306.
3. Bale TA, Rosenblum MK. The 2021 WHO Classification of Tumors of the Central Nervous System: An update on pediatric low-grade gliomas and glioneuronal tumors. Brain Pathol. 2022 Jul;32(4):e13060. doi: 10.1111/bpa.13060. Epub 2022 Feb 25. PMID: 35218102; PMCID: PMC9245930.
4. Brat DJ, Scheithauer BW, Eberhart CG, Burger PC. Extraventricular neurocytomas: pathologic features and clinical outcome. Am J Surg Pathol. 2001 Oct;25 (10):1252-60. doi: 10.1097/00000478-200110000-00005. PMID: 11688459.
5. Ishiuchi S, Tamura M. Central neurocytoma: an immunohistochemical, ultrastructural and cell culture study. Acta Neuropathol. 1997 Nov;94(5):425-35. doi: 10.1007/s004010050729. PMID: 9386774.
6. Rades D, Schild SE, Fehlauer F. Prognostic value of the MIB-1 labeling index for central neurocytomas. Neurology. 2004 Mar 23;62(6):987-9. doi: 10.1212/01.wnl.0000115392.21898.e3. PMID: 15037708
7. Jamshidi J, Izumoto S, Yoshimine T, Maruno M: Central neurocytoma presenting with intratumoral hemorrhage. Neurosurg Rev 24: 48–52, 2001
8. Han L, Niu H, Wang J, Wan F, Shu K, Ke C, Lei T. Extraventricular neurocytoma in pediatric populations: A case report and review of the literature. Oncol Lett. 2013 Nov;6(5):1397-1405. doi: 10.3892/ol.2013.1583. Epub 2013 Sep 12. PMID: 24179531; PMCID: PMC3813794.
9. Rades, D., Schild, S.E. Treatment recommendations for the various subgroups of neurocytomas. J Neurooncol **77**, 305–309 (2006)
10. Kane AJ, Sughrue ME, Rutkowski MJ, Aranda D, Mills SA, Lehil M, Fang S, Parsa AT. Atypia predicting prognosis for intracranial extraventricular neurocytomas. J Neurosurg. 2012 Feb;116(2):349-54. doi: 10.3171/2011.9.JNS10783. Epub 2011 Nov 4. PMID: 22054208.
11. Brandes AA, Amistà P, Gardiman M, Volpin L, Danieli D, Guglielmi B, Carollo C, Pinna G, Turazzi S, Monfardini S. Chemotherapy in patients with recurrent and progressive central neurocytoma. Cancer. 2000 Jan 1;88(1):169-74. doi: 10.1002/(sici)1097- 0142(20000101)88:1<169::aid-cncr23>3.0.co;2-7. PMID: 10618620.