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SUPRATENTORIAL EPENDYMOMA:EXPERIENCE OF A NEUROSURGEON

Neurosurgery

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ABSTRACT

Background-Ependymoma account for 5 to 10 % of all brain tumors in young population however majority occur in infratentorial location and Supratentorial Ependymoma (STE) are extremely rare . Aim of this original article is to describe demographic , clinical features and surgical outcome of these rare tumors operated by Neurosurgeon DSM .

Materials and Method- 10 cases of STE were operated by a single Neurosurgeon DSM from May 2017 to January 2019 at a tertiary care hospital.

Results- Gross total resection was performed in 8 cases and Near total resection was performed in 2 cases . Recurrence was seen in 3 cases during study period and 2 cases required resurgery

Histopathology was anaplastic Ependymoma WHO grade 3 in all the cases.

Conclusion- STE have a better prognosis as compared to infratentorial Ependymoma in view of good chance of gross total excision .

KEYWORDS

Anaplastic Ependymoma, Supratentorial Ependymoma (STE), Extra ventricular, Ependymoma

INTRODUCTION

Ependymoma comprise 5 -10% all tumors in pediatric and adults less than 20 years of age[1–3]. Majority of Ependymoma occur in infratentorial location and supratentorial location is extremely rare. Majority of these tumors arise from ependymal cells of fourth ventricle or central canal of spinal cord .Supratentorial ependymoma (STE) can occur both at intraventricular and extraventricular location[4–6] .World health organization has classified STE into WHO grade 2 (Low grade) and grade 3(anaplastic ependymoma) . Nuclear atypia , necrosis, high cellularity, mitotic activity and cellular proliferation decides the grade of STE .The management of STE is controversial as there are only isolated small case series and case reports in world literature[7–9]. Author present the largest single Neurosurgeon experience of supratentorial anaplastic ependymoma operated at a tertiary care hospital. This original article shares clinical details , surgical strategy , post- operative management , clinical and radiological outcome after surgical excision .

MATERIAL AND METHODS

All cases of histologically proved STE operated by the author Dipanker Singh Mankotia (DSM) were included in the study. Demographic data, clinical details , histopathology, treatment and outcome were recorded p for all the patients included in the study .All patients had a preoperative and post-operative contrast CT or MRI scan. All patients had a clinical and radiological follow up at regular intervals after surgery .Histopathology was done according to WHO classification by an experienced Neuro pathologist. Written and informed consents were taken by each patient or patients relatives included in the study. Only those cases where the minimum clinical and radiological follow up was 6 months ,were included in the study.

RESULTS

10 cases of histology proved STE with diverse age groups were operated by author DSM at a single tertiary care hospital from May 2017- January 2019 . There was a definite male sex predilection in our study with M:F ratio of 4:1(Figure 1). The median age at presentation and surgery was 15 years (1-43 years)(Figure 2). The median duration of symptoms to presentation to hospital was 5 months (2-10 months) (Figure 3) .The tumor size ranged from 5 cm to 10 cm in maximum dimension .All patients had signs and symptoms of raised intracranial pressure in form of persistent headache and vomiting .Focal neurological deficits like hemiparesis were seen in 6 patients . All patients had at least one episode of seizure before presentation to hospital .Majority of the patients (50%) had both solid and cystic component on neuroimaging and there was homogenous enhancement of solid component in all the cases .Purely cortical solid tumor was seen in only 2 cases (20%) with homogenous contrast enhancement . The main aim of surgery was radical tumor excision and a gross total excision could be performed in 8 out of 10 cases. A Near Total Excision was performed in 2 pediatric cases in view of high vascularity of tumor and intraoperative excessive blood loss. There was no mortality or serious morbidity in the current series during hospital stay. The histopathology in all our cases was Anaplastic Ependymoma (WHO grade 3). Microvascular proliferation and foci of necrosis were seen in all the cases confirmed as WHO grade 3 Anaplastic Ependymoma .The classical histological feature of perivascular pseudo rosettes were also seen in all the cases. All adult patients received post operative radiotherapy and chemotherapy . Radiotherapy was not given in 2 cases as children were 1 year old .One patient of giant STE who had a recurrence, was lost to follow up after 1 year .There was radiological recurrence in 3 cases during follow up scans . Re survey for recurrence was performed in 2 cases and one case was lost to follow up .All other 7 cases did not have any signs of radiological or clinical recurrence at atleast 6 months follow up.

Figure 1.Sex Distribution

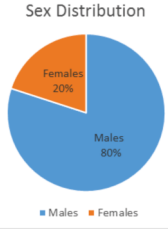

Figure 2.Age Distribution

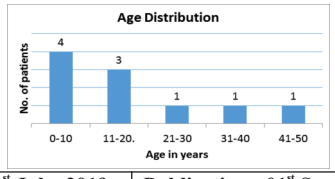

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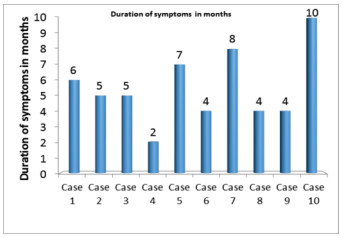
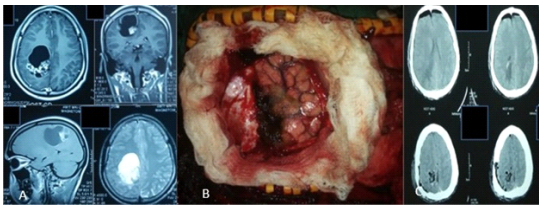
Figure 3.Duration of symptoms



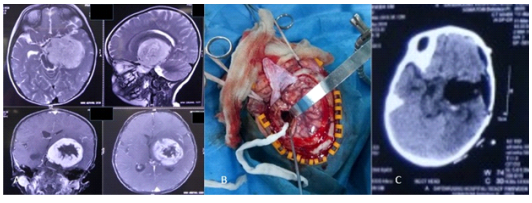


Illustrative case 1 Image A- preoperative MRI with a right posterior frontal STE. Image B- intra operative . Image C- post operative CT Scan

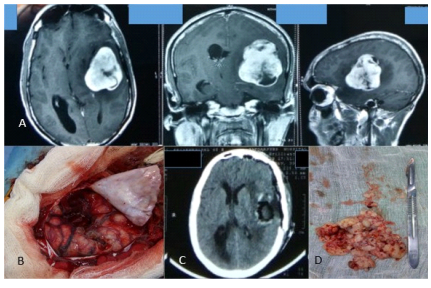
Table 1 below shows details of all cases. Illustrative images below Table 1 shows various images of five of our operated cases.

Sno	Age (years)	Sex	Location	Extent of Resection Gross Total Resection GTR Near Total Resection NTR	Biopsy	Recurrence
1	17	M	Posterior Frontal	GTR	Anaplastic Ependymoma	No
2	9	M	Parieto- temporal	GTR	Anaplastic Ependymoma	No
3	43	M	Parietal	GTR	Anaplastic Ependymoma	Yes
4	1	M	Temporo-parietal	GTR	Anaplastic Ependymoma	No
5	5	M	Giant Frntotemporoparietal	NTR	Anaplastic Ependymoma	Yes
6	14	F	Parietal	GTR	Anaplastic Ependymoma	No
7	19	M	Occipital	GTR	Anaplastic Ependymoma	No
8	35	M	Posterior frontal	GTR	Anaplastic Ependymoma	No
9	1	F	Occipital-parietal	NTR	Anaplastic Ependymoma	Yes
10	27	M	Occipital	GTR	Anaplastic Ependymoma	No

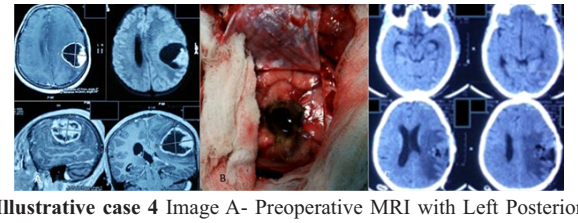
Table 1- Summary of operated case of Supratentorial Anaplastic Ependymoma.



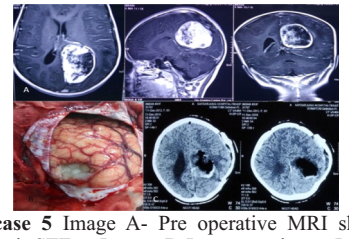
Illustrative case 2 Image A- pre operative MRI with Left temporo-parietal STE . Image B- Intra operative . Image C- post operative CT Scan



Illustrative case 3 Image A-preoperative MRI with Left frontoparietal STE . Image B- Intra operative , Image c- Post operative CT Scan , Image 4- Tumor specimen



Illustrative case 4 Image A- Preoperative MRI with Left Posterior frontal STE, Image B-Intraoperative , Image C- post-operative CT Scan



Illustrative case 5 Image A- Pre operative MRI showing a left occipito-parietal STE , Image B-Intraoperative , Image C- Post operative CT Scan

DISCUSSION

Ependymoma are tumors of central nervous system which arise from ependymal cells[10–12] . They account for roughly one third of all pediatric brain tumors and are still rarer in adults. Majority of these tumors arise from ependymal lining of fourth ventricle and central canal of spinal canal[13–16]. Supratentorial extra ventricular location is extremely rare. STE have been subclassified into intraventricular and purely parenchymal extraventricular form.STE is thought to arise from the ependymal remnants of brain parenchyma[17–20] .WHO has graded ependymoma into three subtypes as myxopapillary ependymoma (Grade 1),low grade ependymoma (grade 2) and Anaplastic ependymoma (grade 3).In contrast to previous published studies on STE histopathology in all our cases was WHO grade 3 Anaplastic Ependymoma[21–24]. Supratentorial and infratentorial ependymoma behave differently and prognosis of former is better than latter[20–22,24,25] .

Majority of supratentorial ependymoma occur in adults in second decade of life with a mean age of presentation being 18-24 years but in our series the median age was a bit younger at 15 years [26–29]. There was a definite male sex predisposition(M: F 4:1) in the current series as opposed to other series with equal distribution. All the patients in our study group presented with signs of raised intracranial pressure like chronic headache , vomiting and had atleast one episode of seizure .All patients received antiepileptic therapy before and after surgery .The incidence of grade 2 and grade 3 STE was equal in world literature however all cases encountered in our study were grade 3 Anaplastic STE[5,7,29,30] .

Pure intraparenchymal forms are extremely rare and exact pathogenesis and origin is not known till date. STE are hypothesized to arise from ependymal cell remnants in brain parenchyma. Glial cells with progenitor cell properties are also thought to be a source of STE . Proginator cells are considered to be probable source of extremely rare extra cerebral epedymoma[16,17,25,31] . The histopathological feature of STE are similar to ependymal tumors found elsewhere in body. These tumors usually arise from either frontal or temporal lobe and present with symptoms of raised intracranial pressure, seizures and headache[18,29,30,32]. Great deal of clinical suspicion is required to diagnose STE before surgery based solely on radiological features which are inconsistent and not classical. The differential diagnosis include pleomorphic xanthochromic astrocytoma, oligodendroglioma , dysembryoplastic neuroepithelial tumor (DNET) and high grade glioma[17,28,30]. Low grade ependymoma are many times confused with angiocentric glioma and sometimes molecular studies are required to differentiate them conclusively.

Prognosis depends on location of tumor, extent of surgical resection, age at diagnosis and histological grade of tumor. In general prognosis of STE is better than infratentorial ependymoma in view of superficial parenchymal location and high chances of complete excision. 5 year survival rate for gross total excision ranges from 66-86% and subtotal

excision is 11-75% in small series published on STE[14,19,20,28]. Radiotherapy in dose of 54Gy is recommended atleast 1 month after surgery in case of anaplastic STE. Recurrence is well known in STE even after gross total resection. Even Post-operative radiotherapy after gross total excision cannot guarantee life without recurrence in anaplastic STE [10,11,26]. In the current series 2 patients required resurgery in view of recurrence. Surgical results in the current series is comparable to other series on these complex lesions. STE who have a benign course ad present late have a favorable course as compared to those that present within 1 month of worsening symptoms[13–16,31].

Surgical excision is the treatment modality of choice and post-operative radiotherapy is recommended for anaplastic astrocytoma grade 3. 8 out of 10 patients received both chemotherapy and radiotherapy in our study . Due to neurotoxicity of radiotherapy in pediatric population it may be withheld in cases with complete resection in grade 1 or grade 2 STE. This series is the largest by a single Neurosurgeon on these complex supratentorial tumors[11–13,31].

CONCLUSION

A differential diagnosis of intraparenchymal Ependymoma should always be considered in lobar tumors. Authors feel that with technical advances in micro neurosurgery and neuroanesthesia maximum safe resection with post-operative chemotherapy and radiotherapy should be offered to such cases. The prognosis of supratentorial ependymoma is better as compared to infratentorial lesions as in majority of cases a radical gross total resection can be achieved in view of easy accessibility and cortical location .This is the largest series on STE by a single Neurosurgeon till date in world literature.

REFERENCES

1. Metellus P, Guyotat J, Chinot O, Durand A, Barrie M, Giorgi R, et al. Adult intracranial WHO grade II ependymomas: long-term outcome and prognostic factor analysis in a series of 114 patients. Neuro-Oncol. 2010 Sep;12(9):976–84.
2. Lavrador JP, Oliveira E, Teixeira JC, Lopes JP, Pimentel J, Carvalho MH. Adult Supratentorial Extraventricular Anaplastic Ependymoma: Therapeutic Approach and Clinical Review. Asian J Neurosurg. 2018 Mar;13(1):105–9.
3. Cage TA, Clark AJ, Aranda D, Gupta N, Sun PP, Parsa AT, et al. A systematic review of treatment outcomes in pediatric patients with intracranial ependymomas. J Neurosurg Pediatr. 2013 Jun;11(6):673–81.
4. Sun S, Wang J, Zhu M, Beejadhursing R, Gao P, Zhang X, et al. Clinical, radiological, and histological features and treatment outcomes of supratentorial extraventricular ependymoma: 14 cases from a single center. J Neurosurg. 2017 Jul 7;1–7.
5. Bi Z, Ren X, Zhang J, Jia W. Clinical, radiological, and pathological features in 43 cases of intracranial subependymoma. J Neurosurg. 2015 Jan;122(1):49–60.
6. Matsumoto Y, Ichikawa T, Kurozumi K, Otani Y, Date I. Clinicopathological and Genetic Features of Supratentorial Cortical Ependymomas. World Neurosurg. 2019 May 28;
7. Hübner J-M, Kool M, Pfister SM, Pajtler KW. Epidemiology, molecular classification and WHO grading of ependymoma. J Neurosurg Sci. 2018 Feb;62(1):46–50.
8. Mangalore S, Aryan S, Prasad C, Santosh V. Imaging characteristics of supratentorial ependymomas: Study on a large single institutional cohort with histopathological correlation. Asian J Neurosurg. 2015 Dec;10(4):276–81.
9. Toescu SM, James G, Phipps K, Jeelani O, Thompson D, Hayward R, et al. Intracranial Neoplasms in the First Year of Life: Results of a Third Cohort of Patients From a Single Institution. Neurosurgery. 2019 Mar 1;84(3):636–46.
10. Pajtler KW, Mack SC, Ramaswamy V, Smith CA, Witt H, Smith A, et al. The current consensus on the clinical management of intracranial ependymoma and its distinct molecular variants. Acta Neuropathol (Berl). 2017;133(1):5–12.
11. Chakraborty A, Harkness W, Phipps K. Surgical management of supratentorial ependymomas. Childs Nerv Syst ChNS Off J Int Soc Pediatr Neurosurg. 2009 Oct;25(10):1215–20.
12. Khatri D, Bhaisora K, Das KK, Pandey S, Sardhara J, Mehrotra A, et al. Surgical outcome in cortical ependymoma: A single centre experience of 18 cases. J Clin Neurosci Off J Neurosurg Soc Australas. 2019 Feb;60:49–57.
13. Zamora C, Huisman TAGM, Izbudak I. Supratentorial Tumors in Pediatric Patients. Neuroimaging Clin N Am. 2017 Feb;27(1):39–67.
14. Nakamizo S, Sasayama T, Kondoh T, Inoue S, Shiomi R, Tanaka H, et al. Supratentorial pure cortical ependymoma. J Clin Neurosci Off J Neurosurg Soc Australas. 2012 Oct;19(10):1453–5.
15. Yin L, Li C, Luo S, Ma Z. Supratentorial Parenchymal Ependymomas in Children: Experience with 14 Patients at a Single Institute. Turk Neurosurg. 2017;27(2):272–8.
16. Hollon T, Nguyen V, Smith BW, Lewis S, Junck L, Orringer DA. Supratentorial hemispheric ependymomas: an analysis of 109 adults for survival and prognostic factors. J Neurosurg. 2016;125(2):410–8.
17. Leeper H, Felicella MM, Walbert T. Recent Advances in the Classification and Treatment of Ependymomas. Curr Treat Options Oncol. 2017 10;18(9):55.
18. Liu Z, Li J, Liu Z, Wang Q, Famer P, Mehta A, et al. Supratentorial cortical ependymoma: case series and review of the literature. Neuropathol Off J Jpn Soc Neuropathol. 2014 Jun;34(3):243–52.
19. Alexiou GA, Stefanaki K, Prodromou N. Prognostic factors in supratentorial ependymomas. Pediatr Neurosurg. 2013;49(2):129.
20. Sala F, Talacchi A, Mazza C, Prisco R, Ghimenton C, Bricolo A. Prognostic factors in childhood intracranial ependymomas: the role of age and tumor location. Pediatr Neurosurg. 1998 Mar;28(3):135–42.
21. Sayegh ET, Aranda D, Kim JM, Oh T, Parsa AT, Oh MC. Prognosis by tumor location in adults with intracranial ependymomas. J Clin Neurosci Off J Neurosurg Soc Australas. 2014 Dec;21(12):2096–101.
22. Aizer AA, Ancukiewicz M, Nguyen PL, Macdonald SM, Yock TI, Tarbell NJ, et al. Natural history and role of radiation in patients with supratentorial and infratentorial WHO grade II ependymomas: results from a population-based study. J Neurooncol. 2013 Dec;115(3):411–9.
23. Leng X, Tan X, Zhang C, Lin H, Qiu S. Magnetic resonance imaging findings of extraventricular anaplastic ependymoma: A report of 11 cases. Oncol Lett. 2016 Sep;12(3):2048–54.
24. Yecies D, Azad TD, Esparza R, Quon JL, Forkert ND, MacEachern SJ, et al. Long-Term Supratentorial Radiologic Effects of Surgery and Local Radiation in Children with Infratentorial Ependymoma. World Neurosurg. 2019 Feb;122:e1300–4.
25. Khatua S, Mangum R, Bertrand KC, Zaky W, McCall D, Mack SC. Pediatric ependymoma: current treatment and newer therapeutic insights. Future Oncol Lond Engl. 2018 Dec;14(30):3175–86.
26. Byun J, Kim JH, Kim Y-H, Cho YH, Hong SH, Kim CJ. Supratentorial Extraventricular Ependymoma: Retrospective Analysis of 15 Patients at a Single Institution. World Neurosurg. 2018 Oct;118:e1–9.
27. Khilji MF, Hamid RS, Qureshi A. Supratentorial extraventricular anaplastic ependymoma in a child. BMJ Case Rep. 2014 Mar 12;2014.
28. Alexiou GA, Moschovi M, Stefanaki K, Panagopoulos D, Tsotra M, Siozos G, et al. Supratentorial ependymomas in children: Analysis of nine cases. J Pediatr Neurosci. 2013;8(1):15–8.
29. Wang M, Zhang R, Liu X, Li D, Zhao P, Zuo Y, et al. Supratentorial Cortical Ependymomas: A Retrospective Series of 13 Cases at a Single Center. World Neurosurg. 2018 Apr;112:e772–7.
30. Landau E, Boop FA, Conklin HM, Wu S, Xiong X, Merchant TE. Supratentorial ependymoma: disease control, complications, and functional outcomes after irradiation. Int J Radiat Oncol Biol Phys. 2013 Mar 15;85(4):e193-199.
31. Ghia AJ, Mahajan A, Allen PK, Armstrong TS, Lang FF, Gilbert MR, et al. Supratentorial gross-totally resected non-anaplastic ependymoma: population based patterns of care and outcomes analysis. J Neurooncol. 2013 Dec;115(3):513–20.
32. Wang M, Zhang R, Liu X, Li D, Qiu C, Zhao P, et al. Supratentorial extraventricular ependymomas: A retrospective study focused on long-term outcomes and prognostic factors. Clin Neurol Neurosurg. 2018;165:1–6.