CASE REPORT

Supratentorial Recurrence of Medulloblastoma, A Rare Case Report

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ABSTRACT

Medulloblastoma is the most common brain tumor in children accounting for approximately 20% of all childhood brain tumors and 40% of all paediatric posterior fossa tumours with 70% of cases below 10 years. The peak age incidence is 5 years. Initial therapy includes surgical resection and radiation of the entire neuro-axis. Recurrence is common and typically occurs within 2 years of initial diagnosis. We report a case of supratentorial recurrence 5 years after initial diagnosis. A 17 year old male presenting 5 years after initial diagnosis with isolated right frontal recurrence. Late recurrence to the supratentorial region is uncommon and long term follow-up is recommended in these patients.

INTRODUCTION

Medulloblastoma is the most common brain tumor in children accounting for approximately 20% of all childhood brain tumors and 40% of all pediatric posterior fossa tumours with 70% of all cases below 10 years[1]. Occurrence in the adult population is also well documented, but only accounts for 1% of adult tumors.[1] Medolloblastoma is very aggressive tumour and recurrence is common even after adequate recommended treatment. Treatment includes surgical resection followed by radiotherapy of the entire neuro-axis with chemotherapy.[2,3,4,5] Recurrence of this tumor is well recognized and may require salvage therapy. Time to recurrence typically occurs within two years of initial diagnosis in the pediatric population. The locations of recurrence most commonly present as posterior fossa, spinal, supratentorial and bony metastases.[5] Spinal seedling is common but supratentorial recurrence is a rare phenomenon. Supratentorial recurrence is reported to be in the frontal and subfrontal region.[4,5]

Case report: A 17-year-old male with a history of surgery for posterior fossa space occupying lesion (Figure 1) at 12 years of age, removed surgically with histopathology suggestive of medulloblastoma and treated with adjuvant craniospinal irradiation with chemotherapy presented with history of convulsion, headache and left sided weakness for 7 days. Patient was conscious and oriented. The right pupil was semidilated and sluggish reacting and left pupil was normal size and normally reacting. The left upper extremity had 4/5 strength. MRI Scan showed a space occupying lesion in right frontal region (figure 2). Patient was operated with near total excision of tumor followed by radiotherapy. Patient is kept on regular follow up and is doing well and post-operative MRI scan suggested total clearance (figure 3)

Figure 1.

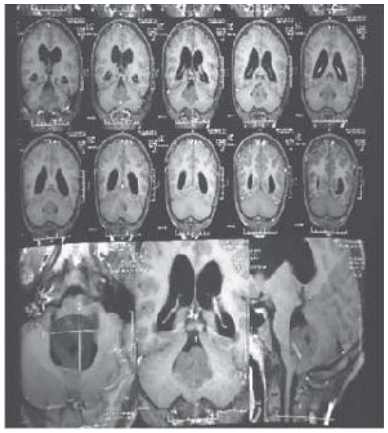

Figure 2.

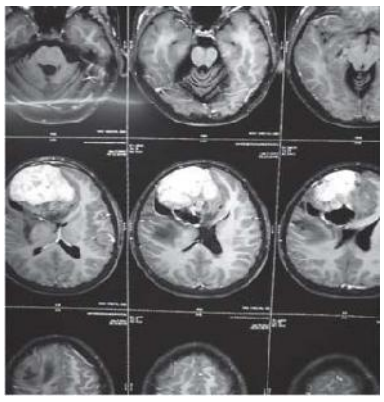

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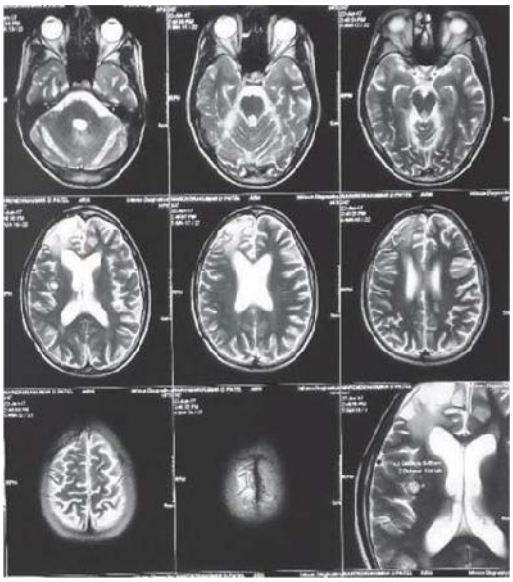
Figure 3



DISCUSSION

Medulloblastoma, a malignant tumor typically arising from the cerebellar vermis in young children and lobes in older children, represents 4–8% of all intracranial tumors, and is the most common malignant central nervous system tumor of childhood, with approximately 80% occurring in patients under 15 years of age.4,6,7 It accounts for 15–25% of all childhood brain tumors in comparison with only 1% of adult intracranial neoplasms, with a slight male predominance in both groups.4,6,7,8 Although primary treatment of medulloblastoma is successful in a high percentage of patients, but early recurrence is seen in adults. Tumor recurrence may be either local or metastatic.9 Presenting features of medulloblastoma are generally related to hydrocephalus (found radiographically in 97% of patients) and cerebellar dysfunction which include: headache, nausea/vomiting, truncal ataxia, and unsteady gait.5 Approximately 83% are found in the midline in children, while 49% of adult tumors are lateral.7

Spinal metastasis are commoner than supratentorial metastasis.10,11 Supratentorial metastasis occur commonly in the frontal lobe, subfrontal region near the orbital roof or cribriform plate.4,5,12 Leptomeningeal metastasis are more common than supratentorial recurrences which appear as mass lesions.13,14 Medulloblastoma tends to seed the cerebrospinal fluid resulting in extensive leptomeningeal involvement, and treatment therefore requires surgical resection followed by neuro-axis radiotherapy and chemotherapy.15,16

With the advent of CT scan and MRI, supratentorial metastasis would be seen more commonly than presumed earlier.17 Frequent supratentorial recurrences seen in the frontal and the subfrontal region have been attributed to the pooling of the tumor cells in the prone position in the frontal region and also due to under-dose of radiation to that region.12,18 Hence it is recommended that medial frontal-basal cisterns to be included in the radiotherapeutic regime.

Age at diagnosis has been shown to be a significant predictor of time until relapse.8 The majorities of pediatric recurrences are within two years of initial diagnosis, and are in the posterior fossa.4,6,8 Those with tumor-free period equal to the age at diagnosis plus nine months may be considered cured (Collin's Law). Treatment of the recurrence with radiation, chemotherapy, or surgery can produce useful palliation in some patients.19 In view of the incidence of local recurrences, Sure (1995) recommended 3 monthly scan of neuraxis in the first three postoperative years and six monthly scans thereafter to catch early recurrences and metastatic disease.18

CONCLUSION

Recurrence of medulloblastoma is common and often occurs in the posterior fossa. Spinal dissemination of medulloblastoma is more common than supratentorial recurrences and known as drop metastasis. Supratentorial recurrence of medulloblastoma in frontal region is a rare phenomenon. We present a case of a 17 years old male who presented 5 years following his initial diagnosis of medulloblastoma with symptoms of supratentorial recurrence. Continued close follow-up is needed in pediatric patients after resection and radiation for medulloblastoma given their unexpected recurrence pattern.

REFERENCE

1. Giordana MT, Schiffer P, Lanotte M, Girardi P, Chio A. Epidemiology of adult medulloblastoma. Int J Cancer. 1999;80:689–92.
2. Brown WD, Tavaré CJ, Sobel EL, Gilles FH. Medulloblastoma and Collins' law : a critical review of the concept of a period of risk for tumor recurrence and patient survival. Neurosurgery. 1995;36:691–7.
3. Buglione M, Triggiani L, Grisanti S, Liserre R, Buttolo L, Gipponi S, et al. Retreatment of recurrent adult medulloblastoma with radiotherapy : a case report and review of the literature. J Med Case Rep. 2013;7:64.
4. Roka YB, Bista P, Sharma GR, Adhikari D, Kumar P. Frontal recurrence of medulloblastoma five years after excision and craniospinal irradiation. Indian J Pathol Microbiol. 2009;52:383–5.
5. Sun LM, Yeh SA, Wang CJ, Huang EY, Chen HC, Hsu HC, et al. Postoperative radiation therapy for medulloblastoma--high recurrence rate in the subfrontal region. J Neurooncol. 2002;58:77–85.
6. Sanders MA, Vitaz T, Rosenblum M, Plaga AR, Parker JC, Jr, Parker JR. 46-year-old man with a spinal cord mass. Brain Pathol. 2011;21:109–12.

7. Sarkar C, Pramanik P, Karak AK, Mukhopadhyay P, Sharma MC, Singh VP, et al. Are childhood and adult medulloblastomas different? A comparative study of clinicopathological features, proliferation index and apoptotic index. J Neurooncol. 2002;59:49–61.
8. Tabori U, Sung L, Hukin J, Laperriere N, Crooks B, Carret AS, et al. Distinctive clinical course and pattern of relapse in adolescents with medulloblastoma. Int J Radiat Oncol Biol Phys. 2006;64:402–7.
9. Albright AL, Wisoff JH, Zeltzer PN, Boyett JM, Rorke LB, Stanley P. Effects of medulloblastoma resection on outcome in children: A report from Children's Cancer Group. Neurosurgery. 1996;38:265–71.
10. Arseni C, Constantinescu A, Danaila L. Supratentorial dissemination of medulloblastoma. Neurol Neurochir Psychiatr. 1976;119:299–306.
11. Garcia EM, Diaz GJ, Soto TO, Simon de las HR, Mateos BF, Munoz A, et al. Cerebellar medulloblastomas in childhood: Supratentorial metastasis. Neurologia. 1993;8:131–4.
12. Hardy DJ, Hope Stone HF, McKenzie CG, Scholtz CL. Recurrence of medulloblastoma after homogenous field radiotherapy. Report of three cases. J Neurosurg. 1978;49:434–40.
13. Meyers SP, Wildenhain S, Chess MA, Tarr RW. Post-operative evaluation for intracranial recurrence of medulloblastoma: MR findings with gadopentatedimeglumine. ANJR Am J Neuroradiol. 1994;15:1425–34.
14. Gerlach R, Kieslich M, Van de Nes J, Galow W, Seifert V. Supratentorial leptomeningeal metastasis of medulloblastoma without cerebellar tumor recurrence. Acta Neurochir (Wien) 2002;144:201–4.
15. David KM, Casey AT, Hayward RD, Harkness WF, Phipps K, Wade AM. Medulloblastoma : is the 5-year survival rate improving? A review of 80 cases from a single institution. J Neurosurg. 1997;86:13–21.
16. Nigri F, Telles C, Acioly MA. Late obstruction of an endoscopic third ventriculostomy stoma by metastatic seeding of a recurrent medulloblastoma. J Neurosurg Pediatr. 2010;5:6 [
17. Clark EE, Hattner RS. Brain scintigraphy in recurrent medulloblastoma. Radiology. 1976;119:633–6.
18. Sure U, Bertalanffy H, Isenmann S, Brandner S, Berghorn WJ, Seegar W, et al. Secondary manifestation of medulloblastoma: Metastasis and local recurrence in 66 patients. Acta Neurochir (Wien) 1995;136:117–26.
19. Mealey JJr, Hall PV. Medulloblastoma in children. Survival and treatment. J Neurosurg. 1977;46:56–64. Mealey JJr, Hall PV. Medulloblastoma in children. Survival and treatment. J Neurosurg. 1977;46:56–64.