Romanian Neurosurgery | Volume XXX | Number 4 | 2016 | October - December

Article

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INDIA

DE GRUYTER OPEN

DOI: 10.1515/romneu-2016-0090

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Adult medulloblastoma

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Abstract: Medulloblastoma is a highly malignant central nervous system (CNS) tumor that arises from the cerebellum. It is the most common primary malignant intracranial childhood neoplasm. In adults, medulloblastoma are much less common, accounting for < 1% of all adult brain tumors. Herein, author has described a rare case of cerebellar medulloblastoma in adult.

Key words: Adult, Medulloblastoma, Posterior fossa

Introduction

Medulloblastoma is the most common CNS tumor of childhood, accounting for 15% to 30% of all childhood brain tumors, and 30% to 40% of all posterior fossa tumors. (1) In adults, the tumor is much less common, accounting for approximately 1% of adult primary brain tumors, 80% of which occur before the end of the fourth decade. (2) The adult tumor is said to occur more often in the cerebellar hemispheres than the vermis and is more often desmoplastic in histology. The exact etiology of medulloblastomas is not known. Herein, author has described a rare case of cerebellar medulloblastoma in adult.

Case report

A 45 years old male presented in neurosurgery department with headache and gait disturbance for last three months. Neurological examination revealed cerebellar signs with ataxic gait.

Magnetic resonance imaging (MRI) of brain showed heterogeneous intensity mass lesion in posterior fossa mainly involving midline (figures 1 and 2). In view of symptoms patient was taken up for surgery.

Gross total excision of the tumour was done via midline suboccipital craniectomy. Intraoperatively tumour was greyish white, mainly present in midline involving vermis and extending into the right cerebellum (figure 3). Tumour was sent for histopathological examination which revealed medulloblastoma desmoplastic variety. After the surgery, the patient was alert, his neurological examination showed only mild truncal ataxia. No obvious mass lesion was present in postoperative imaging of brain (figure 4). To rule out drop metastasis, MRI whole spine was done which was essentially normal (figure 5). After suture removal patient was sent for radiotherapy and no recurrence was found in 6 months of follow up.

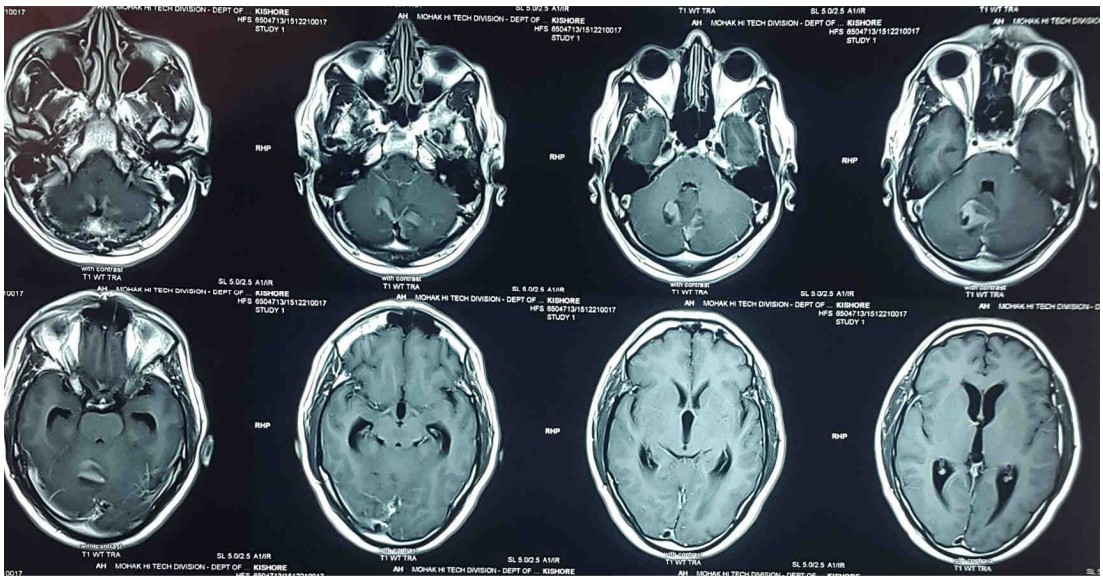

Figure 1 - Post contrast MRI brain axial section showing heterogeneous intensity mass lesion in posterior fossa mainly involving midline

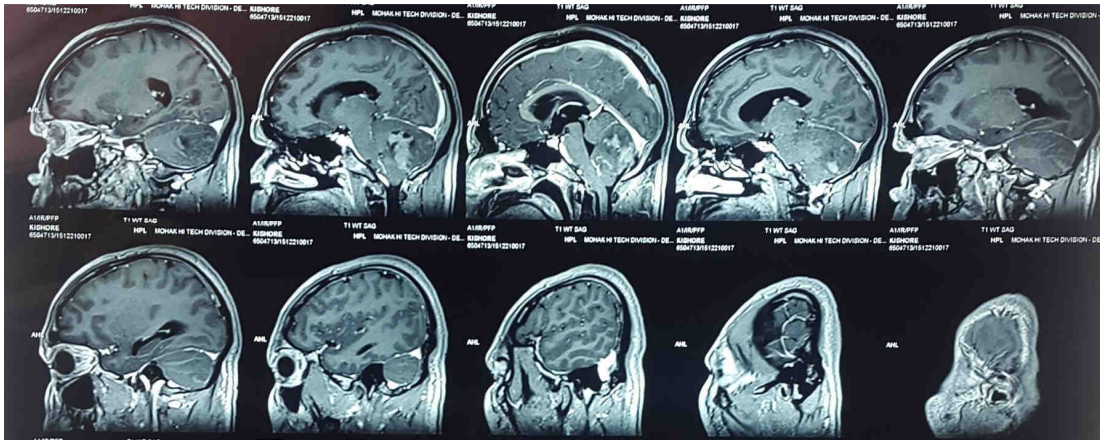

Figure 2 - Post contrast MRI brain sagittal section showing midline cerebellar mass

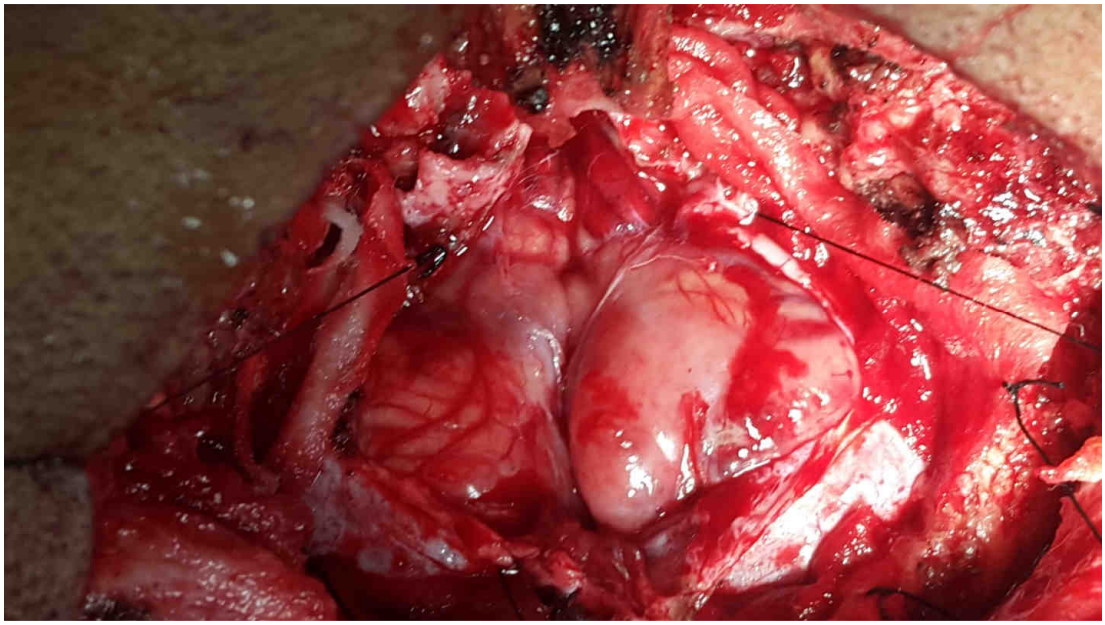

Figure 3 - Intra operative image showing greyish white mass, mainly present in midline involving vermis and extending into the right cerebellum

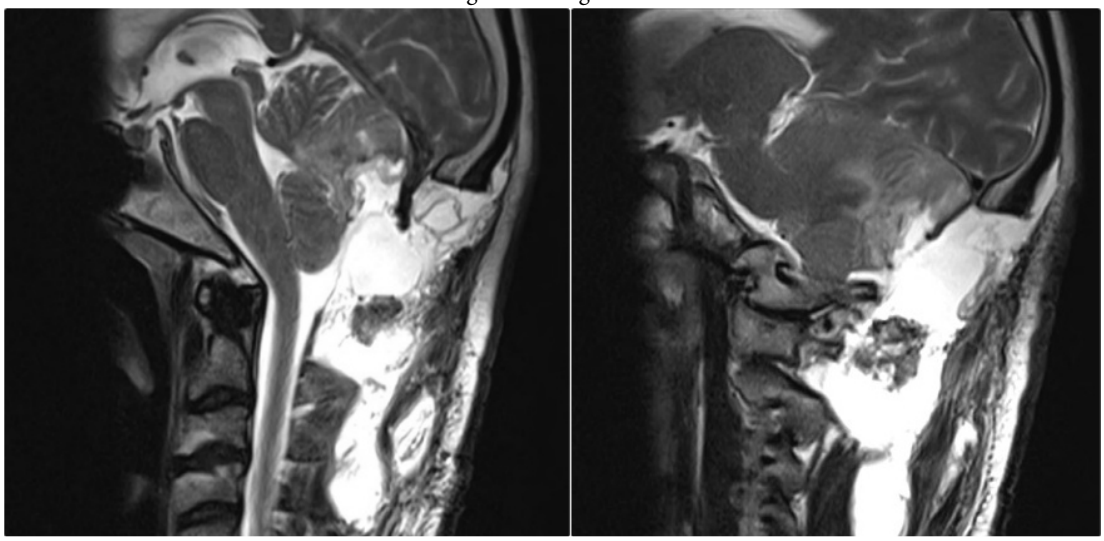

Figure 4 - Postoperative image showing no obvious mass lesion

Figure 5 - MRI whole spine showing no drop metastasis

Discussion

Medulloblastoma is a highly malignant CNS tumor that arises from the cerebellum. It is the most common primary malignant intracranial childhood neoplasm, accounting for 25% of all childhood tumors. (1) More than 80% of medulloblastomas are diagnosed in children during the first 15 years of life, the median age at diagnosis being 5 years. In adults (patients ≥ 16 years of age), medulloblastomas are much less common, accounting for < 1% of all adult brain tumors. The incidence of adult medulloblastomas is approximately 0.5 per million per year, and decreases with increasing age. The exact etiology of medulloblastomas is not known. (2)

Medulloblastoma is thought to arise from a primitive cell in the external granular layer of the cerebellum, but the specific cell of origin has not been identified. Pathologically medulloblastomas consist of small round blue cells. It has two variants - classical and desmoplastic. The latter variant has large amounts of reticulin and collagen.3 Medulloblastoma is located in posterior fossa thus it often produces hydrocephalus and symptoms of increased ICP, as well as cerebellar signs (eg, truncal and limb ataxia, nystagmus) (4) which was present in our case.

Disease may exist as nodular lesions outside the primary site anywhere within the neuraxis, including the spine and supratentorial cerebrum, and may involve the CSF.4 In our case there was no drop metastasis in spine.

Magnetic resonance imaging (MRI) brain is the investigation of choice. Lesions are usually ISO to hypointense on T1WI, have variable signal on T2WI, and frequently demonstrate contrast enhancement. These tumors tend to be homogenous in appearance, with occasional cystic or necrotic areas. (5) Surgery is the treatment of choice. The goal of surgery is to remove all visible tumour. Survival of adult medulloblastoma patients may be influenced by the extent of residual disease following surgery, particularly for patients without evidence of dissemination.

Treatment of associated hydrocephalus can be managed by external drainage with tumor decompression, tumor decompression alone, or by the use of various shunting procedures. (6) In our case gross total excision of tumour was done.

Craniospinal axis radiation therapy is the standard of care for the treatment of adult medulloblastoma patients, as medulloblastomas are quite radiosensitive. Standard radiation doses to the craniospinal

axis involve delivering 35 to 45 Gy to the brain and 30 to 40 Gy to the spine, with a dose of 54 Gy to the primary tumor site. (6) In contrast to childhood medulloblastoma, the role of chemotherapy in adult medulloblastoma patients is undetermined. Prados and colleagues found that those patients who received adjuvant chemotherapy (mostly nitrosourea-based regimens) had a statistically significantly longer survival as compared to those who did not receive adjuvant chemotherapy. (7) In our case, patient was received craniospinal irradiation.

The recurrence rate for medulloblastomas in adults is approximately 50% to 60%. The median survival after recurrence has been reported to be approximately 1.3 years. The most common site of recurrence is the posterior fossa. Other sites of recurrence include the spine, CSF, supratentorial cerebrum, bone, and other extraneural sites. (8) In our case recurrence was not found after 6 month of follow up.

Conclusion

Adult variety of medulloblastomas have notable differences as compared to the pediatric type. There is higher frequency of lateral cerebellar location, desmoplastic histology, and late recurrences in the adult variety. The 5-year overall survival rates are comparable between the adult and pediatric population.The management of adult medulloblastoma patients involves a thorough staging work-up (including brain and whole-spine MRI and CSF cytology analysis), as complete a resection as possible, and postoperative craniospinal axia (CSA) irradiation.

Nitrosourea-based chemotherapy is reserved for recurrent disease, as adjuvant chemotherapy has not shown significant benefit in adult medulloblastoma patients.

Prospective studies are needed to accurately define significant prognostic factors and treatment regimens for adult patients with medulloblastomas.

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