Supratentorial metastasis of medulloblastoma in adults

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Abstract

Two adults, 31 and 20 years of age, developed supratentorial metastasis 3½ years and 11 months, respectively, after gross total removal of their posterior fossa medulloblastoma. The first case developed spinal metastasis as well. Both had undergone craniospinal irradiation. Case 1 underwent laminectomy and case 2 underwent craniotomy because their presenting symptoms required so.

Keywords: Cerebellar mutism, medulloblastoma, supratentorial metastasis

Introduction

Medulloblastoma is the most common brain tumor in children. Surgical gross total resection followed by irradiation and chemotherapy are the treatment modalities. Medulloblastomas are very aggressive tumors (WHO Grade IV) and recurrences are common even after adequate recommended treatment. Most of the recurrences occur within 2 years.[1] Spinal seeding is common; however, seeding into the supratentorial compartment is rare in adults. We report here two such cases with supratentorial seeding. In one of our patient, there was local recurrence and spinal seeding as well and the other developed cerebellar mutism which also is rare in adults.[2]

Case Reports

Case 1

A 31-year-old patient presented with rapidly progressive weakness of both lower limbs of 2 days duration and urinary incontinence of 1-day duration. He had been operated for a posterior fossa mass 3½ years back and the histopathological diagnosis was medulloblastoma which was of the desmoplastic variety. Postoperatively, he had received radiotherapy and he had not developed any new symptoms till 2 days prior to the present admission. On examination, there was flaccid paraplegia and plantars were not elicitable. MRI brain revealed local recurrence, subfrontal enhancing mass and intradural deposit at D11-12 level [Figure 1]. At laminectomy, a vascular solid mass was removed from the dorsal aspect of the cord. His motor power improved to grade III and was given a course of radiotherapy. Histopathology of the tumor was medulloblastoma which was also of the desmoplastic variety. Six months later, he developed hemoptysis, became unconsciousness, and died a few days later.

Case 2

A 20-year-old male presented with headache, vomiting, and diplopia of 1-month duration. On examination, he was conscious, had bilateral papilledema and cerebellar signs more marked on the left side. Computed tomography revealed a midline vermian mass [Figure 2a]. Ventriculoperitoneal shunt was done. Posterior
Fossa surgery was performed in the sitting position and the lesion was removed totally. Histopathological examination confirmed the diagnosis of medulloblastoma which was of the large cell variety. He developed aphasia 24 hours after surgery without any pyramidal or sensory symptoms. After 1 month, aphasia improved to dysarthria. Postoperatively, he received craniospinal irradiation. Eleven months later, he was re-admitted with complaints of headache and occasional vomiting of 2 months duration. On examination, he was conscious and his fundus was normal. He had slurred speech and cerebellar signs were still present. Computed tomography revealed an enhancing temporal lobe mass [Figure 2b]. Operation revealed a firm vascular mass attached to the temporal basal dura. Histopathological examination revealed it to be medulloblastoma of the large cell variety, similar to the previous posterior fossa lesion. Patient was re-admitted 8 months later in a drowsy state. MRI revealed a recurrence of lesion in the temporal, bifrontal, and biparietal region [Figure 2c]. He was treated conservatively and expired 2 weeks later.

Discussion

Although primary treatment of medulloblastoma is successful in a high percentage of patients, it is not successful in adults because of early recurrence. Tumor recurrence may be either local or metastatic.[3] Spinal metastasis are commoner than supratentorial metastasis.[4,5] Supratentorial metastasis occur commonly in the frontal lobe, subfrontal region near the orbital roof or cribriform plate.[6,7,8] Leptomeningeal metastasis are more common than supratentorial recurrences which appear as mass lesions.[9,10] Kunscher et al. (2001) observed that recurrences of medulloblastoma were more common in the posterior fossa (56%) followed by bone marrow (25%).[11] Emmenzuel et al. (2006) reported a metastatic medulloblastoma in the frontobasal region 21 years after the treatment of cerebellar medulloblastoma; however, most of the supratentorial recurrences have been reported within the first two years following surgery for the primary lesion.[1,12]

With the advent of CT scan and MRI, supratentorial metastasis would be seen more commonly than presumed earlier.[13] 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.[1,6] Hence, they recommended that medial frontal-basal cisterns be included in the radiotherapeutic regime. Our patients were operated in the sitting position, so the former explanation would not be applicable to them. The role of ventricular pathway in metastasis has also been suggested.[5]

Treatment of the recurrence with radiation, chemotherapy, or surgery can produce useful palliation in some patients.[14] In view of the incidences 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.

Mutism after posterior fossa surgery is rare and most of the reported cases have been in children. Kai et al. (1997) reported two such cases in elderly and could collect only 10 cases of cerebellar mutism in adults following surgery. The syndrome of cerebellar mutism is not tumor specific; however, it is seen more commonly with medulloblastomas. Mutism is followed by dysarthria and usually recovers completely. However, Steinbok et al. (2003) reported seven cases of cerebellar mutism in which mutism did not recover in all except one case.[15] In one patient, mutism recovered to dysarthria which persisted for ½ years. Splitting of inferior vermis may result in oropharyngeal apraxia with mutism. Damage to superior vermis, paravermian area, superior cerebellar peduncle, and dentate nucleus also results in cerebellar mutism involving dentato-rubro-thalamo-cortical pathways.[16]

Footnotes

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References


**Figures and Tables**
Contrast MRI showing (a) Recurrence in the subfrontal region (3½ years after surgery), (b) Intradural deposit at D11-12, and (c) Local recurrence
Computed tomogram showing (a) Large midline vermian enhancing mass, (b) Enhancing mass in the left temporal region 11 months following surgery, and (c) Contrast MRI showing recurrence in the right frontal and left frontotemporal region with mass effect (20 months after the first surgery)