

## BMR Medicine

### Case Report

# Long Bone Metastasis in a Case of Medulloblastoma - A rare case report

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### Abstract

We reported a case of medulloblastoma with upper end of tibia and lower end of femur metastasis. This was treated with CSI (craniospinal irradiation) two years back. After two year of follow-up he presented with pain over knee joint which was confirmed as metastasis to lower end of femur and upper end of tibia and then treated with palliative radiotherapy. Patient brain MRI shows thalamus mass suggestive of metastasis which was treated with palliative chemotherapy. Our case is unique in presentation because it presented with long bone metastasis.

**Key words:** Medulloblastoma, Long bone metastasis, Radiotherapy

### Introduction

Medulloblastoma accounts for 15% to 20% of all CNS tumours in the paediatric age group. The median age at presentation is 6 years. In the majority of cases the tumour arises in the cerebellar vermis and projects into the fourth ventricle. Patients typically present with symptoms and signs of raised intracranial pressure, that is, headache and vomiting. On CT and MRI medulloblastomas appear as solid masses that enhance usually fairly homogeneously with contrast material. The frequency of spinal seeding at diagnosis is

approximately 30% to 35%, and investigation at diagnosis must include a gadolinium-enhanced MRI of the spinal axis and CSF cytology. Medulloblastoma is one of the few CNS tumours to spread outside the CNS (to lymph nodes, bone), although this is an uncommon event. Those who have undergone complete or subtotal resection with <1.5 cm<sup>2</sup> of residual tumour and no evidence of CSF dissemination (M0) are considered to have standard-risk disease, whereas patients who have larger volume residual tumour and those with evidence of CSF dissemination at diagnosis are characterized as high risk. Surgical resection

followed by craniospinal irradiation is standard of care in medulloblastoma.

### **Case Report**

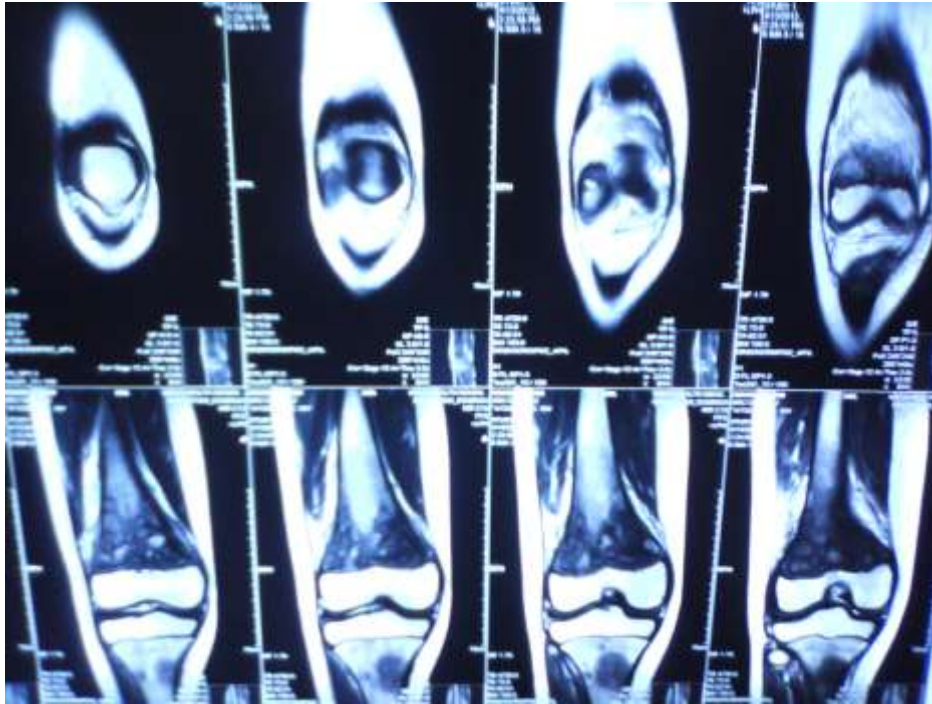
A 9 year old boy presented in neurosurgery OPD with complaining of headache and vomiting. On physical examination his vitals was stable. On Ophthalmological examination his vision was normal and bilateral papilledema was present no other neurological deficit present.

His routine blood count, liver function test, renal function test were with a normal limits. Brain CT scan shows contrast enhancing lesion in brain stem suggestive of brain stem glioma, biopsy was done by Neurosurgeon shows medullablastoma. CSF cytology was negative for malignant cell with this information patient was planned CSI (Craniospinal irradiation) using cobalt-60 machine with a dose of 36Gy in 20 fractions to whole brain & spine and 19.8Gy in 11 fractions boost to posterior fossa in 2011. The course of radiotherapy was uneventful, after completion of radiotherapy,

patient was advised for regular follow-up, and patient was asymptomatic for a period of two year. During last follow-up patient complaining of pain over left knee joint for that patient was investigated with x-ray knee joint shows radiolucent area in lower end of femur & upper end of tibia which was confirmed by MRI, which shows altered signal intensity lower part of shaft of femur in diaphysis region with periosteal elevation, periosteal, subperiosteal oedema and altered signal intensity shows upper part of tibia [Figure No. 1]. Biopsy was done showing metastatic deposit of Medullablastoma. MRI brain also done shows multiple nodules size of 10-15 mm T1W1 hypo intense alter signal intensity lesion are noted in medial aspect of right thalamus, likely to metastasis. He was planned for palliative radiotherapy to lower end of femur & upper end of tibia to a dose of 30Gy in 10 fractions over 2 weeks and for brain metastasis patient was planned for palliative chemotherapy, carboplatin and etoposide based regimen, now patient on this regimen and completed two cycle of chemotherapy.



**Figure:1**



**Figure: 2**

## Discussion

Medulloblastoma is one of most common posterior fossa tumour in childhood which is known to extra cranial metastasis. Bone was most common site of metastasis. Lymph node metastases were second in frequency followed by lung and muscle. The lesions usually involved the axial skeleton, the pelvic and shoulder girdle & adjacent ends of long bones, occasionally; there was periosteal new bone formation. In the majority of cases the tumour arises in the cerebellum vermis and projects into the fourth ventricle. Patients typically present with symptoms and signs of raised intracranial pressure that is headache and morning vomiting. On CT and MRI medulloblastomas appear as solid masses that enhance usually fairly homogeneously with contrast material.

In 1930 Cushing was the first to report the use of total CNS irradiation in medulloblastoma. Currently the standard therapy for medulloblastoma after surgery includes postoperative Cranio-spinal irradiation [1] which has a five year survival rate of between 40 to 60%. Adjuvant treatment in medullo

blastoma is depend on risk factors which includes residual versus non-residual, metastases present or absent (M+, M-)

## Management of Standard-Risk Medulloblastoma:

Until relatively recently, the standard of care for patients older than 3 years with standard-risk disease consisted of postoperative radiotherapy to the craniospinal axis to a dose of 35 to 36 Gy followed by a boost to the whole posterior fossa to a total dose of 54 to 55.8 Gy. In multi-institution studies, such treatment results in long-term event-free survival in 60 to 65% of patients [2][3][4]. Sequelae of treatment include hormonal deficits, decreased bone growth, and neurocognitive deficits that correlate with the age of the child and the radiation dose [5].

## Management of High-Risk Medulloblastoma:

Patients with residual disease  $>1.5 \text{ cm}^2$  and/or those with evidence of leptomeningeal seeding (M+) are considered to have high-risk disease. This is the group of patients in which the use of chemotherapy was shown in the prospective randomized phase III studies conducted in the

1970s to result in significant improvement in disease-free survival would be logical to consider using a radiotherapy dose to residual disease in the posterior fossa higher than the standard 55.8 Gy, and this may be feasible using stereotactic radiotherapy or 3D conformal techniques. Patients with M1 disease may do well using a standard CSI dose of 35 to 36 Gy; in the POG 9031 study, event-free survival at 5 years was 65%. In contrast, results for patients with Metastasis disease remain quite poor, although the use of a higher radiotherapy dose (40 Gy CSI plus a boost of 5 Gy to macroscopic disease) produced excellent early results in POG 9031.

The treatment modalities for CSI in few centres have reported their results using protons for treatment of medulloblastoma. Although protons provide a dose distribution that cannot yet be achieved by the most sophisticated photon beam treatment planning (6), the major disadvantages of proton therapy are the restricted access and high cost. Hopefully, it will be possible in the future, using better dosimetry and IMRT, to achieve equivalent results with photons. Our case is unique in presentation because it presented with long bone metastasis.

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