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Cerebellar Medulloblastoma with Drop Metastasis to Sacral Canal – A Case Report

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Abstract: ***We present a case of 19year old male, known case of cerebellar medulloblastoma, operated one year back and treated with craniospinal radiotherapy which developed drop metastases to sacral canal. Histology study showed medulloblastoma as well as immunohistochemical study for Synaptophysin and Mib-1(Ki-67) was performed which confirmed the diagnosis.***

Keywords: Cerebellum, Medulloblastoma, Sacral metastasis, Immunohistochemistry

1. Introduction

Medulloblastoma is one of the commonest pediatric brain malignancies. Metastases of medulloblastoma can be found in both neuraxial and extraneural locations. Supratentorial metastasis is found in 14.6% of cases, intraspinal metastasis in 12.5% and systemic metastasis in 9.7 %.(1). We present an operated case of cerebellar medulloblastoma with postoperative craniospinal radiotherapy, who developed drop metastases to sacral canal.

2. Case Report

A 19 yr old boy, operated for cerebellar medulloblastoma one year back. (Fig. 1) The lesion was excised totally with histopathology report suggestive of desmoplastic medulloblastoma. Postoperatively, he received craniospinal radiation, although the spine imaging did not reveal any lesion. The CSF showed presence malignant cells. He presented to us with backache and paraesthesia on medial aspect of thigh progressing for the last two months. On examination, there was tenderness on sacral region with hypoesthesia in S2, S3 region.MRI of lumbo-sacral region showed, a well defined lesion in the sacral canal, intradural, extending from S1 to S4, Hyperintense on T2W, and isointense on T1W, with abnormal marrow signal from S2 to S4, suggestive of intradural sacral metastases (Fig. 2). The patient investigated by screening of brain, fundoscopy and other routine parameters and there was no brain recurrence, fundus was normal, and rest of the parameters were normal. He was operated and total excision of the lesion done.

Histopathology confirmed it to be a tumor composed of large, round to oval neoplastic cells arranged in nodules, diffuse sheets, small nests and clusters. The cells are round having high nuclear cytoplasmic ratio with vesicular nuclei & clumped chromatin, containing nucleoli and scanty amount of eosinophilic cytoplasm. Brisk mitotic figures are also noted. The intervening stroma shows blood vessels and areas of heamorrhages. Immunohistochemistry was performed which shows strong positivity for Synaptophysin and very high Mib(ki-67)%-30-35%

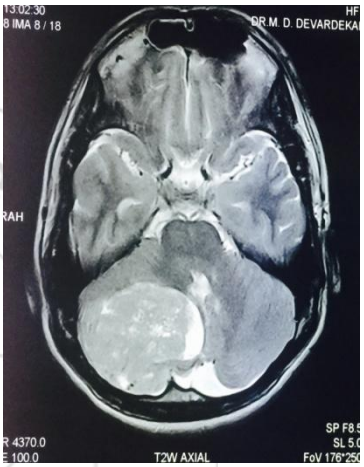

Figure 1: MRI showing right cerebellar hemisphere,intraaxial,hyperintense onT2w &enhancing heterogenously with contrast.

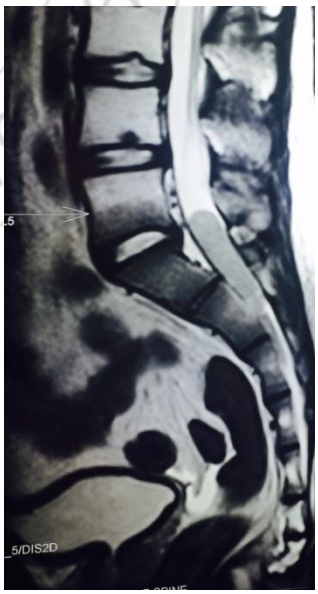

Figure 2: MRI showing lumbosacral spine showing intradural lesion in the sacral canal,iso to hyperintense onT2W images,enhancing with contrast

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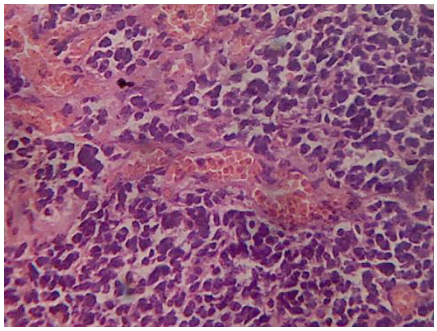

Figure 3: Microphotograph showing malignant round cells with high nucleo-cytoplasmic ratio &scanty amount of eosinophilic cytoplasm.

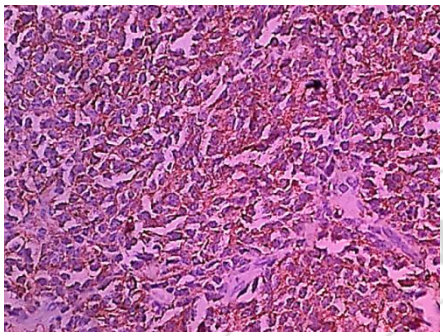

Figure 4: Microphotograph showing strong Synaptophysin

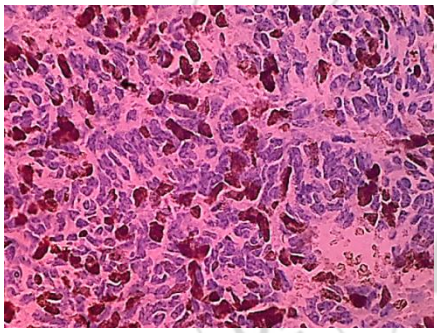

Figure 5: Microphotograph showing high Mib-1(Ki-67)

The patient was then subjected to chemotherapy.

3. Discussion

Common presenting features of medulloblastoma are abnormal gait, truncal ataxia and signs of raised intracranial pressure, including headache, vomiting, and papilloedema. Metastases of medulloblastoma can be found in both neuraxial and extraneural locations. Supratentorial metastasis is found in 14.6% of cases, intraspinal metastasis in 12.5% and systemic metastasis in 9.7 %.1. Medulloblastoma spread by local invasion, hematogenous dissemination, seeding along cerebrospinal fluid pathways and less likely by lymphatic dissemination2. Intraspinal spread from primary intracranial tumors by seeding along CSF pathways is well recognized at autopsy. In the lumbosacral region, nodularity and irregularity of the thecal sac is common, as is diffuse thickening and adhesions of the nerve roots and irregular obliteration of the subarachnoid space. The high incidence of involvement of the lumbosacral region shows the effect of gravity on CSF-borne metastases3. It is rare for them to have intramedullary spinal cord metastasis4. Pezesshkpour and collegues analyzed more than 18,000 primary central nervous system tumors and found that only 0.01% of them had drop spinal metastases, which had caused the presenting symptoms.1 At the time of diagnosis around 10 to 35%of the cases had extramedullary intradural metastases, however, their main presenting symptoms were due to the primary intracranial tumor.5 Stanley and collegues, in 1988, reported on 34 patients with medulloblastoma.6 Fifteen of those had a positive result on myelogram for spinal metastasis and only one patient suffered from lower limb weakness related to spinal pathology. There have been a limited number cases with spinal intramedullary metastasis reported in literature.7 Histopathology and immunohistochemistry are confirmatory for diagnosis.

At surgery, Medulloblastomas are soft, friable tumors, often with necrosis. They are highly cellular tumors with abundant dark staining, round or oval nuclei, and little cytoplasmic differentiation. The spectrum of histopathological appearance ranges from tumors with extensive nodularity to those with large cell/ anaplastic features. The clinical outcome appears to be worse with increasing grade and extent of anaplasia.7 Mitoses are often abundant and neuroblastic Homer Wright rosettes can be found in up to 40 percent of cases .8, 9

Immunohistochemical studies most often demonstrate the expression of the neuronal markers Synaptophysin and Neuron Specific Enolase, and Nestin, a marker of primitive neuroepithelial cells, consistent with their presumed origin from neuronal progenitors in the cerebellum.9 A majority of Medulloblastomas also express markers specific for cerebellar granule cells 10, 11 , supporting the conclusion that they arise most often by oncogenic transformation of cerebellar granule cells. Nuclear β-catenin staining is present in most wingless (WNT) pathway tumors, and p53 immunostaining can be performed to identify tumors with *TP53* mutations.

Several histological variants of medulloblastoma have been described .12 The desmoplastic variant has abundant collagen and reticulin in the interstitial spaces as well as reticulin free "pale islands" .9 This variant is associated with mutations in the patched-1 (*PTCH1*) gene on chromosome 9 and may have a better prognosis.13 A second variant, the large cell anaplastic (LCA) medulloblastoma, is characterized by cerebrospinal fluid dissemination and a more aggressive clinical course.9, 14 The LCA variant is most commonly associated with the group 3 molecular subtype.

In general, medulloblastoma spinal metastasis varied from nodular lesion to complete spinal block in order to have better staging, it is recommended to have a complete spine survey once a posterior fossa lesion with cisternal involvement is diagnosed.

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