Case Report

Spinal Myxopapillary Ependymoma Presentaing with Drop Metastasis & Cauda Eqina Syndrome: Case Report and Review of Literature

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ABSTRACT

Myxopapillary ependymoma is a benign slow-growing tumour, arising predominantly in the region of the filum terminale. It has been designated histologically as grade I neoplasm according to the 2007WHO classification (1). Despite this benign character dissemination and metastasis along the cerebrospinal axis and metastasis to distant sites have occasionally been reported (2). There have been previously reported cases of drop metastasis from MPE, however in 3 of these cases the drop metastasis was diagnosed with concurrent primary spinal MPE. There has been only one previously published case of interval drop metastasis in a case of operated spinal MPE in literature (3,4). We hereby present the second case of interval drop metastasis in a case of conus MPE, with history of having undergone a subtotal resection and post operative adjuvant radiotherapy prior to second surgery. The patient presented months after the primary surgery with symptoms of cauda equina syndrome attributable to the drop metastasis.

INTRODUCTION

Spinal ependymomas are rare tumors, with an incidence of approximately 0.2/100,000 population with a slight male predominance. Approximately 0.5% of all ependymomas are classified as myxopapillary, thus having an incidence of 0.01 per million population 3. No specific risk factors for the development of ependymoma have been identified, although associations with neurofibromatosis type 2 and lack of maternal consumption of prenatal vitamins have been suggested 2. The prognosis for World Health Organization (WHO) grade I ependymoma, of which, MPE is a sub-type, is very good. The 1-year and 10-year survival rates for WHO Grade I ependymoma are 100% and 93% respectively 2.

CASE REPORT

Past Treatment History:

Patient had history of having undergone lumbar spine surgery 1 year ago for complains of low back pain and weakness of bilateral lower limbs. Patient had undergone a D12-L1 laminectomy with excision of a conus intradural extramedullary tumor at L1-L2 level. (Figure 1) Patient had no associated symptoms of bowel, bladder dysfunction at this time. Tumor HPE was suggestive of a Myxopapillary ependymoma (Figure 2). Operative notes suggested, a subtotal resection of the tumor was done owing to the adherence of tumor to the cauda equina roots. A post-operative MRI scan dorso-lumbar spine was performed postoperatively after 1 month which showed an irregular heterogeneously enhancing intradural mass lesion extending from D12-L1 level to L1-L2 level. Lesion was involving the filum terminale and cauda equina roots (Figure 3).

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Fig.1: Initial pre operative mri t2 sagittal showing initial lesion at l1-l2 level
Patient was planned for post operative radiotherapy for residual tumor. Patient was referred to radiation oncology for adjuvant radiotherapy. A treatment plan offering best therapeutic benefit both in terms of optimum dose to target tissue and maximum sparing of the uninvolved organs was selected. Patient was given a total of 54 Gy over 30 cycles (45 Gy/25 # to PTV and 9Gy/5# to the PTV1) on DMX Varian Linear Accelerator. Patient tolerated the adjuvant radiotherapy well and noticed a marked improvement of pain and weakness symptoms 1 month after the radiotherapy. Follow up scans were done 1 month and 6 months after radiotherapy. A small residual lesion was visualized at D12-L1 level on both the scans. However there was no evidence of any new lesions in the spinal cord. Patient had an improvement in the low back pain and the lower limb paresis after radiation therapy. However 8 months after radiation therapy patient started experiencing the low back pain and the lower limb paresis symptoms associated with signs & symptoms of cauda equina syndrome.

**Present Treatment History:**

A 24 year male presented to the neurosurgery OPD with the complaints of low back pain radiating to both lower limbs for the past 5 months. Patient also had complaints of weakness of bilateral lower limbs (left more than right) for the past 2 months. Patient noted decreased sensation over the lower limb and buttocks for the past 1 month. There were episodes of intermittent dribbling of urine and constipation for the past 1 month. A thorough neurological examination was done. Patient was found to have left foot drop with weakness of left knee extensors, bilateral ankle flexor/extensor weakness with bilateral extensor hallucis longus weakness. Patient had decreased muscle tone left lower limb with thinning of the left lower limb. The knee, ankle and plantar reflexes were bilaterally absent. Patient had normal abdominal & cremasteric reflex with absent bulbocavernous and anal reflex. Anal tone was lax. Patient underwent a MRI dorso lumbar spine on present admission as his examination was suggestive of cauda equina region pathology.

MRI showed an irregular enhancing intradural mass lesion along anterior aspect of lower cord and conus extending from D12-L1 level up to the L2 mid vertebral body level. Patient also had an irregular enhancing lesion within spinal canal from L4 to S2 level consistent with drop metastasis (Figure 4). This new lesion developed far below the previous anatomical location of previous tumor. This lesion was seen extending to epidural space with involvement of the posterior body of L4, L5 S1 & S2 vertebra. Patient underwent re exploration, a D11-L3 laminectomy was done with debulking of tumor. Gross total resection was not possible owing to dense adhesion of tumor capsule with surrounding lumbosacral nerve roots. Dural closure was done with macroscopic tumor still present. HPE was suggestive of myxopapillary ependymoma, similar to previously resected tumor.
Patient duly received a radiation oncology referral post operatively and was taken up for local re-irradiation of residual tumor. Patient received a total of 54 Gy over 30 cycles (45 Gy/25 # to PTV and 9Gy/5# to the PTV1) on DMX Varian Linear Accelerator.

Patient had a Nurick Grade 4 (Difficulty in walking needing assistance) prior to radiation therapy which improved to Nurick Grade 3 (Difficulty in walking not needing regular assistance) with no improvement in bladder dysfunction symptoms. The patient had a gradual worsening in motor power 6 months after radiotherapy with worsening of low back pain.

Post radiation MRI scan was done which was suggestive of lower dorsal spine D11-D12 spine involvement with multiple nodular lesions within spinal canal from lumbar to sacral region suggestive of metastatic deposits (Figure 5)

DISCUSSION

Ependymoma is an uncommon central nervous system neoplasm, usually seen in children and young adults. Myxopapillary ependymoma (MPE) was first described as a variant of ependymoma by Kernohan in 1932. MPE is usually located in the cauda equina and filum terminale regions, and characterized by the production of mucin and formation of papillae. Cauda equina myxopapillary ependymomas are assumed to arise from the ependymal cell nests normally present in the filum terminale. The reason for the myxopapillary appearance is not well understood; ultra structural evidence shows that the myxoid material represents excess basement membrane material deposition around the tumor cells induced by the abnormal juxtaposition of collagen that abuts directly onto ependymal cells in the normal filum terminale. MPE is a slow-glowing tumor and is designated histologically as grade I neoplasm according to the 2007 World Health Organization classification.

Pathologically there are four types of ependymomas, namely: typical, anaplastic, subependymoma and myxopapillary. Among these, mainly anaplastic ependymomas are known to spread to multiple sites in the spine and cranial cavity. Rare cases of tumor spread from the spine retrograde to the cranium and extraneural spread to liver and lungs from the spine have been reported in literature. Peripheral metastasis has also been described, including one case of local tumor reoccurrence forty years after initial therapy with concomitant metastases in the lungs, liver, and lymph nodes.

Single institution retrospective studies suggest, younger age, high tumor grade and failure to achieve GTR are poor prognostic factors for patients with ependymoma. However larger population based study from the surveillance, epidemiology end results database suggested that older age rather than younger age group was associated with a poorer prognosis. Recent investigations have started focusing on newer pathologic and immunohistochemical markers of prognosis. A study has shown increased mitotic index, increased cellularity, presence of Ki-67, and presence of cyclin D1 as independent markers of mortality in ependymomas. However, no such studies have focused solely on MPE. The most common presenting symptoms of spinal MPE include pain, weakness, and sensory changes. Symptoms of bowel, bladder, and/or sexual dysfunction may be seen depending on the tumor location. MPE is a benign tumor but has various growth patterns and metastatic modes. Metastatic spread of MPE is infrequent and, when it does occur, the disease tends to spread in the rostral direction within the central nervous system. Intracranial metastasis is the most common site for MPE spread. A recent review described 19 such cases and found a variety of intracranial sites involved including the telencephalon, the brainstem, and the cerebellum. The time span to presentation of metastasis also varied wildly, from 1 month to 13 years following primary tumor resection in one series. Previously published reports of drop metastases in patients with MPE are scarce. Reports mention cases where drop metastases were discovered simultaneously with the primary tumor. Metastases have been reported up to 20 years after initial treatment, emphasizing the role for extremely long-term follow up in patients treated for MPE.

One case reported drop metastases from a T11-L2 primary MPE to the distal thecal sac, similar to the patient reported here. However this patient was a 13 year old girl and the metastasis was found on the initial MRI scan. An additional case report describes a 22-year-old man who presented with MPE lesions in the third ventricle and sacral spinal canal; he was hypothesized to have a drop metastasis by the authors, though both lesions were again found simultaneously.
There has been only one previously reported case where an adult patient presented with secondary MPE drop metastases following an initial subtotal resection and post-operative radiotherapy of the tumor bed of a primary MPE similar to our case⁴. To our knowledge this is the only reported case in literature of a MPE with interval drop metastasis presenting as cauda equina syndrome.

**CONCLUSION**

The various cases mentioned in the recent literature suggest a higher incidence of CNS dissemination of MPEs than previously thought. This holds true at the time of diagnosis and during the subsequent follow up. Screening of the entire CNS axis at the time of diagnosis and during follow up is recommended. The limited data on treatment of MPE indicates, radiotherapy is efficacious for residual tumor following subtotal resection and for metastatic disease. Long-term survival has been documented in MPE patients though continued follow up is warranted to look for interval metastasis.

**REFERENCES**