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Intramedullary Spinal Cord Ependymoma after Posterior Fossa Ependymoma Resection: A Case Report

Case Report
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We experienced a case in which multiple spinal cord dissemination was found 6 years after resection of the fourth ventricle ependymoma. A 17-year-old boy had undergone a subtotal resection of the fourth ventricle ependymoma and radiation therapy to the posterior fossa when he was 12 years old. Follow up CT head detected no residual tumor until he complained of back pain and pain in right thigh 6 years after surgery. MRI revealed multiple mass at D10-12 and L2-3. He underwent laminectomy and excision of tumor. This case reminds us to the necessity for long term radiological follow up including the spinal cord even in benign ependymoma, although it is still not clear for how long and how often we should do it.

Key Words: ependymoma, dissemination, spinal cord

Intracranial ependymomas constitute approximately 3-5% of all intracranial tumours.[3] According to WHO, 4 major subtypes occur, including ependymoma, anaplastic (malignant) ependymoma, myxopapillary ependymoma and subpendymoma.[2] Prognostic factors such as extent of resection, age of the patient, tumor location, histological composition and the role of the adjuvant therapies still remain controversial1 Intracranial ependymomas may spread by local infiltration in the surrounding brain or by dissemination through the cerebrospinal fluid. The true incidence of cerebrospinal dissemination of ependymomas is still unknown. It is commonly known that especially high grade ependymomas tend to disseminate few years after the initial diagnosis. Late recurrence of low grade ependymomas is not unknown, but unusual.[3]

The five year survival rate of patients with low grade tumors was 45-78%, compared to 10-48% for patients with high grade tumors.[5]

We experienced a case with multiple spinal cord disseminations, 6yrs after partial removal of the fourth ventricle low grade ependymoma. This case emphasizes that prolonged neurological and imaging review is reasonable. Furthermore as a matter of fact that long dormant period may be considered as one of ependymoma's behavioural dynamics.

Case Report

A 18-year-old male was admitted to the Neurosurgery, BP Koirala Memorial Cancer Hospital (BPKMCH) in 2016 with a history of back pain for 4months and radiating of to right thigh. There is history of suboccipital craniectomy and total excision of tumor on 11/2/2066 at other hospital. The tumor was diagnosed as grade II Cellular Ependymoma. He was referred to our hospital for radiotherapy. Postop MRI of brain was done in June 2009 (1 month after surgery) and showed residual tumor.CSF cytology was negative. He received 50.4 Gy 30 fraction Radiotherapy in posterior fossa on 8/24/2009. After radiation there was no residual tumor in CT head. Neurological examination was normal. MRI of dorsolumbar spine showed mild contrast

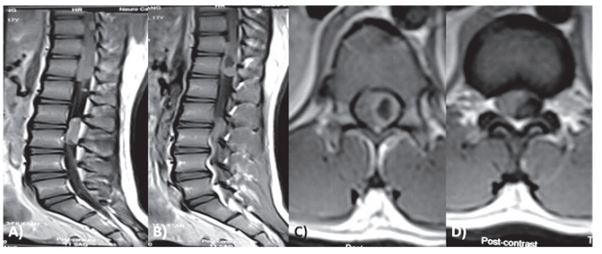

Figure 1: Preoperative MRI T1 of dorsolumbar spine with contrast, sagittal and axial images, showing two contrast enhancing intradural lesions at D10-12 and L2-3

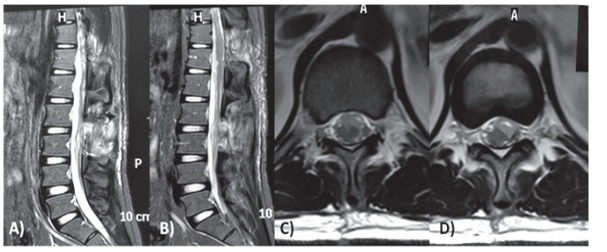

Figure 2: Postoperative MRI of dorsopumbar spine, T2 saggital and axial images, showing complete excision of both the lesions

enhancing solid and small non enhancing cystic lesion displacing conus medullaris toward right side at D12 and L2/3 **(Figure 1)**.

He underwent D10/12 and L2/3 laminectomy and total excision of intramedullary tumor on 20/11/2072 **(Figure 2)**. Postoperatively he complained of weakness of left lower limb with power 3/5 on hip flexion and 4/5 on knee extension and hospital stay was 12 days.

On follow up after 1 month he complained of burning micturition and weakness of left lower limb power 4/5, reflex 3+ on left ankle, decreased sensation on medial part of foot and lateral part of left leg. Wound was healthy. HPE showed Ependymoma Gr II.

Discussion

Proliferation and dissemination kinetics of ependymomas still remain unpredictable. Previously reported extremely rare events emphasized that ependymomas should be followed for a long term period. But the frequency and extent of radiologic follow up is still controversial.[3]

Lyons et al reviewed the literature published before 1990 and found that only 3% of 219 low grade infratentorial ependymoma developed spinal seeding. Vanuytsel et al also reviewed the literature and found that high grade tumor, infratentorial location and residual tumors at primary site were risk factors for seeding. Although most cases of dissemination became symptomatic within five years concomitantly with local recurrences a few cases disseminated later.[5]

Salazar et al reported late recurrence of ependymoma to be rare (90% of them occur before four years after initial diagnosis.[4]

Surgical treatment

The treatment of choice for intracranial ependymomas remains resection as extensive as possible which prolongs OS and decreases the rate of CSF dissemination.[6]

Of particular interst is the fact that 83% of the patient with gross total tumour resections never developed disseminated disease, where as 71% of the patients with subtotal resections demonstrated dissemination. The importance of total resection at the initial surgical attempt must be emphasized because a patient with residual tumour is 5.3 times more likely to suffer from disseminated disease eventually ($p = 0.02$, chi square).[1]

Adjuvant therapy

Considering the fact that intracranial ependymoma relapse occurs predominantly at the primary tumor site radiotherapy should be focused on local control, especially for Gr II ependymoma.[6]

Benefits of postop radiation are controversial for Gr II ependymomas. When low grade ependymomas are completely resected, the prognosis is fairly good without radiation. Thus radiotherapy is indicated in cases of malignant ependymoma and partially resectable primary or recurrent benign ependymoma.[6]

CSF Dissemination

Rezai et al reported that younger patients whohad undergone subtotal resection or harbored high grade tumour had a substantial risk for suffering disseminated disease during their clinical course. Thus it is very important to diagnose the histological characteristic of ependymomas accurately to predict the future risk of CSF dissemination, which is almost always fatal.[6]

Surveillance with periodic contrast enhanced MRI is mandatory at least once every 3 months even after imaging proven complete remission.[6]

Conclusion

Since partially resected low grade ependymomas can recur frequent imaging is necessary to find out local recurrence and spinal dissemination.

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