Case Report

Recurrence of Ependymoma in A Young Adult

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Abstract

We report the case of a young man of 28 years, who was hospitalized with complaints of headache for 5 days and weakness in both upper and lower limbs for 10 days. He has a history of Ependymal excision at 4th ventricle in the past. The young man was found to have a recurrence of Ependymoma with infiltration to the spinal cord after doing MRI of brain and spinal cord. Ependymoma is a rare type of brain tumor. It arises from Ependymal cells which line the spinal cord and ventricles of the brain. Ependymomas can develop in both children and adults. Ependymal tumors represent approximately 1.6% of all primary central nervous system tumors. Occurrence of Ependymal tumor in both brain and spinal cord is very rare, this review emphasizes that recurrence of Ependymoma after surgery can occur affecting both brain and spinal cord.

Key words: *Ependymoma, Recurrence, Spinal Metastasis.*

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Introduction

Ependymomas are glial tumors that arise from ependymal cells lining the ventricles and central canal of the spinal cord.[1] They account for approximately 8–10% of all central nervous system (CNS) tumors in children and young adults. Among these, posterior fossa ependymomas—most commonly found in the fourth ventricle—are a significant subtype, especially in pediatric and young adult populations. While initial management typically includes maximal surgical resection followed by adjuvant radiotherapy, these tumors are notorious for recurrence, sometimes at distant sites within the neuroaxis due to cerebrospinal fluid (CSF) dissemination.[2-4]

Spinal drop metastases or secondary spinal involvement from an intracranial ependymoma, though uncommon, represent a serious complication and a therapeutic challenge. Recurrent disease in the spinal cord can lead to severe neurological impairment and significantly affect the quality of life, necessitating timely diagnosis and multidisciplinary management. This case report highlights a rare instance of spinal cord recurrence of a posterior fossa ependymoma in a young adult, several years after the initial treatment, emphasizing the importance of long-term surveillance and tailored treatment strategies in recurrent ependymomas.[5,6]

Case Report

A 28 years young man was brought to the emergency department of TMC and RCH, Bogura with the complaints of headache for 5 days and weakness in both upper and lower limbs for 10 days. He gave the history of Ependymal excision at 4th ventricle 15 days back where he had suffered with surgical site infection. While I examined the young man, all the vitals were normal, deep tendon reflexes were absent and Planter response was also absent bilaterally, tone of the muscle was diminished, power was reduced to 2/5 on both limbs, Pupil was reacting to light. His milestone of development was normal. There was no facial asymmetry and no any history of involuntary movements during admission. Then the routine investigations were done where CBC revealed Hb% reduced to 9.0 gm/dl, ESR was raised to 76mm. Chest xray, ECG, serum creatinine, serum electrolytes revealed no any abnormalities. Then the MRI of brain was done which revealed post-operative state of 4th ventricular ependymoma, residual disease involving left side of inferior cerebellar vermis, superior cerebellar peduncle, left middle cerebellar peduncle, right cerebellum, presence of lesions in pineal gland region, compression on 4th ventricle obstructive dilatation of 3rd-lateral ventricles. (Figure 1) (Figure 2) & (Figure 3). Histopathology report shows the WHO grade-2 Ependymoma.

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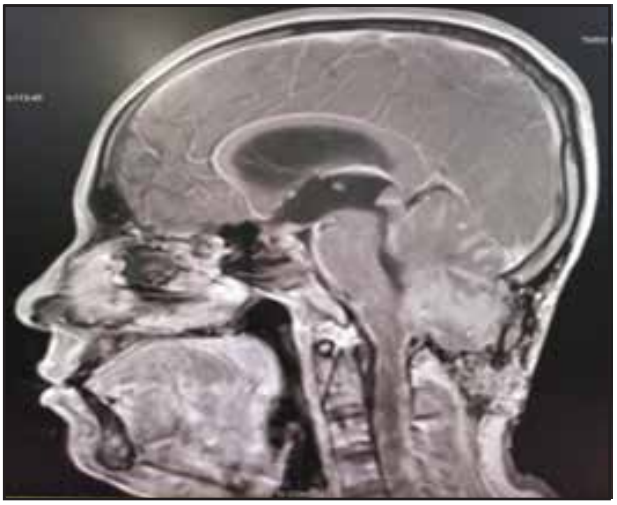

Figure 1: Coronal contrast MR scan showing enhancing lesion in cerebellum extending into peduncle.

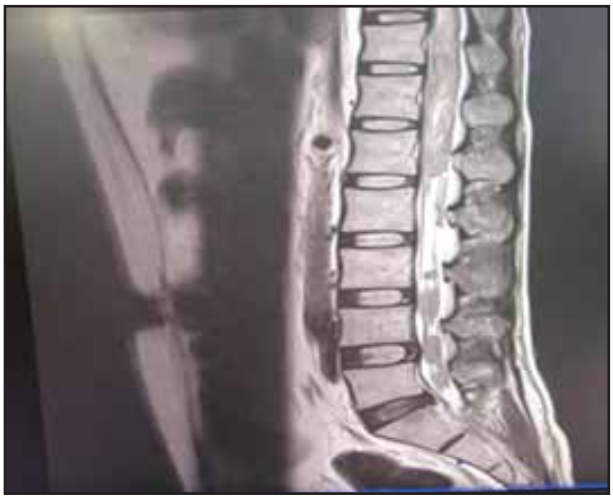

Figure 2: Sagittal T2WI MR image showing iso to mixed hyperintense lesion in lower part of spinal cord and along visualized nerve roots.

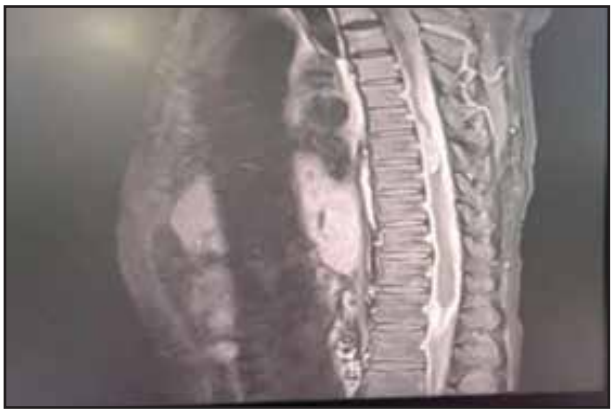

Figure 3: Sagittal T2WI MR image showing mixed hyperintense lesion in Dorsal part of spinal cord.

Discussion

An Ependymoma is a neuroepithelial brain tumor that arises from the Ependymal tissue (tissue of central nervous system). The location of the Ependymoma is usually intracranial in case of pediatric groups and in spinal in case of adults. The common intracranial site of Ependymoma is fourth ventricle. The cause of Ependymoma is unknown, it is assumed that it may be due to gene mutation. Researchers have found that people with neurofibromatosis type-2 are prone to develop Ependymoma.[2, 4]

Central Brain Tumor Registry of USA shows annual incidence of Ependymoma ranges from 0.29 to 0.6 per 100,000 persons3. Males are slightly more affected than females. Ependymoma makes up of about 5% of adult intracranial gliomas and up to 10% in case of childhood tumor of central nervous system. Ependymoma most often occurs at the age of 1 to 5 years of life intracranially and in adults spinal ependymoma is more common usually at the age of 35 to 45 years of life which especially occurs in the filum terminale of spinal cord where they have myxopapillary histology.4 In children, these tumors are likely to be recurrent even after treatment which is usually in the same spot of original tumor. There also appears to be a racial disparity, with an incidence rate per 100,000 of 0.40 in white versus 0.27 in African Americans.[5]

Ependymal tumors are classified by World Health Organization (WHO) according to their histology into three distinct grades of malignancy: WHO grade I, II and III. The sub-types of Ependymoma with their location and molecular features can predict better prognosis than grade alone. According to 2021 WHO the main sub-types of Ependymoma are Supratentorial ependymoma(ZFTA fusion positive), Supratentorial ependymoma(YAP1 fusion-positive), Posterior fossa group A (PFA) ependymoma, Posterior fossa group B (PFB) ependymoma, Spinal ependymoma, Spinal ependymoma (MYCN amplified), Myxopapillary ependymoma, Subependymoma.[5,6]

Clinically Ependymoma presents with severe headache, vomiting, visual loss, diplopia, nystagmus, epilepsy, drowsiness, papilloedema, constipation, gait change (rotation of feet when walking), back flexibility, irritability, insomnia, back or neck pain, muscular weakness, dizziness, difficulty in urination.[3] The most common presenting symptoms of both intracranial and spinal Ependymoma is pain. Changes in personality, mood and concentration can be early indicators. MRI with contrast enhancement is the choice of diagnosing for Ependymal tumors. Intracranial Ependymoma appears as a well-circumscribed mass lesions having heterogeneous appearance on T1 and T2. DWI may be useful for differentiating pilocytic astrocytomas, medulloblastomas and ependymomas in posterior fossa. Whereas spinal cord Ependymomas display more distinct borders than diffuse astrocytomas. The formation of cyst and T2 hypointensity of the cyst wall due to blood products are suggestive of Ependymoma.[4]

The mainstay treatment of Ependymoma is surgery both in case of children and adults, where complete resection of the tumor has better outcome and better survival rate than partial resection. The retrospective studies has shown the improvement in survival rate who has gone surgical resection of tumor along

with adjuvant radiation therapy.6 In adults with anaplastic ependymoma WHO grade III and incomplete resection of WHO grade II ependymoma radiotherapy is employed. Chemotherapy is given for younger children of age less than 12 months and also for adults with the recurrent disease where further surgery and irradiation are no longer possible.[2,5,6]

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