Case Report

Paediatric Ependymoma- A Rare Case Report

Dr. Rishab Singh Deo[1], Dr. Deshpande V. G.[2], Dr. Machale G. B.[3], Dr. Suresh Waydande[4], Dr. Sunil Natha Mhaske[5]

[1]Junior Resident, [2]Associate Professor, [3]Assistant professor, [4]Professor and Head, Department of Paediatrics, [5]Professor Paediatrics and Dean, DVVPF's Medical College and Hospital, Ahilyanagar-414111, Maharashtra, India

Abstract:

Ependymomas are rare central nervous system (CNS) tumours originating from ependymal cells lining the ventricles and spinal cord. This case report details a unique presentation of a sellar ependymoma in a 2-year-old male presenting with acute symptoms, including inability to walk and vomiting. Magnetic resonance imaging (MRI) revealed a large sellar and suprasellar mass causing hydrocephalus. The patient underwent a parieto-occipital craniotomy with complete tumour excision and placement of a ventriculoperitoneal shunt to manage hydrocephalus.

Key words: Paediatric ependymoma, Sellar tumour, Intracranial tumours, Hydrocephalus management

Introduction:

Ependymomas are tumors in the central nervous system that develop from the ependymal lining of the ventricle and spinal cord. They account for around 2% of all intracranial tumors, with the majority affecting children. In adulthood, they account for 3–5% of glial tumors. [1,2] The majority of ependymomas form infratentorially, although they can also emerge supratentorially and mostly along the ventricle walls, which is compatible with the location of the ependymal cells from which they originate.[3] Ependymomas form when cells called ependymal cells develop uncontrollably. Ependymal cells surround the ventricles and pathways of the brain and spinal cord, producing cerebrospinal fluid (CSF), which serves as a cushion to protect the brain and spinal cord from damage. Ependymomas can spread when the CSF transports ependymoma cells to different areas of the CNS. Ependymomas seldom spread beyond the CNS.[4] The symptoms of ependymoma vary depending on the age of the kid and the location of the tumor. Children's symptoms include weariness, vomiting, eyes that remain down, irritability, slower growth or increased head size, loss of balance or difficulty walking, neck discomfort, seizures, frequent headaches, and fuzzy vision.

We present a case of sellar ependymoma (SE) in a 2-year-old male with a history of incapacity to walk and vomiting for 15 days. Magnetic resonance imaging (MRI) revealed a mass in the sellar and suprasellar regions of the brain and management modalities is discussed

Case Report:

2 years an old male child reported to department of Paediatrics with chief complaints of inability to stand and walk for 5 days and episodes of vomiting for 2 days. Child was apparently alright 15 days back when child's parents noticed that the he was unable to stand and walk which he used do previously. There was no history of trauma, fever or any other muscle weakness, loss of consciousness. For the above complaints they visited physician who suggested to do a MRI Brain plain imaging and was referred to higher center. Child also started to have episode of vomiting mostly early in the morning which was non projectile in nature

Currently on examination, General condition of patient was fair, with Heart Rate 108 bpm, Respiratory Rate 26/min with Spo2 97 %, Blood pressure 88/64 mmHg was comfortably sitting on mother's lap. Neurologically child was conscious, he responded to painful stimuli with crying.

Corresponding Author: *Dr. Rishab Singh Deo*

Email ID: *rishabdeo241297@gmail.com*

Address: *Department of Paediatrics, DVVPF's Medical College and Hospital, Ahilyanagar - 414111, Maharashtra, India*

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He was oriented and followed colorful objects., child's muscle nutrition was normal, power in upper limb is 5/5 and lower limb power was 3/5, tone in upper limb was normal whereas tone in lower limb was hypotonia, sensory system was normal, cranial nerve examination was normal.

Child was admitted in PICU started with antibiotics; basic blood investigation was also sent. MRI Brain plain and contrast imaging with spectroscopy was done which was suggestive of 58x50x71mm large solid heterogeneously enhancing cauliflower like lesion involving vermis 4th ventricle and extending to infra and supratentorial space causing moderate hydrocephalus with raised level of choline in lesion mass. Hence child was referred to neurosurgeon, where child was operated. A parieto-occipital craniotomy was performed. When the dura was opened during the operation, the extra-axially located, well-circumscribed, dirty yellow-white tumor was easily dissected from the surrounding tissue. The tumor was excised entirely by microdissection and ventriculo peritoneal shunt was done. There were no intraoperative and postoperative complications documented. Histopathological examination revealed a glial tumor with focal infiltration and an extensive, expansive growth pattern in the brain parenchyma suggestive of grade 2 ependymoma. Postoperatively child was shifted back to PICU where child was kept intubated on mechanical ventilator, later gradually as sensorium improved, he was extubated and was taken on oxygen nasal prongs, was weaned off later. Patient was monitored for vitals, input and output. Child was initially kept NBM post operatively later was gradually started with liquid diet and later shifted to soft diet. Child was started with limb physiotherapy and was started to ambulate. No metastasis was found in additional radiological imaging in terms of seeding metastasis. Eventually as condition improved child was discharged and called for follow up. At 6-month follow-up, the patient's clinical condition subsided utterly, and a magnetic resonance (MRI) showed no evidence of residual tumor.

Discussion:

Pediatric ependymomas are most commonly located in the posterior fossa, although they can also occur in supratentorial regions, as observed in this case.[5,6]

This report presents a unique instance of sellar ependymoma in a 2-year-old child who presented with acute symptoms, including an inability to walk and persistent vomiting. Such symptoms are consistent with increased intracranial pressure and brainstem compression, which are common in posterior fossa tumors. The rapid onset of neurological deficits in this patient underscores the aggressive nature of the tumor and the critical need for prompt diagnosis and intervention.

Magnetic Resonance Imaging (MRI) is the imaging modality of choice for the diagnosis of ependymomas. In this case, MRI revealed a large mass in the posterior cranial fossa with dilation of the third and fourth ventricles, indicating obstructive hydrocephalus.

The surgical approach, involving a parieto-occipital craniotomy, allowed complete excision of the tumor with no intraoperative complications. Complete tumor resection significantly improves survival outcomes, particularly in pediatric patients. Histopathological examination confirmed a World Health Organization (WHO) Grade II ependymoma, which typically has a favorable prognosis compared to higher-grade variants. Postoperative care, including the use of a ventriculoperitoneal (VP) shunt, was essential in managing hydrocephalus and preventing recurrence of symptoms.

The patient's postoperative course was remarkable for recovery, with gradual improvement in neurological function. The absence of residual tumor on follow-up imaging six months post-surgery is a positive indicator, as the extent of resection is closely correlated with long-term survival. It is also noteworthy that no evidence of cerebrospinal fluid (CSF) dissemination or metastatic spread was observed, which is a critical concern in ependymomas due to their potential to disseminate along the CSF pathways (ependymoma case).

Despite advancements in surgical and adjuvant therapies, the management of pediatric ependymoma remains challenging, particularly in cases with incomplete resection or anaplastic features. The role of adjuvant radiotherapy in children under the age of three is controversial due to the potential for long-term neurocognitive deficits; however, it may be considered in cases of residual disease or recurrence. In this case, the decision to monitor without immediate postoperative radiotherapy appears justified given the complete resection and the young age of the patient.

Conclusion:

This report highlights the aggressive nature of paediatric ependymomas, timely intervention and the critical need for vigilant follow-up to ensure long-term survival and quality of life.

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