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Hydrocephalus Secondary to Intraventricular Myxopapillary Ependymoma: Case Report

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Abstract

Ependymomas are a somewhat diverse category of glial tumors that often develop from the lining of the brain's ventricles, or the spinal cord's central canal. They make up 5% of all neuroepithelial tumors, 10% of paediatric brain tumors, and up to 33% of brain tumors in children under the age of three. Hydrocephalus is one of the complications, and it can be identified as progressive macrocephaly or increasing head circumference crossing percentiles, nausea, vomiting, poor appetite, irritability, and regression of developmental milestones.

Keywords

Hydrocephalus, Myxopapillary Ependymoma (MPE), Paediatrics, Neurosurgery, Glioma, Intracranial Hypertension

1. Narrative

We present a case of a 5-month-old Caucasian female with complaints of hypersomnia and vomiting. Due to relentless vomiting, the patient quickly manifested signs of severe dehydration, somnolence, was lethargic and adynamic, hence intracranial hypertension was suspected. Contrast and non-contrast Magnetic Resonance Imaging (MRI) was obtained, which showed evidence of a cyst located in the frontal horn of the right ventricle with an abnormal formation. This was identified as the route of the hydrocephalus. Additionally, a contrast imaging study revealed a myxopapillary ependymoma (MPE) compressing

the ventricular system. A ventriculoperitoneal shunt was placed, and symptoms improved. Verbal consent was acquired however, all documentation could not be provided.

Intracranial ependymomas are an uncommon subclass of glial tumors, which have been difficult to diagnose amongst neuroradiologists. They can be confused for an abscess, astrocytoma, medulloblastoma, cerebral neuroblastoma, or choroid plexus papilloma. The presence of this tumor led to the dilation of the ventricles, thus causing the transient accumulation of CSF within the neonate's brain. The tumor's growth induced this patient's hydrocephalus and other clinical abnormalities. Due to the peritoneum's ability to absorb fluid, the ventriculo peritoneal (VP) shunt is the diversion technique that is most frequently used to cure this neurological disorder.

2. Introduction

Hydrocephalus is defined as a neurological disorder caused by the excessive buildup of cerebrospinal fluid (CSF) within the ventricles of the brain. This may cause the ventricles to widen causing compression of surrounding brain structures [1]. This was first noted by the scientist Walter Dandy in the 19th century, who named the two conditions communicating and non-communicating hydrocephalus [2]. The histological appearance of MPEs serves as the basis for the term "myxopapillary." Their branching vasculature causes them to manufacture mucin and create tumor cells that are organized in papillae. They can occasionally spread with CSF, and 14 - 43% of cases of intracranial secondary lesions in the central nervous system are observed [3].

This paper highlights the significance of prompt diagnosis and treatment, especially the implantation of a ventriculoperitoneal shunt, in the management of this uncommon subclass of glial tumors in paediatric patients. Compared to adults, MPE is less common among the paediatric population [4], and that is why this case is presented as unique. The objective of this paper is to discuss the case study of a 5-month-old child who had hydrocephalus as a result of intracranial ependymoma symptoms, specifically a myxopapillary ependymoma.

3. Clinical Presentation

Symptoms of hydrocephalus differ by age. Adults typically present with a headache, blurred vision, nausea, unpleasant changes in day-to-day activity as well as focal neurological signs [1]. Infants, however, have different physiology and are not able to communicate as clearly as adults, leading to the condition presenting with different symptoms. These may include progressive macrocephaly or increasing head circumference crossing percentiles, nausea/vomiting, poor appetite, irritability, and regression of developmental milestones [5].

Ependymomas are a somewhat diverse category of glial tumors that often develop from the lining of the brain's ventricles or the spinal cord's central canal. They make up 5% of all neuroepithelial tumors, 10% of paediatric brain tumors,

and up to 33% of brain tumors in children under the age of three. Although ependymal tumors can develop anywhere in the neural axis, their distribution and molecular characteristics tend to vary depending on the anatomical compartment in which they are discovered and are categorized into Posterior fossa (PF) (60%), Supratentorial (ST) (30%), and Spinal cord (10%) subtypes (**Figure 1**) [1] [2] [6].

Site of tumor	Subtype	Anatomically-related ependymoma genes	Posterior fossa groups
Supratentorial	• Ependymoma • Subependymoma	• ZFTA Fusion • YAP1 Fusion	N/A
Spinal	• Ependymoma • Subependymoma • Myxopapillary ependymoma	• MYCN non-amplified	N/A
Posterior fossa	• Ependymoma • Subependymoma	• H3K27me3	• Group PFA (1) • Group PFB (2)

1: H3K27me3 los; 2: H3K27me3 retained.

Figure 1. WHO 2021 classification of ependymomas.

4. Classification System

According to the histopathologic traits, the 2021 categorization allows for the assignment of grade 2 or grade 3. Grade 2 or 3 ependymomas tend to be anaplastic in nature (**Figure 2**), which reduces life expectancy amongst diagnosed patients. PF ependymomas differ from ST ependymomas in that they lack recurrent hallmark genetic events [5] [7]; yet they can be divided into 2 main groups according to their epigenetic characteristics: Histone H3 K27-trimethylation is absent in PF type A (PFA) cancers, whereas histone H3 K27-trimethylation is highly expressed in PFB tumors [8].

5. Management Strategies

Management is usually surgery followed by radiation therapy [9]-[11]. Chemotherapy has thus far failed to show a significant effect on overall patient survival in both adults and children. With current facts stated, it is still controversially discussed for its potential role in radiotherapy deferral strategies in infants [12]. Clinical therapy of primary cerebral ependymomas (WHO grade II/III) is difficult since they frequently appear with locally invasive development patterns. The size of the resection is the most reliable independent prognostic predictor, and surgery is crucial for local tumor management [13] [14].

6. Case Report

We present a case of a 5-month-old female with complaints of hypersomnia and vomiting. Feeding was impaired due to vomiting. The child was conceived following in vitro fertilization after the mother suffered from infertility for 20 years.

The mother was 38 years old, and this was her second pregnancy but first delivery. Twin delivery was expected though she had suffered from a spontaneous abortion years before. During prenatal care, it was noted that the fetus had severe anemia and polyhydramnios. The mother contracted COVID-19 during the third trimester of pregnancy which called for a cesarean section to be performed at 36 weeks of pregnancy. The patient was the second born and had a birth weight of 2.1 kg and a height of 44 cm. Her APGAR score was 6/7.

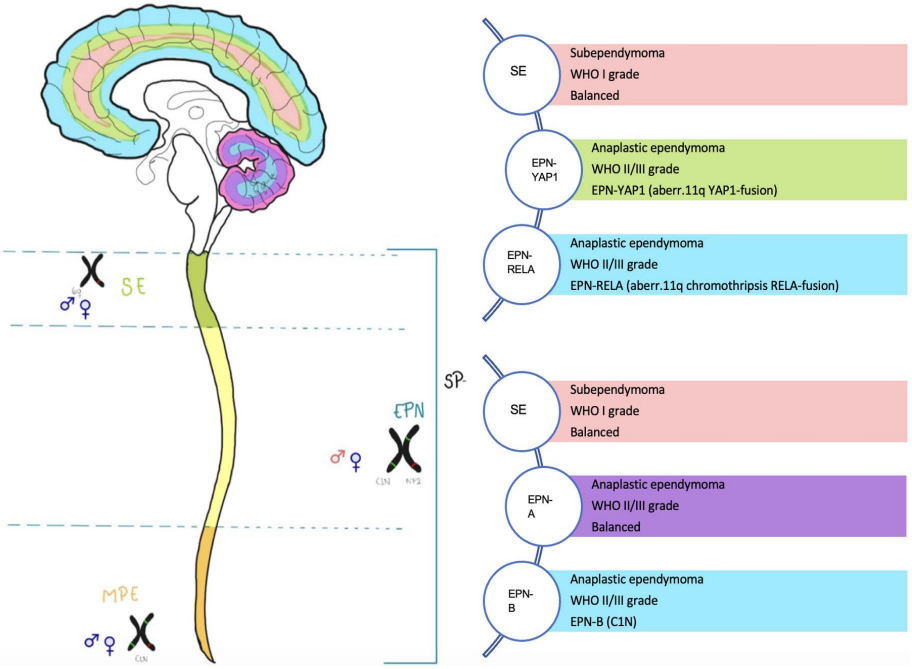

Figure 2. Anatomic and genetic classification of ependymomas.

Intracranial hypertension was suspected, and a neurologic ultrasound of the patient's head was performed and indicated a round-shaped cystic hollow mass with a capsule sized 41 × 43 mm in a right temporal area with delayed liquefaction and hydrocephalus at the level of the fourth ventricle. A contrasted and non-contrasted Magnetic Resonance Imaging (MRI) was obtained, evidence of a cyst located in the frontal horn of the right ventricle with an abnormal formation causing hydrocephalus, Furthermore, a contrast imaging study revealed a tumor compressing the ventricular system and further evaluation was conducted. The selected procedure was a ventriculoperitoneal shunt to drain the excess cerebrospinal fluid and decrease intracranial pressure. Tumor extirpation by craniotomy was performed and biopsy showed intraventricular MPE. The patient required mechanical ventilation which lasted 5 days and was extubated due to clinical improvement. She was discharged and close observation was conducted 3 months after intervention with no complications.

7. Discussion

MPE can be classified as a grade I glioma. Incidence ranges between 0.05 - 0.8

per 10,000 people per year. The presence of this tumor led to the dilation of the ventricles and thus, the transient accumulation of CSF within the neonate's brain, leading to intracranial hypertension. The choice of operative management for this neonate is beneficial for treating tumors obstructing the lateral ventricles, third ventricles, and those within the posterior fossa.

MPE is a rare condition; however, various genetic predispositions have been identified in the literature that increases the risk of paediatric patients developing intracranial ependymomas. A study by Yao Y *et al.*, found a gain of chromosome 1q in 20% of intracranial ependymoma cases. Additionally, the deletion of chromosomes 6q and 9 has been implicated in the development of intracranial ependymomas [15]. Hypermethylation and deletion of the CDKN2A gene, responsible for the tumor suppressor genes p16 and p14, have been observed in over 90%e of paediatric supratentorial ependymoma cases [16]. In contrast, spinal ependymomas tend to arise more in patients who possess mutations in the Neurofibromatosis type 2 (NF2) gene, a regulatory factor for the cell cycle. A study by Lee CH *et al.*, stated that mutation in the NF2 gene is responsible for 30 - 71% of sporadic spinal ependymoma cases in paediatric patients [4]. Specific genetic predispositions for MPE require further investigation, as current evidence is limited.

Due to early intervention, the prognosis for our patient after surgery was predicted to be high. A lack of treatment would have led to an increased intracranial pressure and thus, interference with the newborn's developmental milestones [9] [10]. Other treatment options include radiotherapy, where patients have demonstrated better outcomes when in combination with tumor resection, decreasing local failure and increasing global survival rates [17]. Due to the peritoneum's ability to absorb fluid, the ventriculo peritoneal (VP) shunt is the diversion technique that is most frequently used to manage intracranial hypertension. MPE is regarded as a surgical disease, with surgery representing the primary method of treatment, followed by radiation therapy as an adjunctive therapy for optimal outcomes. It is worth noting that many patients may require repeat bouts of surgery, particularly if they have initially undergone subtotal resection instead of gross total resection as a form of treatment [3].

Mutations in specific genes associated with human development have been linked to the diagnosis of MPE. However, other potential contributing factors, such as environmental and social influences, have not been extensively discussed as attributable factors, which may affect the incidence of paediatric MPE [4]. Furthermore, ongoing research requires investigating new therapies based on the genetics and molecular biomarkers of tumors, taking into consideration the differences in genes for intracranial and spinal ependymomas.

8. Conclusion

This case study emphasizes the significance of early identification and treatment for intracranial ependymomas, especially in paediatric patients who seem to have a presentation consistent with hydrocephalus. The efficacy of surgical re-

section and ventriculoperitoneal shunting in symptom relief and enhancing patient outcomes is demonstrated by their successful implementation. It is necessary to continue researching the genetic and molecular foundations of ependymoma to improve strategies for therapy and prognostic precision in clinical settings.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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