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Case Report

Eye a gateway to brain-nystagmus as a presentation of brain ependymoma

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ABSTRACT

Ependymomas are the third most common brain tumours in children, with the posterior fossa being the most frequent site. Early diagnosis can be challenging due to nonspecific symptoms. Authors report the case of a 6-year-old male who presented with a two-month history of intermittent occipital headaches and a one-month history of vomiting. Neurological examination revealed bilateral horizontal nystagmus on lateral gaze, but no gait abnormalities or other cerebellar signs. MRI of the brain demonstrated a large, heterogeneously enhancing lesion in the posterior fossa (5.7×5.2×4.3 cm), causing mass effect. The patient underwent midline suboccipital craniotomy with gross total resection of the tumour and placement of a right parietal Ommaya reservoir. Histopathology confirmed a WHO Grade 3 ependymoma. Postoperatively, the child remained neurologically intact and was discharged symptom-free. This case highlights the importance of early recognition of posterior fossa tumours in children with subtle but progressive symptoms. Timely neuroimaging and surgical intervention can lead to favourable outcomes even in high-grade pediatric ependymomas.

Keywords: Pediatric ependymoma, Posterior fossa tumour, Intracranial mass, Nystagmus

INTRODUCTION

Pediatric brain tumours are most common solid tumours in children. The incidence of pediatric brain tumours ranges from 0 to 2.11% and account for around 10-21% of total intracranial tumours.[1]

The most common pediatric brain tumours are astrocytic tumours (34.7%), followed by medulloblastomas & supratentorial tumours (22.4%), craniopharyngiomas (10.2%) & ependymomal tumours (9.8%).[2]

Brain tumours can present in the form of headaches, loss of balance, seizures, head tilt, irritability (personality change), failure to thrive, neck pain, speech problems, coordination problems, double vision, nystagmus, limb weakness, loss of bowel bladder control, voluntary control.[3] Symptoms vary depending on size and location of tumour.

CASE REPORT

We present the case of a 6-year-old male child diagnosed with a posterior fossa ependymoma. The child presented with a two-month history of intermittent occipital headaches, occurring every 2-3 days and lasting 1-2 minutes, along with vomiting for the past month. Additionally, there was a notable weight loss of 3 kg over the last two months, accompanied by anorexia. The child had no history of chronic illnesses and was developmentally normal.

On examination, the child was alert, well-oriented to time, place, and person, with a head circumference of 51 cm. Neurological examination revealed bilateral horizontal nystagmus on lateral gaze, but the rest of the cerebellar assessment was normal, with no gait disturbances. Ophthalmologic evaluation showed no signs of papilledema. Based on these findings, the child

was referred for further surgical evaluation. MRI of the brain revealed a large, heterogeneous enhancing lesion in the posterior cranial fossa, primarily on the left side, measuring 5.7×5.2×4.3 cm. The mass showed multiple foci and caused significant mass effect.

On follow up the patient had undergone midline suboccipital craniotomy for resection of the grade 3 ependymoma, with placement of a right parietal Ommaya reservoir. The patient is now symptom-free and is being discharged without any neurological deficits.

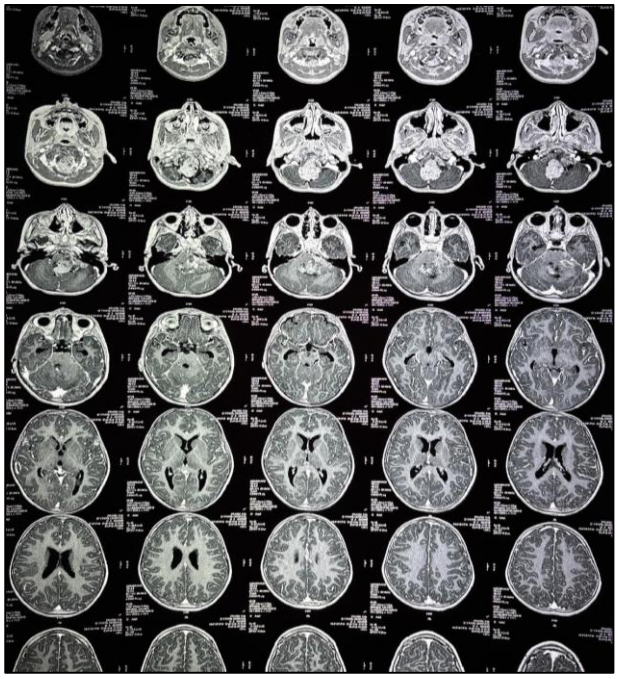

Figure 1: MRI brain -showing findings most likely suggestive of malignant neoplastic etiology likely posterior fossa ependymoma.

DISCUSSION

Ependymomas are rare central nervous system tumors, with an estimated annual incidence of 0.26 per 100,000 children.[4] These tumors originate from ependymal cells, which line the ventricular system and central canal of the spinal cord. Ependymomas are most commonly diagnosed in children under five years of age and occur more frequently in males. In pediatric populations, approximately 10% of all ependymomas are located in the posterior fossa (cerebellum), while around 25% arise in the supratentorial region.[4] There are three main anatomical subtypes of ependymomas, posterior fossa ependymomas-the most common type in children, supratentorial ependymomas, spinal cord ependymomas.

Posterior fossa ependymomas typically present with symptoms related to increased intracranial pressure, such as nausea and vomiting due to obstructive hydrocephalus. Additional features may include ataxia and cranial nerve palsies. In contrast, supratentorial ependymomas often present with headaches, seizures, and focal neurological deficits. The two most commonly reported symptoms at diagnosis are headache (61%) and vomiting (53%). Neurological deficits are present in approximately 20.4% of cases.[1] Visual disturbances, including diplopia and nystagmus, may also occur.

Nystagmus is characterized by involuntary, rhythmic, and repetitive oscillations of one or both eyes. It is a relatively frequent finding in pediatric brain tumors. A study conducted in Denmark reported that nystagmus was observed in 13.7% of children diagnosed with brain tumors, and in 83.3% of these cases, it was identified incidentally during clinical examination.[5] Supratentorial ependymoma on MRI, these tumors typically appear as well-defined masses with heterogeneous enhancement, often accompanied by peritumoral edema and cystic areas. They may exert mass effect, displacing nearby brain structures or ventricles; while in the posterior fossa, ependymomas are seen as well-circumscribed lesions arising from the fourth ventricle. These tumors exhibit heterogeneous signal intensity and post-contrast enhancement, often causing obstructive hydrocephalus and surrounding edema, with possible cystic changes.

Treatment usually begins with surgery to remove as much of the tumor as possible. If necessary, radiation or chemotherapy is used, with radiation being carefully considered in young children. Chemotherapy is commonly used for tumors that are difficult to remove or have recurred. Emerging options, such as targeted therapies and immunotherapy, may be considered for certain tumor types. Continuous follow-up care is crucial for monitoring recurrence and managing long-term effects.

CONCLUSION

Pediatric headaches, especially when accompanied by neurological signs such as nystagmus, warrant thorough investigation. Nystagmus may indicate underlying central nervous system pathology, including space-occupying lesions or other neurological disorders. A detailed clinical examination, along with advanced radiological imaging, is crucial for identifying potential causes and ensuring timely and accurate diagnosis. Early detection and intervention are key to preventing long-term complications and optimizing outcomes in affected children.

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